Chapter 18: Non-neoplastic salivary gland disease

A. G. D. Maran

As with ear disease, people do not normally die of non-neoplastic salivary gland disease and thus some basic principles of pathology are still ill understood.

Pseudoparotomegaly

Hypertrophy of the masseter

This is a condition which occurs almost exclusively in females. It is very distressing because it lends a square shape to what should otherwise be an oval-shaped face. It usually affects younger females and so they are more sensitive about their facial appearance. It looks like bilateral parotid enlargement, but the diagnosis becomes obvious when the patient is asked to clench the teeth and a bulging rippling masseter becomes obvious. It is an important diagnosis to make if only to stop unnecessary parotidectomies being performed. In the group of patients in whom it occurs, it must be distinguished from true parotomegaly that results from bulimia.

The reason for the muscular hypertrophy is not truly understood. It is too simplistic to blame all cases on bruxism which is grinding of the molar teeth. Although some patients do this, it is unlikely that this can stand as a cause unless the cusps of the molars show signs of distinct wear. It is sometimes seen in patients who have undergone long-term orthodontic treatment, perhaps as the result of the masseter setting the jaw in the new position. The most recent explanation is that it is due to kissing and that this is why it is predominantly a disease of females because the male has the stronger jaw.

Treatment is difficult but often very necessary because of the distress of these young females. There is little point in approaching the problem from an external route. This causes a facial scar, requires an unnecessary parotidectomy and puts the facial nerve at risk. It is unlikely that sufficient masseter could be removed without causing at least a temporary paralysis of the facial nerve and the disease certainly does not warrant this approach.

An intraoral approach is the recommended one. The jaw is splinted open and a cut is made along the ascending process of the mandible. The three heads of the masseter are identified and the inner two heads are removed. This leaves the outer head protecting the facial nerve. Care must be taken in this operation, however, not to penetrate the posterior limit of the masseter or else the main trunk of the facial nerve will be at risk.

Ageing

With the absorption of adipose tissue in the ageing process, the salivary glands become more obvious. It is usual that one can palpate the glands in the submandibular area in elderly patients. Patients are sometimes referred to otolaryngology/head and neck units with suspected metastatic disease of the submandibular nodes on the basis of absorption of fat from the submandibular area, leaving the glands obvious and palpable.
**Dental causes**

Dental infection can spread to the lymph nodes either within or surrounding the parotid gland and also to the lymph nodes in the submandibular area. Drainage of the lower incisor or canine teeth below the mylohyoid line can, on occasion, cause Ludwig's angina which involves the submandibular glands although the swelling is much more brawny. Tissue oedema within the infratemporal fossa and between the heads of the masseter muscle in turn causes facial swelling. This is probably the basis of the old-fashioned 'gum-boil'.

**Tumours in the parapharyngeal space**

These are dealt with elsewhere in this volume. Chemodectoma, glomus vagale tumours, schwannomata of the vagus or sympathetic trunks and enlargement of lymph nodes with cyst, tuberculosis or metastatic disease can fill this space and push the parotid gland outwards. This gives fullness in the parotid area that is not a true parotid swelling, but only a displacement. These tumours can also present between the tail of the parotid and the tail of the submandibular gland at the angle of the jaw and displace both structures.

**Tumours of the infratemporal fossa**

The infratemporal fossa is anterior to the parapharyngeal space and tumours in this area can mimic parotid swellings by exiting from the space through the mandibular notch or under the zygomatic arch. The author has had personal experience of this occurring with a haemangioma, a haemangiosarcoma, a leiomyosarcoma and a hydatid cyst.

**Mandibular tumours**

Although tumours of the mandible are relatively rare, both osteosarcoma and chondrosarcoma of the ascending process of the mandible can mimic parotid enlargement as can tumours of the horizontal portion of the mandible in relation to the submandibular gland.

**Mastoiditis**

Mastoiditis in any form is now extremely rare in western and north Europe, but there are parts of the world where it still occurs. A well pneumatized mastoid, if infected, can cause a subperiosteal abscess which, in turn, can drain into the sternomastoid muscle or the digastric muscle lifting the tail of the parotid and mimicking parotid enlargement.

**Intraparotid lesions**

1. Neuroma of the facial nerve
2. Aneurysms of the temporal artery
3. Lymph node enlargement in or around the parotid gland
4. Parotid cysts.
Parotitis

Pathogenesis

Parotitis is probably the most common infectious disease in childhood and is due to the mumps virus in this age group. What is less well known is that the incidence of viral parotitis in young adults is rising and is due to infection with one of the many strains of the echo- or coxsackieviruses. Bacterial parotitis used to be a common premortem event prior to the advent of antibiotics. The cause of this was an ascending staphylococcal infection along the parotid duct in the dehydrated patient who lacked resistance to infection. As a common oral infection, fungal parotitis is not as common as one would expect given the vicinity of thrush to the duct.

Parotitis can occur secondary to obstruction of the duct either by a stone, which is common in the submandibular gland, epithelial debris, which is common in the parotid gland, and stenosis of the parotid duct due to interdental problems. The patient usually recovers from the pain within a period of minutes or hours but, on occasion, infection can supervene and may even progress to a parotid abscess formation.

Infection of the parotid gland can be due to lymph nodes. There are 6-10 lymph nodes around the parotid gland and 4-6 within the parotid gland. These drain the skin of the side of the face, the scalp, the ear, the eye and the posterior part of the oral cavity. Lymphadenitis can thus be secondary to skin lesions and, in this regard, infected pierced ears are probably the commonest cause. Otitis externa frequently presents with preauricular pain and swelling due to lymph node enlargement. The relationship with dental infection of the molar teeth has already been mentioned and preauricular pits as a result of malformation of the branchial apparatus can result in recurrent infections which drain to the parotid lymph nodes. These nodes may resolve or they may proceed to abscess formation.

Chronic infection of the salivary glands can occur as the result of tuberculous infection. It is rare for tuberculosis to affect the stroma of the gland and, if the salivary glands do become affected by tuberculosis, then it is the surrounding lymph nodes that are infected. Sarcoid can similarly affect the area and rarely the parotid and submandibular glands can be affected by actinomycosis, leprosy or tularemia.

The symptoms from parotitis prior to abscess formation come when saliva is produced. This happens when the patient attempts to eat and creates secretomotor stimulation of the salivary glands. If the ducts are oedematous or blocked with stone or debris, then the flow of saliva is obstructed and the tense glands swell even more, causing severe pain. The parotid and the submandibular glands are covered with the investing fascia of the neck. Although this is not very obvious at surgery, it causes constriction of the salivary glands in the case of infection. It then becomes very difficult for the glands to expand causing severe pain.

Clinical features

Parotitis causes severe pain and elevation of temperature. It is made worse by eating and the patients are very hungry but do not eat because of the fear of pain. The pain and surrounding swelling cause spasm of the masseter, the temporalis and the pterygoid muscles;
this causes trismus. The effect of trismus in a viral parotitis can be to create the environment in which a superadded opportunistic bacterial infection can occur due to bad oral hygiene. The area over and around the affected salivary gland is extremely tender. The diagnosis can be substantiated by asking the patient to sip a little lemon juice when there will be an acute worsening of the pain. This will not be the case, however, if the swelling is due to a lymph node.

Apart from the examination of the salivary glands, the oral cavity should be examined for the presence of dental infection and the molar teeth especially should be palpated, moved and tested with hot and cold stimuli. Some attempts should be made to see what material comes out of the salivary ducts with moderate pressure over the glands. Pierced ears and otitis externa should be looked for.

It should also be borne in mind that the painful parotid gland may be a manifestation of Sjögren's disease, but this is not at all common.

**Laboratory investigations**

There will be an elevated white cell count with lymphocytes predominating if it is a viral infection, and neutrophils if it is a bacterial infection. The erythrocyte sedimentation rate will also be raised in keeping with the general condition of the patient but, if it is very elevated, then the possibility of Sjögren's disease arises.

Viral titres should be measured in every case and, although the mumps titre is reliable, there are so many strains of the echo- and coxsackieviruses that it is felt to test for each strain would be too expensive in terms of reagents and manpower.

**Bacteriology**

Secretions from the ducts can be used to try to identify viral infection and also can be plated for bacterial culture and sensitivity. Mycology should also be examined in every case.

If tuberculosis or sarcoidosis is suspected then the opinion of an ophthalmologist should be secured to see if the patient has uveitis. This often accompanies parotitis in these two conditions and if present completes Heerfordt's syndrome.

**Radiology**

**Plain X-rays**

Plain X-rays are useful in identifying the presence of stones. Occlusal films in the submandibular region show stones very well and are much better in this regard than lateral films. It is important to be able to assess whether or not the stone is in the oral cavity or in the gland.

Parotid stones are radiolucent and will not be seen on plain X-ray.
Sialography

The timing of a sialogram in parotitis is debatable. It would be universally agreed that a sialogram should not be performed during the acute phase. A week or two later, however, it might be therapeutic in washing out the duct system.

This examination is very useful because it will give some idea of duct blockage and will certainly give a diagnosis of sialectasis, if this is present. It may show duct distortion although this is such a variable feature that it lacks any diagnostic specificity.

Scanning

Parotid scanning with technetium-99 has largely been abandoned because of so many false positive and negative results.

Fine needle aspiration biopsy

This is only possible in departments with an experienced cytologist. The diagnosis in parotitis is usually so obvious that fine needle aspiration is not required, but it may be carried out for confirmation.

Conservative treatment

The patient will feel so ill that he probably should be confined to bed. In spite of trismus and pain, a good standard of oral hygiene should be maintained and, although eating and drinking are very painful, the least painful nutrition is with high calorie milk drinks which do not carry any flavour. All patients will require the appropriate degree of analgesia and local heat applied to the affected gland is often comforting. Adrenaline should be applied to the appropriate salivary duct in the hope of reducing the oedema and causing some lessening of tension with drainage of saliva. It is doubtful if any antibiotic is truly effective, but the only antibiotic that is secreted in saliva is clindamycin and this can be used according to the circumstances. If a diagnosis of tuberculosis is established then the appropriate chemotherapy can be begun with a high resolution rate.

Similarly, with leprosy, actinomycosis and tularaemia, a reasonable response to chemotherapy can be expected.

If the infection is due to lymphadenitis, then the primary source of infection should also be dealt with either surgically or with antibiotics.

Surgical treatment

The most successful surgical intervention is where a stone blocks the submandibular duct with secondary sialadenitis. The stone can be removed perorally to eradicate infection, but since abscess formation is often loculated any serious attempt at drainage would involve lifting a facial flap and carrying out multiple incisions over the gland. This is seldom warranted.
**Metabolic parotomegaly**

The following conditions have traditionally been recorded as causes of parotomegaly - gout, Cushing's disease, myxoedema and diabetes mellitus. Investigators who have examined large series of patients with these endocrine disorders with special reference to their salivary glands, have not substantiated the original relationships and endocrine parotomegaly may be anecdotal.

The parotid gland is much more closely related to nutritional abnormalities. Since the Second World War, parotomegaly and enlargement of the submandibular glands have been noted in prisoners of war and other groups subject to starvation. This is probably a similar process to the one mentioned earlier in relation to ageing, namely the persistence of the substance of the salivary glands with the disappearance of surrounding adipose tissue.

The metabolism of fat within the parotid glands is ill understood. As well as the salivary glands maintaining their shape and form and the surrounding adipose tissue disappearing, fatty infiltration is very common in obese individuals. Furthermore, if these individuals lose a lot of weight then they are left with the original fat deposition in the salivary glands causing residual parotomegaly.

Although salivary gland enlargement is not noticed in anorexia nervosa, parotomegaly is a feature of the recently recognized condition of bulimia where binge eating is followed by self-induced vomiting. Several of these patients have presented with parotomegaly and have not disclosed their binge eating. As a result some normal parotids have been removed. Histological examination of these glands, furthermore, has failed to reveal any abnormality or abnormal deposition of fat.

**Drug-induced parotomegaly**

In the laboratory, it has been shown that both isoprenaline and thiouracil make a rat's parotid swell. Nowadays, with the expert control of thyroid disease, it is very seldom that a patient with parotomegaly due to thyroid-related medication or isoprenaline is seen. The list made up by the Committee for the Safety of Medicines, however, shows over 40 drugs as affecting the salivary glands. Many of these reports are anecdotal and coincidental and there is little in the way of scientific evidence to show that drugs do cause parotomegaly. Although drug 'allergy' is mentioned in many reviews of parotomegaly, again there is no evidence for such an entity.

In clinical practice, the only drugs associated with parotomegaly or painful parotid with any frequency are dextropropoxyphene (Distalgesic) and high oestrogen oral contraceptive pills. The method of action of these latter drugs is to create epithelial shedding within the duct system, the creation of epithelial mud and the blockage of salivary ducts with the possible creation of stones in the submandibular area.
Sialectasis

Pathogenesis

The cause of sialectasis is unknown. It is probably best regarded as a salivary gland analogue of bronchiectasis. There is a progressive rotting and disintegration of the alveoli which ultimately coalesce forming cysts. The debris from these cysts passes along the duct and intermittently blocks areas of the duct causing hypertrophy, stenosis and duct dilatation. This is exactly what happens in bronchiectasis and, as with this disease, some have it from birth, and others develop it for no very good reason. In a few this debris is secondary to known obstructions, but occurrence in both the salivary glands and the lung is the exception rather than the rule. Congenital sialectasis has no adequate embryological explanation.

The parotid is a serous gland and is low in calcium. The epithelial debris, which could become calcified to form a stone, does not have the stimulus of the correct environment for calcification and remains as ‘mud’. This is effective in blocking the duct system but not as effective as a stone would be. It is softer and does not impact so easily and is more easily removed with a build-up of saliva. Stones can form, however, in the parotid gland but they are of low density and are radiolucent.

The submandibular gland is a mixed seromucinous gland and is high in calcium. Epithelial debris here, therefore, calcifies easily and this is why stones are more common in the submandibular than in the parotid gland. The stones are of high density and are, therefore, radiopaque.

Although some texts list calculus disease as a separate entity, it is unlikely that stones can form de novo. They are probably all formed in a radiologically negative sialectatic gland.

The symptoms are produced when the ducts are blocked. If the main duct is blocked then the whole gland will swell up in response to the secretomotor stimulation of eating, or drinking. This is especially marked with citrus drinks or fruits which cause maximal salivary stimulation. The blockage may be in a more distal duct, in which case only a portion of the gland will swell up.

In most instances, the gland will clear itself. Sometimes the swelling stays for days but usually only for minutes or hours. If the swelling stays for some days, then it may become secondarily infected and abscess formation occur. The abscess may rupture or be drained but more likely it will heal by fibrosis.

Clinical features

History

The patient typically complains of pain and swelling of the gland during a meal. The swelling is visible and can remain up for minutes, hours or days. While the gland is swollen it is painful but, when the swelling goes down the gland is not painful. The patient does not feel unwell and does not have an elevation of temperature.
The condition can be particularly troublesome in children. This is the group of children that are diagnosed as having 'mumps' more than once. Fifty per cent of these cases resolve in time and only a few adult cases require surgical treatment.

Examination

On examination, a stone may be seen in the submandibular duct or palpated by bimanual palpation within the gland. The mouth of the duct may be oedematous and pouting.

The parotid duct may have the same appearance and it is useful to massage each parotid to see if there is any drainage of saliva from the duct.

Investigations

Laboratory investigations

There is little information to be gained from blood tests.

Radiology

The sialogram is diagnostic of this condition. A plain X-ray should be performed in all cases to see if a radiopaque stone is visible and then one should proceed to a sialogram.

A sialogram may show six pictures:

(1) it may be normal

(2) it may be overfilled, an overfilled sialogram is often reported as sialectasis and one must be aware of this picture

(3) the radiologist may fail to cannulate the duct; most radiologists know that they should not persist with difficult cannulations for any period of time because the duct becomes oedematous and it then becomes virtually impossible to cannulate; if cannulation in these cases is eventually successful then the radiologist's report will be of duct stenosis but it will be iatrogenic rather than real

(4) the fourth possible picture is that of an obstructed duct; this will be obvious in the submandibular gland if a stone is seen on the plain film but in the parotid gland, the dye may enter a little way along the duct and come to a halt, in which case it can be presumed that epithelial mud is blocking the main duct

(5, 6) there are two classical pictures of sialectasis: the first is cystic and the second is globular or saccular. Only one of these truly represents the pathology of the condition. Cystic sialectasis where the alveoli coalesce and form large spaces together with duct, stenosis and dilatation, is the true picture of sialectasis. Thackray (1955) has shown that globular sialectasis represents no abnormality of the duct other than abnormal leakage where the lipiodol or radiopaque medium comes out of the alveoli and lies in the stroma of the gland. Although popularly called saccular sialectasis, it is not related to the pathology that was described akin to bronchiectasis.

As stated previously scans are unreliable and a sialogram together with a computerized tomography (CT) scan is of little value in this condition.
Treatment

No treatment

In many cases the sialogram is therapeutic. It washes out the duct and alveolar system and, thereafter, the patient may be advised to finish each meal with a citrus drink which will encourage the production of saliva, and then to massage the affected gland in order to wash out any epithelial debris and stop collections. This is successful in the vast majority of cases.

Peroral removal of a calculus

This can be carried out if a calculus is seen, usually in the submandibular gland. The duct is blocked proximally to stop the stone disappearing back into the gland during manipulation and the duct is marsupialized after the stone is removed.

Marsupialization of the duct

This must accompany any peroral removal of the stone and it can be undertaken in the parotid duct stenosis of dental origin. A cannula is placed into the duct and a 2.5-5 cm (1-2 inch) segment is opened and stitched carefully to the adjacent mucosa with 6-0 absorbable suture material.

Ligation of the duct

This is mentioned only to be dismissed as an illogical and inadequate present-day treatment.

Duct dilatation

The same applies to this method of treatment.

Tympanic neurectomy

This procedure was popular during the 1950s and 1960s. The aim of treatment is to divide Jacobson's nerve which crosses the promontory to form the tympanic plexus. It joins the jugular plexus and glossoharyngeal nerve to the greater superficial petrosal nerve and forms part of the reflex arc. Some surgeons combine this with division of the chorda tympani nerve.

The procedure certainly works for up to 6 months but, like all autonomic surgery, alternative pathways develop and symptoms recur.

Removal of the submandibular gland

This is a straightforward procedure and carries little risk with it. If the gland shows evidence of sialectasis, or if there is a stone in the body of the gland, then the surgeon should have no hesitation in removing the submandibular gland as a whole. If it has undergone numerous attacks of sialadenitis, then the removal may be difficult, but care should be taken
to avoid paralysing the mandibular branch of the facial nerve. Furthermore, one should make sure that the remnant of the submandibular duct in the oral cavity is clear of stones when the gland is removed, because it is quite possible to push stones from the gland into the duct remnant during manipulation. This is of little clinical significance, but the patient may be surprised to spit out a stone some days after the operation.

**Total parotidectomy**

Superficial parotidectomy is illogical for a disease which affects the whole parotid gland. By removing half of a sialectatic parotid gland, there is a high risk of fistula. In a superficial parotidectomy for tumour, a denervated normal deep lobe is left behind. This usually ceases to function and thus fistula is rare. In sialectasis a cystic diseased deep lobe is left behind for which denervation does little. It continues to produce mucus and saliva and causes a salivary fistula.

Total parotidectomy is, therefore, the only logical operation for sialectasis and, in the gland which may be heavily fibrosed due to recurrent sialadenitis, the facial nerve is at more risk than it is in surgery for benign tumours.

**Sjögren's syndrome**

In 1888, Dr Mikulicz described the case of a 42-year-old East Prussian farmer with swelling of the submandibular, parotid and lacrimal glands. He removed two-thirds of the submandibular gland and found it to be infiltrated with lymphocytes, and then removed the whole gland some months later with recovery of the patient. This was known as Mikulicz's disease and to it were added all the symptoms of other non-neoplastic salivary gland disease over the next 50 years. Mikulicz's syndrome included tuberculosis, sarcoid, actinomycosis, gout, etc. In 1925, Gougerot, a French dermatologist, introduced the concept of dryness when he described a series of patients with dry mouth, vulval dryness, skin dryness, etc. In 1933, Henrik Sjögren, a Stockholm ophthalmologist, described 33 women with the syndrome of xerostomia and keratoconjunctivitis sicca. Twenty-three of these patients had rheumatoid arthritis. No mention was made of parotid gland disease. In 1952, Godwin at the Armed Forces Institute of Pathology in Washington, described the concept of enlargement of the parotid glands due to lymphocytic infiltration and related this to the future development of lymphoma. It was named benign lymphoepithelial lesion. In 1974, Anderson and Talal described a further variety of this called aggressive lymphocytic behaviour.

The classification now is as follows:

1. primary Sjögren's syndrome (sicca complex) - this consists only of xerostomia and xerophthalmia with no connective tissue component

2. secondary Sjögren's syndrome - this consists of xerostomia, xerophthalmia and a connective tissue disease which in nearly 50% of cases is rheumatoid arthritis but may also be systemic lupus erythematosus, scleroderma and polymyositis

3. benign lymphoepithelial lesion, otherwise known as myoepithelial sialoadenitis, which is localized to the parotid glands and some regard as a prelymphomatous condition
aggressive lymphocytic behaviour which again is confined to the parotid glands and is almost a pseudolymphoma.

**Epidemiology**

Sjögren's syndrome is more common in the northern than the southern hemisphere and is more common in northern than in southern Europe. A proportion of older people have many of the symptoms of Sjögren's disease, but do not have the immunological profile. In a study of octogenarians, Whaley showed that one in six males and females had keratoconjunctivitis sicca. Three per cent of men and 20% of women had xerostomia, but only 2% had the immunological profile of the sicca syndrome. Eleven per cent of patients with rheumatoid arthritis had keratoconjunctivitis sicca, 1% had xerostomia but 100% had lymphocytic infiltration of the submaxillary glands when examined at autopsy. Thirty per cent of patients with rheumatoid arthritis will develop Sjögren's disease. It has a very wide range of autoantibodies and is the second most common autoimmune disease after rheumatoid arthritis.

**Clinical features**

Sjögren's syndrome is a multisystem disease affecting every system in the body but particularly the oral cavity, the eyes and the salivary apparatus.

The oral symptoms are those of dry mouth with secondary candidiasis, stomatitis, glossitis and subsequent dental caries.

The eye symptoms are keratoconjunctivitis sicca; the patient has a foreign body sensation in the eye, burning, redness, itching, photosensitivity and an inability to tolerate contact lenses.

Only 40% feel salivary gland enlargement and only 20% show it clinically. It is nearly always in the parotid and those patients with parotomegaly from Sjögren's disease have a much higher chance of developing lymphoma. Two-thirds of the patients never have salivary gland enlargement.

Other associated systemic problems are primary biliary cirrhosis, chronic hepatitis, vasculitis, chronic graft versus host disease, cryoglobulinaemia, hypergammaglobulinaemic purpura and polyarteritis. Fifteen per cent will have thyroiditis and many will develop pancreatitis.

Achlorhydria, disorders of oesophageal motility and web formation may present to the otolaryngologist as may nasal crusting, epistaxis, serous otitis media, laryngitis sicca and a persistent cough with tenacious sputum, glazing of the oral mucosa and sticky secretions in the nasopharynx, etc.

**General examination**

The presence or absence of a connective tissue disorder should be established as should the presence of any of the above-mentioned abnormalities of other organs.
**Investigations**

**Blood examination**

The erythrocyte sedimentation rate is usually raised. A protein profile will show elevation of all the immunoglobulins especially IgG. Rheumatoid factor and antinuclear factor will probably be positive and there may well be a wide range of autoantibodies.

**Specific immunological tests**

These can only be carried out in a few places in the UK. When class 2 antigens such as HLA A1 and B8 and DR3 are examined, then almost three times as many patients with sicca syndrome have these antigens when compared with patients with the secondary syndrome. Specific antigens for Sjögren's syndrome are called SSA and SSB. Again these are more common in patients with the sicca syndrome than in those with secondary Sjögren's disease with rheumatoid arthritis. The immediate clinical relevance of these immunological abnormalities is not known and it may be that they are _in vitro_ epiphenomena.

**Schirmer's test**

This is carried out by putting special strips into the lower fornix. Wetting of less than 5 mm in 5 minutes represent a diagnosis of xerophthalmia. A diagnosis of keratoconjunctivitis sicca, however, cannot be made until the ophthalmologist examines the eye with Rose Bengal dye to see the filamentary keratitis.

**Salivary flow rate**

This is measured using Carlsson-Crittenden cups; these are suction cups placed over the parotid duct. Maximum stimulation is created by getting the patient to suck a lemon. A flow of less than 0.5 mL in a minute represents xerostomia.

**Labial biopsy**

This is performed by obtaining four globules of fat from the back of the lower lip. It can be performed under local anaesthetic and is the diagnostic test for Sjögren's disease. The pathologist must grade it according to the rules laid down.

- Grade 1: slight lymphocytic infiltration
- Grade 2: less than 50 lymphocytes per 4 mm²
- Grade 3: 50 lymphocytes per 4 mm²
- Grade 4: more than 50 lymphocytes per 4 mm².

The distribution of lymphocytes is important also because they cannot be diffuse, but must be periductal. In this test, false positives can be obtained in rheumatoid arthritis, scleroderma, subacute lupus erythematosus, sarcoid, amyloid and graft versus host disease.
Radiology

Sialography either shows a normal sialographic pattern or that of 'globular sialectasis'. This does not imply that the patients with Sjögren's disease have sialectasis. What it does imply is that there is an abnormality in the duct allowing leakage of lipiodol into the stroma of the gland.

Natural history

One in six patients with Sjögren's disease will go on to develop lymphoma. This will be a B-cell type non-Hodgkin's lymphoma. The immunological abnormality in Sjögren's syndrome is a loss of suppressor T-cell activity and an alteration in the T-suppressor-helper cell relationship. As well as a non-Hodgkin's lymphoma they can develop Waldenström's macroglobulinaemia and immunoblastic sarcoma.

The present suggestion as to aetiology is that the cytomegalovirus infects salivary ducts and ducts elsewhere in the body. The ducts act as the antigen and B-lymphocyte proliferation occurs. As well as lymphoma, a peculiar type of anaplastic carcinoma can also develop in these patients. It has been reported predominantly but not solely in Eskimos.

Treatment

There is little of a specific nature that can be done to help these patients. Bouts of parotid swelling may be treated with steroids but the bouts are seldom so severe that they require other immunosuppressive drugs. Artificial tears and synthetic saliva provide limited comfort and bromhexine 40 mg/day sometimes helps a tenacious cough.

The most important feature of treatment, however, is to put these patients on a lymphoma follow-up. Those who have parotid enlargement are at a higher risk of developing lymphoma and diagnostic parotidectomy should be considered.

Salivary gland cysts

Similar to vascular and lymphatic malformations, salivary gland cysts are benign swellings, and merit inclusion in this section.

Most cysts within the parotid or submandibular glands are secondary to sialectasis, or salivary tumours (pleomorphic adenoma, Warthin's tumour, cystic duct adenoma, mucoepidermoid tumour, adenocarcinoma). Cysts not related to other disease more commonly arise from minor salivary glands. In a series of 483 salivary gland cysts, 77% were minor salivary gland mucoceles, and the remainder included parotid duct cysts (10.5%), lymphoepithelial cysts (6%), and ranulae (5%) (Seifert et al, 1986).

Mucoceles are spherical, painless swellings that contain mucus. There are two histological types - extravasation mucoceles (80%), which probably follow repeated minor mucosal trauma, and retention mucoceles (20%) due to duct obstruction by microliths, or inspissated secretions or to bends in the duct.
Extravasation mucocoeles (sometimes called a mucous granuloma) are commoner in younger adults, and show a predilection for certain sites (lower lip 80%, cheek and floor of mouth 15%, and palate, tongue and upper lips 5%). In contrast, retention mucocoeles present in older patients and do not have the marked predilection for certain sites. Treatment of either variety is simple excision.

A ranula is a specific type of salivary gland cyst which arises from the sublingual gland, and is discussed in Chapter 4.

Salivary fistulae

A salivary fistula usually originates from the parotid gland, although it sometimes arises from the submandibular gland. It can be internal or external, congenital or acquired. Internal fistulae drain into the mouth and are therefore often not noticed. Congenital fistulae are rare, and may arise from aberrant or accessory salivary tissue, or be associated with branchial cleft anomalies.

A parotid fistula may be due to surgery, facial trauma, or sepsis within the gland parenchyma. A fistula which follows a partial parotidectomy (especially a lumpectomy) usually arises from the gland parenchyma, and generally drains through the suture line. This type of fistula will close spontaneously in most cases, and until this time, minor leakage will occur with meals. To help prevent such a fistula from developing, the parotid duct is often divided well forward, and ligated in some cases. Saliva may collect underneath the skin flap after surgery, and should be aspirated and a pressure dressing applied. Saliva has a high amylase content compared with fluid from a seroma.

In contrast to the above, a fistula which arises from the main duct system of the parotid leaks profusely, even at the thought of food, and invariably needs an operation to close it. This type of fistula is usually due to a deep facial wound, and the facial nerve may also be damaged. Such fistulae can be difficult to control, and three categories of treatment are described:

1. reduction of saliva production; by drugs, irradiation, gland denervation, and duct ligation

2. operations on the fistula; excision, diversion into the mouth, reconstruction of the damaged duct

3. removal of the gland; partial or total conservative parotidectomy.

Reducing the output of saliva may promote closure of the fistula, and tympanic neurectomy provides a simple effective way of achieving this. Irradiation has been used, but the outcome is uncertain, and there is a risk of later carcinoma. A damaged main duct may be suitable for repair, and this may be demonstrated by a sialogram. However, the outcome of such surgery is uncertain, especially if the fistula has been present for some time, an in such cases parotidectomy may be necessary.
A salivary fistula from the submandibular gland is a much easier problem to deal with. If it does not close spontaneously the gland should be excised.