Chapter 15: Benign diseases of the neck

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Thyroglossal cysts

Terminology

Since it has not been established whether or not the tract connecting the thyroid gland to the foramen caecum persists as a solid tract, a hollow tube or a duct that becomes obliterated, terms such as 'thyroglossal duct cyst' or 'thyroglossal tract cyst' are best avoided and the general term 'thyroglossal cyst' used.

A sinus is an opening between an internal structure and an epithelial surface whereas a fistula is a connection between two epithelial surfaces. The term 'thyroglossal fistula' is often used but it is erroneous. Only one case of a congenital thyroglossal fistula has been described and its existence is rather tenuous. What most authors mean is a thyroglossal sinus and this is the preferred term. A fistula would suggest an opening between the base of the tongue and the skin surface of the neck and this clearly never occurs.

The discharge from a thyroglossal sinus is secreted mucus not saliva.

Embryology

The branchial arches and pharyngeal pouches develop at the beginning of the fourth week of embryonic life. Among other things the first arch forms the lingual swellings that make up the bulk of the anterior two-thirds of the tongue; it is completed by the tuberculum impar which forms a median eminence behind the lingual swellings and eventually joins them. The posterior third of the tongue is formed by the merging of the ventral portions of the second and third branchial arch.

During the fourth week the thyroid anlage forms an outpouching from the floor of the pharynx between the tuberculum impar and the posterior third of the tongue. It enlarges caudally as a bilobed diverticulum following the descent of the heart and great vessels and grows into the loose parapharyngeal connective tissue. As it moves down it leaves a tract behind.

The hyoid bone develops later and joins from lateral to medial. It is possible, therefore, for the tract to be caught in this, resulting in the tract running through the bone. More commonly, however, the hyoid rotates to achieve its adult position and draws the thyroglossal tract posteriorly and cranially at the inferior edge of the body. Apart from this 'notch' behind the body or partly included in the body, the thyroglossal tract lies ventral to the body and the thyrohyoid membrane.

There is no natural opening of the tract. The tongue and foramen caecum form later than the descent of the thyroid and the blind tract, sometimes found in association with the foramen caecum, is the lingual duct and represents the point of union between the paired anterior and posterior segments that form the tongue base.
The thyroglossal tract normally atrophies and disappears between the fifth and tenth weeks, but the caudal attachment may remain as the pyramidal lobe of the thyroid gland.

Pathogenesis

The prevalence of thyroglossal cysts depends on the author and the institution. While many authors claim it is the most common non-neoplastic neck mass and others that it represents 40% of all primary tumours in the neck, it is salutary to note that Sistrunk, whose name is still attached to the operation for removal of the cyst, reported only 31 thyroglossal cysts in a series of 86,000 general paediatric patients.

The sex distribution is equal and the age range is from birth to 70 years with a mean age of 5.5 years. Of about half the published cases, 31.5% were under the age of 10, 20.4% were in the second decade, 13.5% in the third decade and 34.6% were older than 30 years. Ninety per cent lie in the midline and 10% are to one side of the midline; of those, 95% are on the left and 5% are on the right.

If this is a developmental abnormality and thus a congenital lesion, why should almost one in three not present until the patient is over the age of 30 years? The reason is not known but two possibilities are cited. The first suggests that recurrent throat inflammation may stimulate the epithelial remnants of the tract to undergo cystic degeneration. The second possibility is a retention phenomenon. A blocked thyroglossal duct may expand to form a cyst because of an accumulation of secretion. Most proponents of this theory, however, implicate the foramen caecum in the obstruction. The lingual duct has mucous and serous glands and the cyst may be a lingual cyst and not a true thyroglossal cyst. This might account for those cysts found in the tongue above the hyoid.

In the published series, where the position is maintained, 2.1% are intralingual, 24.1% suprahyoid, 60.9% thyrohyoid and 12.9% suprasternal. This means that about one in four is above the hyoid and three out of four are below.

A cyst may form on both sides of the hyoid resembling a dumb-bell lesion. Missing part of the dumb-bell may account for some recurrences after a Sistrunk operation. Sometimes multiple cysts are found. These are probably pseudocysts because they have no real lining and are only granulation tissue with extravasated mucus. These probably arise from continual mucus production in a blocked duct that gradually balloons out at its distal end to form a cyst that ruptures into the surrounding tissue.

Sinus openings are always secondary due to spontaneous or surgical drainage after infection. Sinus formation may also occur after a 'lumpectomy' operation leaving the hyoid and part of the tract if it is mucus secreting.

Thyroid tissue is present in the cyst wall in more than 60% of cases. Its presence is most probably due to the origin of the duct in relation to thyroid tissue, but benign thyroid metastases (compare endometriosis) have been postulated. It is from this tissue that thyroid carcinoma can arise.
The epithelial lining is variable; most commonly it is pseudostratified ciliated columnar but it may be squamous. Squamous carcinoma has also been reported in these cysts.

**Clinical features**

The patient will present with either a painless or an infected lump. One death has been reported due to respiratory obstruction from an intralingual cyst. If uninfected, the cyst may be soft, fluctuant and mobile, but very often it is so tense that it seems solid and, in this state, it can be wedged between the hyoid and the thyroid cartilage and thus appear fixed. It is, however, in the midline or just to the left and very few malignant lesions are in this site. Most cysts are prehyoid or infrahyoid and, since all are attached even by a tract to the hyoid, they move both on swallowing and protruding the tongue. Mobility depends on size.

If infected, the lump will be painful, the patient will have odynophagia and the overlying skin will be red. Misguided attempts at abscess drainage here can cause problems when excision is attempted and the recurrence rate is measurably higher.

Sometimes a tract is seen or palpated from the cyst to the hyoid. Such a tract may also be found clinically if there is a sinus.

The differential diagnosis is from a dermoid cyst, an infected lymph node, lipoma, minor salivary gland tumour, sebaceous cyst, cartilaginous tumour of the thyroid, hypertrophic pyramidal lobe and choristomata.

If the cyst is intralingual, it might be a lingual thyroid and, if it is below the thyroid cartilage, it may be a thyroid adenoma. Finally, the possibility of a carcinoma in the thyroglossal remnant must be borne in mind.

It is almost impossible to differentiate a midline dermoid from an uninfected thyroglossal cyst before operation. The content of a dermoid is different. Instead of thick, viscous mucus, dermoids contain cheesy semisolid material. The epithelial lining is keratinizing squamous epithelium with skin appendages on the wall. There is no evidence of a duct nor of inflammation.

Lipomata are more ill defined at the edges, but are tantalizingly fluctuant and so may cause diagnostic difficulty. They also move on swallowing.

Some of these differences may be noted on needle biopsy.

Consideration should be given to ordering a $^{131}$I scan in all suprahyoid and infrathyroid lumps. In 65-75% of patients with a lingual thyroid, there is no other thyroid tissue and it represents a complete failure of the gland to descend. It is three times more common in females because of the increased demands of the thyroid at puberty, pregnancy and menopause. There is not the same necessity to scan patients with lumps in the usual position between the thyroid and front of the hyoid.
Treatment

Wenglowski, who performed much of the embryological study on neck cysts, first suggested that, not only the body of the hyoid, but a core of tissue between this and the foramen caecum be removed. Schlange was the first to remove the body of the hyoid, but Sistrunk still has his name applied to the present day operation. He adopted Wenglowski’s suggestion and removed a core of tissue between the hyoid and the foramen caecum.

A horizontal incision is made at the inferior border of the mass and the skin carefully dissected off the mass. Sometimes there is a close connection between the mass and the skin and cyst rupture is a possibility. This makes further dissection difficult and there is then a risk of leaving part of the wall or pseudocysts behind.

The cyst is mobilized and the tract is often found in these cysts lying low in the neck, but in the higher ones there is little point in looking for a tract in and around the hyoid. Sometimes the tract is multiple and in sections its very thin wall is sometimes striking. Furthermore, although it has been described as being in the hyoid bone, van Nostrand’s series showed the tract never to be in the bone.

The body of the hyoid between the lesser horns is divided with shears or Mayo scissors. Medial to the lesser horns there is no danger of damaging the hypoglossal nerves. The genioglossus, mylohyoid and geniohyoid are attached to the body of the hyoid bone and a segment of these muscles is removed in continuity with the bony segment. There is no possibility of identifying a tract in this core of muscle, but failure to remove the core involves the risk of recurrence. The core is removed in a line drawn at 45% to the body of the hyoid aiming at the foramen caecum. There is no need, however, to open into the oral cavity.

The infected cyst

It is unwise to operate on an infected cyst. On the other hand, these may be very tense and painful and antibiotic preparation may be poor. Aspiration of pus may improve antibiotic penetration and allow resolution with a view to later removal.

Treatment of recurrence

If the hyoid body is not removed, the recurrence rate is 85%. Even if it is removed the reported recurrence rate varies from 2% to 8%. Recurrence is easily understood if the body of the hyoid is not removed, but it is not so easily explained if Sistrunk’s procedure is performed. Possible causes of recurrence are:

(1) missing a dumb-bell cyst deep in the back of the hyoid pushing the thyrohyoid membrane back

(2) dealing with a cyst that has ruptured to form thin-walled pseudocysts

(3) rupturing a cyst and leaving part of the wall behind

(4) failing to realize that the tract may be multiple and taking too thin a core.
If Sistrunk's procedure has not been performed then revision surgery is simple and the body of the hyoid removed along with a core. If, however, Sistrunk's procedure has been carried out, further surgery is difficult and the removal of the recurrent cyst plus tract must be improvised.

**Treatment of sinus**

A sinus is due to spontaneous or surgical drainage and is always secondary. Sistrunk's operation is performed, but an ellipse of skin is removed around the sinus.

If the sinus is high in the neck then the scar is a problem. If a healing scar is mobile then hypertrophy will possibly occur. The hyoid area moves constantly during swallowing and the patient should be warned of the scar problem. Dermabrasion, or breaking the line of the scar may be necessary.

**Thyroglossal carcinoma**

Two types of carcinoma have been described - one from the thyroid elements and one from the squamous elements.

The thyroid carcinomata are the more common and over 90 have been reported. Some have been diagnosed before operation for removal of thyroglossal cyst and some have been associated with coexisting thyroid gland cancer and only seven have had metastatic neck nodes. The age range is between 6 and 81 with an average age of 39. The sex incidence is equal. The age and sex distribution is the same as for thyroid carcinoma. Eighty-five per cent are papillary adenocarcinoma and the remaining 15% are follicular adenocarcinoma, adenocarcinoma and squamous carcinoma.

With so few cases reported, no treatment plan has evolved and prognosis cannot be delineated. It does appear, however, that Sistrunk's operation plus suppressant doses of thyroxine offers a reasonable chance of a cure.

**Branchial cysts, sinuses and fistulae**

**Terminology**

It is now thought that branchial cysts, sinuses and fistulae are not all variants of the same thing originating from the branchial apparatus. The original papers linking them with the branchial apparatus did so more by acclamation than experimentation.

The cysts, also known as lateral cervical cysts, usually present in the lateral part of the neck deep to the sternomastoid at the junction of the upper third and lower two-thirds. A few cysts have a definite tract into the area of the posterior pillar of the tonsil but most have no evidence of a tract. There is doubt if these are of branchial origin.

Sinuses, or branchial pits, open along a line between the tragus and the sternoclavicular joint at the anterior border of the sternomastoid. These are almost certainly failure of completion of development of the branchial apparatus. As such, they are present at
birth and reflect failure of development of the first, second, third and fourth arches. Sinuses may also occur with an internal opening only. These, however, may not always come to light but some may produce mucus, block off and become cysts with an identifiable tract.

On rare occasions, the estimated pit is demonstrated clinically and/or radiologically to have an internal opening at the posterior tonsillar pillar. Thus with two openings between epithelial surfaces there is a fistula. No case has been described with an external opening, an inclusion cyst in the tract and an internal opening.

**Embryology**

A 2-week embryo has on each side six branchial arches, five branchial clefts and five pharyngeal pouches. These arrangements are not parallel but tend to come together at the sixth arch. The first and second arches are important, the third and fourth less so, and the fifth and sixth vestigial.

The second pharyngeal pouch forms the palatine tonsils; the second arch grows downwards on its lateral side to meet the fifth arch thus enclosing the second, third and fourth clefts forming the cervical sinus of His. By the sixth week, the branchial apparatus has disappeared having formed the ear, tongue, hyoid, larynx, tonsils and parathyroids.

**Theories of origin**

The debate as to the origin of a branchial cyst reached a climax in the 1920s and 1930s. There are four theories of origin of branchial cysts, but because of the complicated development of the neck none has been proven by embryological investigation. Most of the theories have been an attempt to correlate clinical findings with known embryological facts and none can stand close scrutiny.

**Branchial apparatus theory**

These cysts may represent remains of the pharyngeal pouches or branchial clefts or a fusion of these two elements. When branchial cysts have an internal opening, it is in the region of the tonsillar fossa indicating an origin from the second branchial pouch. Fistulae and sinuses from the second pouch would necessarily pass between the external and internal carotid arteries.

Origin from the third or fourth pouches is unlikely, as they would have to pass over the hypoglossal nerve to reach the skin and would be severed by the upward movement of that nerve during development.

A fourth arch tract would also have to pass below the subclavian artery on the right and the aortic arch on the left.

A third arch tract should have its internal opening in the pyriform fossa and a fourth arch tract below this. These have never been described, so that origin from these pouches can be discounted.
Origin from the first pouch is possible because high branchial cysts have been described lying under the parotid gland with an internal opening between the bony and cartilaginous meatus. If the branchial apparatus theory were to be upheld, a lot more cysts would be expected to have internal openings; it is a popular misconception that many branchial cysts have an internal opening. More cysts would also be expected to be present at birth, but this event has been described only once. The peak age incidence is in the third and fourth decades which is late for a congenital lesion (compare thyroglossal duct cysts).

**Cervical sinus theory**

This is an extension of the previous theory and considers that branchial cysts represent remains of the cervical sinus of His which is formed by the second arch growing down to meet the fifth. It is unlikely that this is true for those with an internal tract, since this is closed by fusion of its ectodermal lining from within towards the surface. This makes an internal opening difficult to achieve.

**Thymopharyngeal duct theory**

Cysts may be a remnant of the original connection between the thymus and third branchial pouch from which it originates. The originator of this theory presumed that the hyoid bone constituted the lower level of branchial derivatives. Not only is this false but a persistent thymic duct has never been described. Furthermore, no branchial cyst has even been described deep to the thyroid gland nor have there been any examples of tracts between the pharynx and thymus.

**Inclusion theories**

King (1949) stated that there was insufficient evidence to show that cysts arose from the branchial apparatus and suggested that the cyst epithelium arose from lymph node epithelium. The following facts support this theory:

1. most branchial cysts have lymphoid tissue in the wall and are found in the parotid and pharynx as well as in the lateral neck
2. the peak age incidence is later than expected for a congenital lesion
3. a branchial cyst in a neonate is almost unknown
4. most branchial cysts have no internal opening, or at best a tract with an ill-defined termination.

**Pathogenesis**

Cysts and sinuses are lined by stratified squamous epithelium but, on occasion, by non-ciliated columnar epithelium. The appearance of this latter epithelium probably represents a glandular metaplasia as a result of infection. This could account for the mucus production from sinuses but, if it were the sole cause of cyst formation the cysts would be expected to be filled with thick, viscous mucus like thyroglossal cysts. This is not the case, however,
because the cysts contain straw-coloured fluid containing cholesterol crystals. It is the type of fluid that could only be derived from blood rather than from a mucous gland secretion. If mucus production from metaplastic epithelium was postulated as a cause of the cysts then it is unlikely that the wall would also contain lymphoid tissue, as more than 80% of cysts do. The lymphoid tissue often shows evidence of germinal centres which could only happen if the cyst formed inside a node.

Squamous cysts within lymph nodes are also found in sites far removed from the branchial apparatus, such as the posterior triangle of the neck, the pharynx, the parapharyngeal space and even within the substance of the parotid gland.

Heterotopic salivary, thyroid and squamous epithelium within lymph nodes is well documented. Salivary tissue within lymph nodes can undergo neoplastic change to form a monomorphic adenoma. Thyroid tissue may undergo carcinomatous change in the lateral neck and ectopic thyroid cancer is well recognized. Similarly, therefore, the squamous epithelium can undergo cystic change (as in branchial cysts) or neoplastic change as in branchiogenic carcinoma.

Branchial sinuses and fistulae are present at birth as one would expect for a developmental defect of the branchial apparatus. The peak age incidence for branchial cysts is in the third decade, the range being 1-70.

**Clinical features**

Sixty per cent are in males and 40% in females. The peak age incidence is in the third decade for cysts; sinuses are noted at birth and no cases are reported of sinuses appearing later in life unless by virtue of spontaneous or surgical drainage, always after infection. Sixty per cent are on the left and 40% on the right. A few are bilateral. Three-quarters are in the classical upper lateral neck position, the remainder being in the lower neck, the parotid, the pharynx and the posterior triangle.

The presenting features are:

- continuous swelling 80%
- intermittent swelling 20%
- pain 30%
- infection 15%
- pressure symptoms 5%

Seventy per cent are cystic on palpation and 30% are firm, but this is probably just a measure of fluid content. Before the widespread use of fine needle aspiration biopsy, some patients had the long work-up for a metastatic node from no identifiable primary. Indeed, the author has removed a branchial cyst after such a work-up in man of 70 who presented with a neck mass and a 6-week history.

It is unknown why some cysts present suddenly as infected masses with overlying skin erythema. The infection might be blood borne, might reflect an internal opening and infection
from the pharynx or it might be a chemical reaction within a squamous-lined cyst in a lymph node.

The differential diagnosis depends on the age of the patient.

In the patient under the age of 10 years, and especially the very young, the neck is relatively smaller than in the adult and division of the lateral neck into thirds is easier on paper than on the patient. In the newborn, a lymphangioma or dermoid must be suspected; a lymphangioma is much softer than a branchial cyst and a dermoid is very firm and tense. A lymphangioma does not have well-demarcated edges while a dermoid and a branchial cyst do. If the child is a little older, rhabdomyosarcoma is a possibility and, if it is tender, it might be lymphadenitis from the tonsil or even the teeth and pharyngeal spaces.

In the patient between the age of 15 and 40 years, the most likely alternative diagnosis is adenitis from viral or bacterial causes, tuberculosis, lymphoma or a nerve sheath tumour.

In the patient above the age of 40 years, a metastatic node from a head and neck primary neoplasm is the prime diagnosis. Alternatives are lymphoma, tuberculosis, lipoma or nerve sheath tumour.

Diagnosis is by clinical examination and from needle aspiration biopsy if necessary. Radiology is not usually helpful but if it pulsates then carotid angiography might be considered.

There is no differential diagnosis for branchial sinuses or fistula. To differentiate between a sinus and a fistula is important because all the tract must be removed to avoid recurrence. A sinogram will give this information.

**Treatment of branchial cysts**

These should be removed if they present as a mass both for diagnosis and cosmetic reasons. If left there is also the danger of infection.

A transverse incision is made in a neck crease and the sternomastoid retracted. When the cyst is mobilized, attempts are made to find a tract. This is not usually possible and very often surgeons who feel they 'ought' to find a tract, manufacture compressed areolar tissue and fascia into one going through the external and internal carotid artery. If a true tract exists it traverses this path to the posterior pillar of the tonsil. Before the operation, careful examination of this area of the fauces under anaesthesia may reveal whether or not there is an internal sinus.

If the cyst presents as an infected mass it should be aspirated and treated with antibiotics. When the infection has settled excision is planned, but sometimes there is no mass to find when the time comes for incision. If this is the case, exploration of the neck should not be performed because it will be impossible to tell which of the many lymph node contains the cyst and which are reactive.
**Treatment of branchial sinus and fistula**

The mouth of the sinus is encompassed in an elliptical incision and the tract, which is often as thick as a medium-sized artery, is found just underneath the skin. It is dissected as high as possible and then another incision is made higher in the neck. Dissection is continued to the tonsillar area where the tract usually disappears.

If a periauricular pit is noted, the surgeon should be well versed in parotid surgery and also ear anatomy, because while the tract often goes up towards the temporal region, it may also go to the junction of the bony and cartilaginous external auditory meatus. The troublesome route, however, is when the tract goes towards the facial nerve. In this instance, the surgeon should stop following the tract into the parotid and should formally identify and dissect the facial nerve.

**Branchiogenic carcinoma**

In the 1940s, many cases of carcinoma in nodes in the neck were reported and the fashion was to ascribe the development of the squamous carcinoma to neoplastic growth of heterotopic squamous epithelium within lymph nodes: hence the name branchiogenic carcinoma.

It became obvious that many of these cases were metastatic deposits in lymph nodes from primary tumours in the head and neck. This was dawn of modern head and neck surgery and in order to discourage the haphazard treatment of metastatic neck nodes, it was decreed that before a branchial carcinoma could be claimed, four postulates had to be attained:

1. the carcinoma should be demonstrated as arising in the wall of a branchial cyst
2. the tumour should occur in a line running from a point just anterior to the tragus along the anterior border of the sternomastoid to the clavicle
3. the histology should be compatible with an origin from the tissue found in the branchial vestigia
4. no other primary should become evident in a 5-year follow-up.

It is patently obvious why virtually no branchiogenic carcinomata were reported over the next 20 years. The first postulate would be a matter of exquisite timing. The second is virtually meaningless. The third merely means squamous carcinoma, but was sufficiently vague as to be menacing, and the fourth is essential.

In spite of the above, several cases of branchiogenic carcinoma have been reported in the last 20 years that undoubtedly are real. There is no doubt that heterotopic squamous epithelium can exist within lymph nodes. It would also be within the scope of normal biological developments for this squamous epithelium to undergo malignant change just as heterotopic tissue anywhere can.
While emphasizing that branchiogenic carcinoma cannot be claimed until the possibility of an undiagnosed primary has been completely excluded, it is a real entity that is underdiagnosed. Perhaps many of the long-term survivors who originally present with a metastatic neck node with a primary tumour that never comes to light, have this tumour.

Treatment is that of a node with no discoverable primary, that is either radical neck dissection and postoperative radiotherapy or excision biopsy and postoperative radiotherapy.

**Neurogenous tumours**

*Terminology*

These tumours arise from the neural crest which differentiates into the Schwann cell and the sympathicoblast; this latter cell gives rise to paraganglionic cells from which arise carotid body tumours, glomus jugulare tumours, glomus vagale tumours and ganglionic cells from which arise benign and malignant gangliomata. The Schwann cell gives rise to the neurilemmoma (schwannoma) and the neurofibroma.

Neural crest:

- Schwann cell
- Neurofibroma
- Neurilemmoma
- Sympathicoblast - Paraganglionic cells
- Carotid body tumours
- Glomus vagale
- Glomus jugulare
- Glomus tympanicum.

The thin outer sheet of nerve is called the neurilemmoma and the inner sheath of Schwann is the neurolemma. Tumours arising from the inner nerve sheath are often called neurilemmomata which is incorrect, as is neuronoma. The preferred term is 'schwannoma' for tumours arising from nerve sheath. A schwannoma shows well-developed cylindrical bands of Schwann's cells and delicate connective tissue fibres with a tendency towards pallisading of the nuclei about a central mass of cytoplasm (Verocay bodies). This form is known as Antoni type A tissue, whereas Antoni type B tissue is a loosely arranged stroma in which the fibres and cells form no distinctive pattern. The two types may also be mixed. It has no clinical significance, no surgical significance and no prognostic significance and is only mentioned because it forms part of the pathological catechism of these tumours.

Neurofibromata are often seen in association with von Recklinghausen's disease if they are multiple, but they need not be multiple and can exist as discrete entities. It is suggested that they arise from a disseminated neuroblastoma or aberrantly migrating neural crest cells. It is said that they are not as encapsulated as schwannomata and have an 8-10% chance of becoming malignant. Histologically they are characterized by loose structure, abundant matrix, stout bundles of collagen fibres and spindle-shaped cells sometimes with waveform nuclei.

The difference between neurofibroma and schwannoma is histological, but it is not as clear cut as it might seem to the non-pathologist. A recent study by Horak et al (1983) has
shown that these histological pictures can be mixed in the one tumour. From a prognostic viewpoint, these authors also show that the important difference is in the degree of cellularity rather than the classification.

Both of these tumours can show a plexiform pattern of growth. This applies especially to the cellular ones rather than to those showing the typical histological picture. The typical growth pattern is a lump that either arises from the sheath and grows outwards leaving the trunk of the nerve intact, but more often the growth involves the nerve trunk and fibres can be splayed around the tumour with apparently normal clinical function. In plexiform growth, the abnormal nerve tissue grows into adjacent tissue planes and is rather like a neural lymphangiomia. It is difficult to remove and is liable to recur. These are known as plexiform neuromata.

Gangliomata or ganglioneuromata are very rare. They usually arise from a cervical sympathetic ganglion and are firm, smooth and well encapsulated. Microscopically they contain ganglion cells and neurites.

Postoperative neuromata are the result of uncontrolled growth of axons from the proximal stump of a nerve that has been cut. These are not true tumours, but represent attempts by the damaged nerve to repair itself; the axon cylinders become enmeshed in Schwann's cells and scar tissue. If the process becomes hyperactive, the neuroma becomes clinically obvious and the patient experiences localized pain and tenderness.

Carotid body tumours were first termed 'chemodectomata'. This term has now lost favour because carotid body tumours appear to arise from paraganglionic cells rather than chemoreceptor cells. Paraganglionic cells are epithelioid in appearance, are derived from the neural crest and migrate in close association with autonomic ganglionic cells. They are located chiefly along the aorta and great vessels with the largest accumulation in the adrenal medulla where they are chromaffin positive producing catecholamines, adrenaline and noradrenaline and may give rise to phaeochromocytomata.

Formerly, tumours of the extra-adrenal chemoreceptor system were described as non-chromaffin paragangliomata, but in recent years catecholamines and secretory granules like the adrenal medulla have been found in the carotid body. Functioning tumours producing hypertension have been reported in the carotid body and the jugular body. Phaeochromocytoma have been described in association with carotid body tumours.

Paragangliomata have been reported in the following sites - aortic bodies, superior vagal ganglion (glomus jugulare), auricular branch of the vagus (glomus tympanicum), inferior vagal nodose ganglion (glomus vagale), superior laryngeal nerve (glomus laryngicum), mandible (alveolar body), ciliary ganglion (ciliary body), bifurcation of the pulmonary artery, pleura, femoral artery, retroperitoneal tissue, mesentery, coccyx and pineal body.

**Pathogenesis**

The growth pattern of nerve sheath tumours is either fusiform or plexiform as outlined in the previous section. Nerve paralysis on presentation is very rare, even though the nerve is found to be grossly distorted at surgery with the fibres widely stretched over the tumour.
The histology has also been described and the fact highlighted, that features of neurofibroma and schwannoma can exist in the one tumour. These tumours both arise from the Schwann cell. On occasion, peripheral nerve tumours may be difficult to distinguish from other spindle cell mesenchymal lesions. The neural crest marker antigen, S100, is common to the supporting cells of the peripheral and central nervous system. Immunocytochemical staining using antibody to S100 is positive in the majority of tumours of Schwann cell derivation, although expression of the antigen is reduced in Antoni A or malignant areas. The use of antibody to neuron specific enolase may help to distinguish neurofibromata although the method is less tissue specific.

They arise from any cranial or spinal nerve that has a sheath and this means any motor or sensory nerve other than the optic or olfactory. In the head and neck (apart from the acoustic nerve), the vagus is affected more commonly than any other nerve, but nerve sheath tumours have been described on the hypoglossal nerve, the facial nerve, the spinal accessory, the sympathetic chain, the glossopharyngeal and branches of the cervical plexus.

While multiple neurofibromata can occur in association with von Recklinghausen's disease, little is known of the aetiology of other nerve tumours. In a long-term follow-up of over 2000 patients who had been irradiated for tonsil and adenoid enlargement, Shore-Freeman et al (1983) found 29 schwannomata, two neurofibromata and one ganglion neuroma. These tumours can, therefore, be radiation induced and they can continue to occur for at least 30 years after the radiation exposure. They are more common in women than in men. Most cases present in the 30-50 year age group. Malignant change in nerve sheath tumours in the head and neck is very rare. Das Gupta et al (1969) described only one sarcomatous change in a series of 303 solitary nerve tumours. It is said that malignant change occurs in 10-15% of patients with multiple neurofibromatosis. The only thing that differentiates a neurogenous sarcoma from a fibrosarcoma is its origin from a nerve trunk. Malignant change is suggested by rapid growth, pain and paraesthesia. They do not metastasize to regional lymph nodes but do to the lungs.

The microscopic appearance of carotid body tumours does not relate to the future behaviour of the tumour. They are poorly encapsulated and extremely vascular, like a haemangiomia, with an enveloping network of capillaries and areolar tissue arranged in concentric circles similar to an onion. They encroach upon and gradually surround the carotid bulb, invade it and extend along the carotid artery for long distances drawing blood from the vasa vasorum. The carotid system becomes progressively distorted and extended with the internal carotid artery being attenuated but never occluded. Adjacent cranial nerves are encased as may be muscles and even the base of the skull.

Glomus tumours of the vagus grow in the same manner and totally distort anatomy at the skull base with bleeding tissue making dissection extremely difficult.

Less than 10% are frankly malignant with regional and distant metastases to lung and bone.

Due to the fact that chronic hypoxia at high altitudes leads to carotid body hyperplasia, there is a high incidence of carotid body tumour in Peru where the majority of population live.
at altitudes of 2000-5000 metres. The average age of presentation ranges from 35 to 50 years, the youngest reported being 12 years old. The sex incidence is equal.

There is a striking family history of up to 10% and also a tendency to bilateral tumours, tumours of other similar cells and phaeochromocytoma. Twenty-five per cent are bilateral in those with a positive family history, compared with 3% in those with no family history.

**Clinical features**

Nerve sheath tumours of the vagus present as parapharyngeal space masses. They are nearly all fusiform and so do not expand out of the parapharyngeal space to cause a discrete lump at the angle of the jaw, but usually push the sternomastoid laterally. This gives an ill-defined neck mass deep to the muscle which may, however, have an anterior border but certainly no palpable superior border. Since the vagus lies deep in the parapharyngeal space it pushes the pharynx medially. Tumours of the deep lobe of the parotid push the tonsil and soft palate medially but vagal and carotid tumours push the posterior pillar and posterolateral oropharyngeal wall forward. They are painless masses that rarely cause nerve paralysis.

Tumours of the sympathetic trunk present in the same way, but tumours of the hypoglossal, accessory and cutaneous cervical nerves merely present as neck lumps in the appropriate area. Facial nerve tumours are invariably diagnosed as parotid tumours.

A postoperative neuroma is very tender and, on palpation, the patient experiences an electric shock type of sensation. There must be a previous history of neck surgery, such as a parotidectomy, or a radical neck dissection. The nerves most usually affected are the cutaneous branches of the cervical plexus, that is the great auricular, the lesser occipital and the anterior cutaneous nerve of the neck. They rarely form on a stump of the hypoglossal nerve after a XII-VII anastomosis.

Carotid body tumours present in the same space, but it is as usual for them to form a discrete neck mass as it is for them to present as a parapharyngeal mass pushing the pharyngeal wall medially. Noradrenaline-secreting tumours have been described which had properties similar to phaeochromocytomas. There is also an association with the neural crest lesions and the syndrome of multiple endocrine adenomatosis should be suspected. Cranial nerve paralysis is more common in carotid body tumours than in nerve sheath tumours.

Glomus vagale tumours may or may not present with vagal nerve paralysis but all present with a neck mass high under the sternomastoid.

On clinical examination, it is impossible to differentiate these tumours if they present, as they usually do, in the parapharyngeal space. Old clinical aids, such as moving masses side to side but not up and down, are fairly useless because any lesion attached to a structure that run vertically in the neck like a vagus nerve, a carotid artery or the sternomastoid muscle will behave in such a fashion. Pharyngeal presentation will be the same for carotid or vagus masses, namely behind the posterior tonsillar pillar. Carotid body tumours are not pulsatile.
A computerized tomography (CT) scan will show if there is a mass in the parapharyngeal space and this will be very useful for smaller tumours that only give a suspicion of a lump clinically. A CT scan will not, however, always differentiate between a nerve sheath tumour and a carotid body tumour. Angiography is nearly always advisable if there is real suspicion of a carotid body tumour. This is not only diagnostic but gives an indication of the resectability of the tumour.

Fine needle aspiration biopsy is always worth carrying out but open biopsy should be avoided. Although much less frequent now, there is ample scope for surgical disaster by exploring these tumours unprepared for major vascular surgery. The disaster potential can be maximized by an approach through the mouth.

**Treatment of nerve sheath tumours**

There are three approaches to the parapharyngeal space:

1. transparotid
2. transmandibular
3. transcervical.

The transcervical route is best employed for these tumours. The sternomastoid muscle is retracted backwards, the tail of the submandibular gland swung forwards and the tail of the parotid gland lifted upwards. Adequate access to the skull base can be achieved by this route. The author's experience is that it has not been possible to remove nerve sheath tumours from this space, keeping the nerve intact. The first step is to make sure it is not a carotid body tumour or a glomus vagale. Once this is done, the nerve is dissected off the artery and excised with the tumour. The vocal cord paralysis is subsequently rehabilitated by a Teflon injection or a laryngoplasty.

**Treatment of carotid body tumours**

Carotid body tumours should be removed if technically possible. If the tumour is not removed, then the patient is left with a progressively enlarging neck tumour of uncertain biological behaviour. The factor that limits resection is extension to the skull base. Unless there is an inch (2.5 cm) or so of carotid artery free of tumour at the skull base, it is impossible to attach a graft and resection should be abandoned. Usually it is possible to dissect these tumours from the carotid after freeing its whole length and rotating it. This is to allow access to the posterior aspect where a plane can often be entered. At all of these operations, however, there should exist the facility for immediate vascular grafting if required.

Radiation should not be used as a primary treatment. It should be reserved for poor risk patients, inoperable patients, malignant tumours and those who refuse surgery.
Lymphangiomata

Terminology

There are three types of lymphangioma:

(1) lymphangioma simplex
(2) cavernous lymphangioma
(3) cystic hygroma.

Embryology

The lymph system arises from five primitive sacs (two jugular sacs, two posterior sciatic sacs and a single retroperitoneal sac) developed from the venous system. Endothelial buds from these extend centrifugally to form the peripheral lymphatic system.

There are two theories of the origin of lymphangiomata: either they are sequestrations of lymphatic tissue derived from portions of the primitive sacs, which retain their rapid and proliferative growth potential and have no connection to the normal lymph system, or they arise from endothelial fibrillar membranes which sprout from the walls of the cyst, penetrate surrounding tissue, canalize and produce more cysts.

Pathology

Lymphogenous conditions have been classified into three groups:

(1) lymphangioma simplex - composed of thin-walled capillary-sized lymphatic channels

(2) cavernous lymphangioma - composed of dilated lymphatic spaces often with fibrous adventitia

(3) cystic hygroma - composed of cysts varying in size from a few millimetres to several centimetres in diameter.

All these can be regarded as one entity, but site may play some part in the final version - the smaller lymphangiomata occur in the lips, tongue, cheek and where the tissue planes are tighter, whereas the cystic hygroma has more space to expand into the tissue planes of the neck. Simple lymphangioma can occur anywhere in the mouth as pale soft fluctuant lesions and form one-third of all lymphangiomatosus tumours. More common are cavernous lymphangiomata which form 40% of these lesions, mainly in the tongue. At the base of the tongue they must be differentiated from a lingual thyroid, a lingual carcinoma or an internal laryngocoele. They also occur on the lateral border. Some cheek lesions reach an enormous size and are very difficult to eradicate since total excision produces an unacceptable cosmetic defect.

A cavernous lymphangioma of the floor of the mouth can be part of a cystic hygroma or a ranula. Macrocheilia usually affects only the upper lip.
Cystic hygroma consists of large multinodular cystic masses which may communicate or be isolated. The walls are thin and the contained fluid can be clearly seen. The walls are thin and the contained fluid can be clearly seen. A hygroma occurs in the cervicofacial region spreading into the cheek, mouth, tongue, parotid and even the ear canal.

Histologically the cyst is lined by a single layer of flattened endothelium with fetal fat and cholesterol crystals. They are rare tumours forming 0.5% of large series of neck lumps. There is no sex or side predominance. Two out of three are noted at birth and nine out of 10 before the end of the second year.

Thirty-five per cent of lymphangiomata of all types occur in the cheek, tongue and floor of the mouth, 25% in the neck and 15% in the axilla.

**Clinical features**

Most of these tumours manifest themselves at birth or shortly afterwards. Lymphangiomata in the mouth can first appear in adult life, as can recurrences of cystic hygromata after surgery in infancy. Recurrences usually occur on the periphery of the facial area where the main mass originally presented, such as the ear, parotid or posterior triangle.

While size alone is the prominent first symptom and sign, if the cyst is big enough it can cause stridor. In very large cysts, a lateral displacement of the trachea and even mediastinal widening may be seen on the radiograph.

Sudden increase in size by spontaneous haemorrhage may be fatal. Brachial plexus compression with pain and hyperaesthesia may also occur.

The most common site is in the posterior triangle of the neck. Large masses can extend into the anterior triangle and across the midline. These anterior tumours may involve the floor of the mouth and the base of the tongue. Cystic hygromata often extend up into the cheek and parotid gland or down into the mediastinum or axilla.

If it is not the gross congenital swelling characteristically seen at birth, it can be discovered as a painless soft or semifirm swelling in the neck. The tumour usually progressively enlarges, although some fluctuation in size is common. Depending on its mass and direction of growth, the lesion can encroach on the trachea, pharynx and oesophagus causing dyspnoea and dysphagia. A sudden increase in size of the tumour may be secondary to infection or haemorrhage and has caused death. Tumour swelling is usually related to upper respiratory tract infection and tumour pain, which is an unusual complaint, occurs only in the presence of infection. One case of facial nerve paralysis due to enlargement of the hygroma has been recorded.

The diagnosis is usually made on clinical grounds. Prior to the CT era, the radiologist could contribute little. All he could identify would be displacement of the trachea or oesophagus or mediastinal extension but the size could not be imaged. Injection of radiopaque material into the cyst is unrewarding and angiography only showed an avascular mass lesion. Computerized tomography can, however, show the extent and size of the lesion and also it relation to the important structures at the skull base.
Treatment

The treatment of lymphangiomata is surgical. Intraoral lymphangiomata should be removed from an external approach since they are almost certainly much more extensive than expected. Remnants will also be left in the tissue planes of the tongue. Lymphangioma of the base of the tongue can often be dealt with by coagulation diathermy, repeated if necessary. Cryosurgery may also be helpful. Lymphangioma of the upper lip should be dealt with by a lip shave and vermillion advancement and excision of muscle and cyst to reach an acceptable size.

Recurrences usually appear within the first 9 months in about 10-15% of patients. The recurrence rate is higher with cavernous lymphangioma than with cystic hygroma.

No patient with cystic hygroma should have surgery unless he has had a chest X-ray to check for mediastinal extension. Since the child at birth looks like a monster to the mother, surgery should not be delayed. Total removal is impossible, but surgery is easiest at the first attempt, before there is infection and scarring, so as much as possible is removed. It is essential not to damage the child and so the carotid arteries, jugular vein, vagus nerve and facial nerve, if necessary, are dissected with great care. The first excision should be limited to the cervical area. After removal it will become apparent that there is a large amount of the tumour left in the parotid and cheek. Excision of this should be delayed as long as possible, taking into account the effect of an asymmetrical face on a child's psyche. The longer one waits, the bigger the facial nerve branches become and the safety of dissection increases. In the initial excision the nerves most likely to be damaged are the lower branch of the facial nerve, the spinal accessory and the vagus.

Radiotherapy should not be used because of the possible damaging effects on the growth of local structures and the potential induction of malignancy.

Injection of sclerosants has been suggested and tried, but the scarring is unpredictable because of the multiplicity of cysts and further surgery may be made very difficult.

Repeated aspirations should only be performed in the event of rapid increase in size causing pressure effects. The danger is infection and possibly haemorrhage into the cysts.

Broomhead (1964) claimed that 15% of cases undergo spontaneous regression. This view has not met with universal agreement.

After surgery, about one-third of patients will have nerve paralysis and more than half will have to undergo further surgery.
Dermoid cysts

Pathology

In the head and neck there are three varieties of dermoid cyst.

Epidermoid cyst

The epidermoid cyst has no adnexal structures, is lined by squamous epithelium and may contain cheesy keratinous material. This is the most common variety.

True dermoid cyst

The true dermoid cyst is lined by squamous epithelium and contains skin appendages such as hair, hair follicles, sebaceous glands and sweat glands. These are either congenital or acquired.

The congenital type derives from ectodermal differentiations of multipotential cells pinched off at the time of closure of the anterior neuropore. It occurs, therefore, along the lines of fusion.

The acquired type is due to implantation of epidermis at the time of a puncture type of injury and is often solid with areas of cystic spaces containing sebaceous material.

Teratoid cyst

The teratoid cyst can be lined with squamous or respiratory epithelium and contains elements formed from ectoderm, endoderm and mesoderm - nails, teeth, brain, glands, etc. This is the rarest variety in the neck and is nearly always diagnosed in the first year of life. Less than 10 cases of teratoid tumours of the neck have been described in adults.

Twenty per cent of all dermoid cysts are found in the neck, and 30% in the neck and face. Dermoids form 28% of all midline cysts, and there is no sex predominance.

Clinical features

These cysts present as solid or cystic masses in the midline of the neck between the suprasternal notch and the submental region. They can also occur lateral to the submandibular gland. Painless swelling is the only symptom, but if the cyst is large, minor obstructive symptoms can occur.

About 20% of dermoids occur in the mouth, either deep to the mylohyoid (sublingual) or superficial to it (submental). They present in the second and third decades but have probably been present since birth.
Treatment

Complete excision is usually easy and should be carried out in all cases.

Infective neck masses

Tuberculosis

Pathology

The condition is not common in USA or Europe, but it is still common in Asia or Africa. There are 32,000 new cases of tuberculosis in the USA each year and 5% of these (1,600) develop cervical lymphadenitis.

Where the incidence of tuberculosis is low, primary infections are acquired later and so it is young adults who acquire tuberculosis nodes. In the UK, the maximum age incidence is 5-9 years, but in 30% it occurs in patients over the age of 25 years.

The bacillus, which is usually the human variety, reaches the lymph nodes by direct drainage or by haematogenous spread. The incidence of coexisting pulmonary tuberculosis is less than 5%. In one series, almost half of the tonsils removed showed evidence of tuberculosis and it thus appeared that the tonsil was the source and that the cervical adenitis was precipitated by an attack of acute tonsillitis. Once the bacillus has entered the host, further exposure is not necessary to trigger off the adenopathy.

Clinical features

Most patients give a fairly long history and usually seek medical advice because the lumps have become painful. In Asia, the presentation is different: 20% have discharging sinuses, 10% a cold abscess and 10% are adherent to the skin; in these patients all have a negative chest X-ray.

Ninety per cent are unilateral and 90% involve only one node group, the most common being the deep jugular chain followed by the nodes in the submandibular region and then those of the posterior triangle.

Diagnosis is by a positive tuberculin skin test and demonstration of acid and alkali fast bacteria in the biopsy. In the USA, patients should also have histoplasmin and coccidioidin skin tests. The differential diagnosis is between lymphoma and metastatic cancer. The absence of a primary tumour in a young adult and the length of history usually makes the latter diagnosis improbable.

Treatment

The treatment is an excisional biopsy followed by 9-12 months of antitubercular chemotherapy. If the glands are very large and matted, local removal is dangerous since the nodes are often attached to the internal jugular vein. A functional neck dissection should then be carried out preserving the sternomastoid, accessory nerve, and jugular vein if possible. In
a child, it is usually wise to remove and histologically examine the tonsils before removing the lymph nodes.

If removal is not followed immediately by chemotherapy, a sinus forms with persistent drainage and, later, ugly scars.

**Sarcoidosis**

Sarcoidosis presenting as cervical adenitis with no other manifestation of the disease is extremely rare. The neck nodes are not often involved in this condition, even when it is generalized. It is almost impossible, therefore, to make a preoperative diagnosis and a biopsy is always needed. The histological characteristic is the absence of caseation. Diagnosis may be confirmed by the Kveim skin test.

**Neck space infections**

Neck space infections are very rare and there is confusion about how many neck spaces there are: estimates vary from 13 to 20. A fascial space is an area of loose connective tissue bounded by dense connective tissue called fascia. It is a matter of opinion how thick connective tissue must be before it is called fascia, and this is where the disagreement as to the number of spaces arises. Knowledge of the anatomy of the areas in which infection tended to collect was important in the pre-antibiotic days from the point of view of routes of spread, complications, and surgical drainage, but nowadays knowledge of three spaces (retropharyngeal, lateral pharyngeal and submandibular) will allow management of 90% of patients.

**Anatomy**

The spaces listed are described by Hollinshead and are shown in Table 15.1.

Nowadays, abscesses usually only occur in the retropharyngeal, lateral pharyngeal (parapharyngeal) and submandibular spaces.

**Retropharyngeal space**

This space lies between the pharynx and the posterior layer of the deep fascia which bounds the prevertebral space. It separates the pharynx from the vertebral column and extends from the base of the skull to the posterior mediastinum as far as the bifurcation of the trachea. Anteriorly, it connects with the pretracheal space so that infections can spread by way of this latter space to the anterior mediastinum. But mediastinitis due to a retropharyngeal abscess is rare. In the infant this space contains one or two lymph nodes.

**Lateral pharyngeal space**

This is more commonly known as the parapharyngeal space; it lies lateral to the pharynx connecting with the retropharyngeal space posteriorly. Laterally, it is bounded by the lateral pterygoid muscles and the sheath of the parotid gland. It extends from the base of the skull to the level of the hyoid bone where it is limited by the sheath of the submandibular
gland. This sheath is also connected to the sheaths of the stylohyoid muscle and the posterior belly of the digastric muscle.

The carotid sheath is bounded anterosuperiorly by the pterygomandibular raphe and the spaces around the floor of the mouth anteroinferiorly.

This space is prone to infection because of its close connection to the tongue, teeth, parotid, submandibular gland and tonsils.

Submandibular space

This is bounded above by the mucous membrane of the floor of the mouth and tongue and below by the deep fascia that extends from the hyoid to the mandible.

It is divided into two by the mylohyoid muscle and so the submandibular gland, which is wrapped around the mylohyoid muscle, extends into both parts of the space. The space superior to the mylohyoid muscle contains most of the sublingual gland. The space inferior to the muscle contains the submandibular gland. Anteriorly lies the submental space between the two anterior bellies of digastric.

Infections of this space are known as Ludwig’s angina.

Table 15.1 Anatomy of neck spaces

Below the hyoid
- carotid sheath
- pretracheal space
- retrovisceral space
- visceral space
- prevertebral space

Above the hyoid
- mandibular space
- submaxillary space
- masticator space
- parotid space

Peripharyngeal area
- retropharyngeal space
- lateral pharyngeal (parapharyngeal) space
- submandibular space

Intrapharyngeal area
- paratonsillar space.
**Clinical features and management**

**Retropharyngeal abscess**

This abscess in infants is due to a lymphadenitis secondary to an upper respiratory tract infection. The child has a sore throat; examination shows a swelling behind an otherwise normal tonsil. The temperature is elevated to 38-39°C (101-102°F) and the child is ill. The swelling may obstruct the posterior nares and push the soft palate down. Respiratory obstruction is an ever-present danger because the child’s spine is short and his larynx is high. (In a 9-month-old infant, the epiglottis is at the level of the atlas.)

Radiographs of the neck show a large retropharyngeal swelling. Treatment is by incision and drainage in the tonsil position.

In an adult, a retropharyngeal abscess usually signifies a tuberculous infection of the cervical spine. It is of insidious onset with a low grade fever. Pus must be obtained to confirm the diagnosis which is also suggested on a radiograph of the cervical spine. Treatment is by antituberculous chemotherapy.

**Parapharyngeal abscess**

This is more common in adults than children. It is a complication of tonsillectomy or tonsillitis in about 60% of patients and a complication of infection or extraction of the lower third molar in a further 30%.

Infection of the petrous apex can rarely rupture directly into the space. Infection of the mastoid tip can also enter the space by way of the digastric sheath.

There is fever and marked trismus because of involvement of the medial pterygoid muscle. The tonsil is pushed medially but looks normal. The most marked swelling is in the neck at the posterior part of the middle third of the sternomastoid. Each patient should be given at least 48 hours’ treatment with an antibiotic, but by this time most patients have a swollen neck and incision and drainage will be required.

**Ludwig’s angina**

This is a rapidly swelling cellulitis of the floor of the mouth and submandibular space secondary to soft tissue infection, tonsillar infection and infection of the lower premolar and molar teeth. Over 80% of patients have dental disease and, in these patients, the lower molars are set eccentrically with the roots closer to the inner than the outer side of the jaw, or the roots of the second and third molars may lie inferior to the mylohyoid line. Root abscesses of these teeth, therefore, drain into the submandibular space. This space may be affected with minimal discomfort from the tooth; pain comes from tension within the bone, but if this gives way and drains there is no dental pain. In cases of dental origin the most usual organisms are *Streptococcus viridans* and *Escherichia coli*.

When infection spreads to the sublingual space, the floor of the mouth becomes very swollen and appears as a roll of oedematous tissue rising to the level of the biting edge of the
teeth. The tongue is elevated posterosuperiorly and respiratory obstruction is a danger. The patient is very ill with a temperature of over 38.3°C (101°F) with pain, trismus and salivation.

Treatment is by antibiotics; incision and drainage should be postponed as long as possible.

Laryngocoele

Pathology

In the UK, the incidence of laryngocoele is approximately one per 2.5 million population per year. The sex incidence is 5:1 in favour of men and the peak age incidence is at 50-60 years. Only one case has been reported in a neonate, but it is possible that this was a laryngeal cyst; 825 were in Caucasians; 85% were unilateral and 15% bilateral. They can be external (30%), where the sac arises from the laryngeal ventricle and expands into the neck through the thyrohyoid membrane, internal (20%) where it arises from the laryngeal ventricle and stays within the larynx presenting in the vallecula, or combined (50%). Laryngocoeles are lined by columnar ciliated epithelium, whereas simple laryngeal cysts are lined by squamous epithelium.

It has long been held that laryngocoeles are due to 'blowing' hobbies or jobs such as trumpet playing or glass blowing. A careful review of the published cases reveals at most four patients subject to these habits so that this theory appears to be untrue. Of more importance is the coexistence of a carcinoma or papilloma of the larynx which acts as a valve allowing air under pressure into the ventricle. External laryngocoeles can be found in 16% of laryngectomy specimens in laryngeal carcinoma, as opposed to 2% in laryngectomy specimens for pyriform sinus cancer. The ventricle in these cases of laryngeal cancer was also significantly higher than that in patients with pharyngeal cancer due to increased air pressure consequent upon obstruction by the laryngeal carcinoma.

Lower animals have air sacs, for example the cheek pouch of monkeys, the fish pouch of pelicans, the tracheal sacs of emus, the syrinx of male quacking birds, etc. Lateral laryngeal sacs are well developed in certain anthropoid apes and are a means of enabling the animal to rebreathe while holding its breath for long periods.

Laryngocoeles in man, therefore, are almost certainly atavistic remnants corresponding to these lateral air sacs. On occasion they become manifest due to an increase in intralaryngeal air pressure due to blowing or coughing.

Clinical features

The most common presenting features are hoarseness and a swelling in the neck. The third most common symptom is stridor, which can come on very suddenly over a period of a few days or even hours in a patient who had previously only mild symptoms for months or years. Other presenting symptoms are dysphagia, sore throat, snoring, pain or cough. Ten per cent present with infected sacs (pyo cocoes) and, because of the mixture of infection and air on the radiograph, a diagnosis of gas gangrene is sometimes made. On palpation, the swelling which is usually large and over the thyrohyoid membrane, can be emptied easily. A
plain radiograph of the neck is diagnostic showing an air-filled sac. Nothing else except gas gangrene can produce this picture.

The most common presenting symptom of laryngocoeles is hoarseness with apparently normal vocal cords. To diagnose smaller laryngocoeles, therefore, every patient with hoarseness and normal vocal cords should have a plain anteroposterior neck X-ray with and without a Valsalva manoeuvre. Especially if a unilateral laryngocoele is present, the patient should have a laryngoscopy to make sure there is no underlying carcinoma of the laryngeal ventricle. These tumours can act as valves allowing air into, but not out of, the saccule thus dilating it.

If an enlarged saccule does not penetrate the thyrohyoid membrane, it travels up behind the thyrohyoid membrane and the hyoid and bulges into the vallecula. This is an internal laryngocoele and, if the mouth of the sac is blocked, then it may be full of mucus and not radiolucent. These are often diagnosed during routine otolaryngological examination and may be quite symptomless. On occasion they can reach a reasonably large size and the patient complains of a feeling of a lump in the throat.

**Treatment**

Laryngocoeles discovered radiologically and that are contained within the larynx, require no treatment. The patients have the cause of the hoarseness explained to them and are kept under annual review. If surgery is attempted on these patients, the uninflated saccule is almost impossible to find and oversew.

Internal laryngocoeles may be uncapped to see if marsupialization and scarring will stop recurrence. If they recur then they should be excised with the approach used for the external laryngocoele.

An infected laryngocoele should be aspirated and treated with antibiotics. When the infection has subsided, formal excision should be carried out.

The best operation for a laryngocoele aims at excising the saccule at its neck. This is accessed by removing the upper half of one thyroid ala, or fracturing it downwards and replacing it. The method used depends on the state of ossification of the thyroid cartilage. Once access to the supraglottis is obtained, it is an easy matter to follow the neck of the laryngocoele down as far as possible. The neck is transected and closed in layers and oversewn. If the thyroid ala is not replaced then the thyroid perichondrium is sewn into the area.

Recurrence after this operation is extremely rare.