Management of malignant salivary gland tumours

ED Vaughan

Malignant tumours of both major and minor salivary glands are extremely rare: the parotid gland is most frequently affected by malignant change, followed by the submandibular gland. Malignant tumours of minor salivary glands may occur anywhere in the upper aerodigestive tract, but more common sites are the upper lip, junction of hard and soft palate and paranasal sinuses.

Malignant salivary gland tumours account for 5.5% of head and neck cancers and represent 0.14% of all cancers. The majority occur in the parotid gland, accounting for 20% of all parotid masses, with mucoepidermoid carcinomas being the predominant malignant type.

In the submandibular gland, approximately 50% of submandibular masses are malignant, with adenoidcystic carcinomas being the most common type (40%). In the minor salivary glands, the commonest tumours include mucoepidermoid carcinomas and adenoidcystic carcinomas, and are extremely rare with approximately six cases being seen annually in Merseyside, which has a population of 2.2 million people.

AETIOLOGY

The cause of malignant change in salivary tissue is unknown. However, there is strong histological evidence of benign pleomorphic adenomas undergoing malignant transformation to carcinoma ex pleomorphic adenoma. It has been estimated that the risk of malignant transformation is 20% over 30 years, according to Hollander and Cunningham (1973).

It would appear that there is no substance to the claims of an association with breast cancer (Bigger et al, 1983).

The role of radiation in the aetiology of malignant salivary gland disease is confused, with a reported increased incidence following high dose exposure to radiation. However, no significant difference was reported in a series compiled by Watkin and Hobsley (1986).

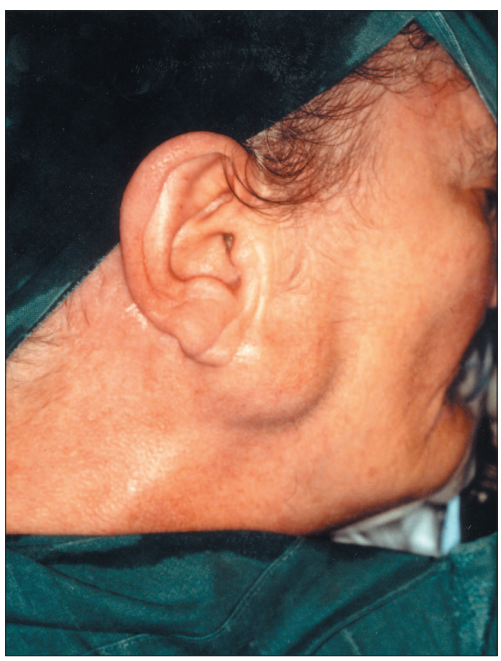
CLINICAL PRESENTATION

In the vast majority of cases, malignant tumours present as painless, asymptomatic lumps in the substance of the parotid or submandibular gland (*Figure 1*). Generally speaking, the patients are usually in the 50–60-year-old age group, with mucoepidermoid carcinomas particularly affecting children.

Facial paresis as a presenting symptom is uncommon, tends to carry a poorer prognosis (*Figure 2*) and occurs in approximately 8–33% of cases, depending on the type of tumour. Again, pain is an uncommon feature and should not be considered pathognomic of malignant disease.

Change in the rate of growth, with sudden increase in size, is considered by some to indicate malignant change. However, this feature

Figure 1. Asymptomatic parotid mass, which was malignant histologically.



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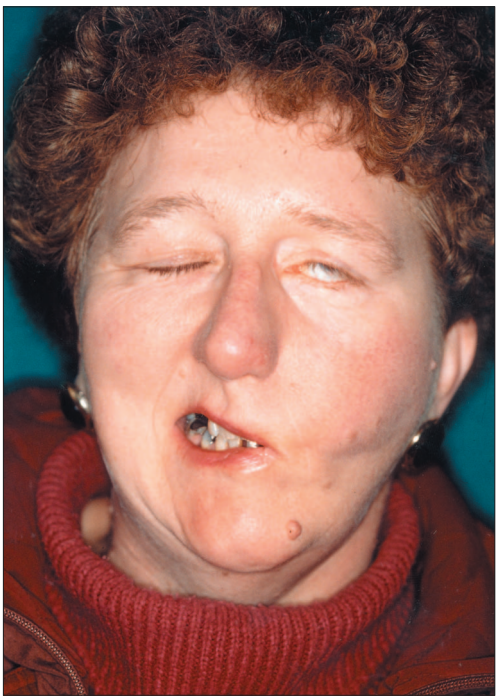


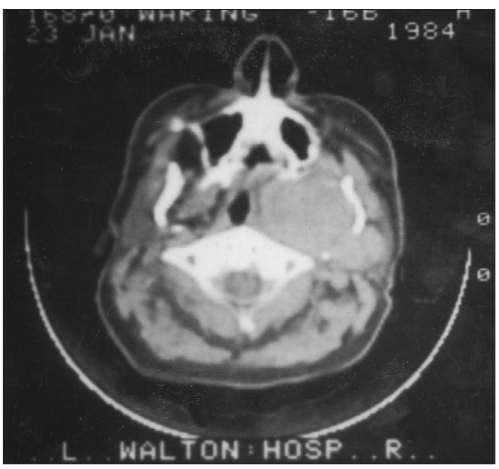
Figure 2. Facial paresis as a presenting symptom of malignant tumours.

occurs as commonly with benign tumours as with malignant tumours.

Generally speaking, should the tumour be located in the deep lobe of the parotid gland, it will be larger and have infiltrated further before being diagnosed, as a result of the loose areolar tissue present. It may present with distortion of the lateral pharyngeal wall or soft palate on intra-oral examination (*Figure 3*).

Trismus and pain in the distribution of the second and third divisions of the trigeminal nerve are worrying signs of infiltration within the infra-temporal fossa.

Figure 3. Axial computed tomography scan of deep lobe parotid tumour.



Overt involvement of the cervical lymph nodes is uncommon and denotes high grade advanced disease. However, 20% of cases have histological evidence of nodal involvement (Ball and Meirion Thomas, 1995).

In the case of minor salivary gland malignant neoplasms, presentation depends on the anatomical position of the tumour, with oral lesions presenting as non-tender sessile swellings, which may have a pink, red or blue appearance depending on the amount of mucoid material contained within the tumour (*Figure 4*). In time, the surface becomes ulcerated and secondarily infected (*Figure 5*).

Tumours arising in the floor of the mouth may become quite large, because of the laxity of the soft tissues there. Clinically, if the location is sino-antral, it may be difficult to distinguish common squamous cell carcinoma from symptoms related to the mouth, including loose teeth or persistence of an oro-antral fistula from tooth extraction. If there is involvement of the nasal cavity, unilateral nasal obstruction and epistaxis are not uncommon but tend to be late features.

Other late manifestations include swelling of the cheek, loss of sensation in the distribution of the infra-orbital nerve. If the tumour extends upwards, then orbital and ocular symptoms may predominate, with proptosis, epiphora and diplopia.

Figure 4. Oral lesion presenting as non-tender sessile swelling.

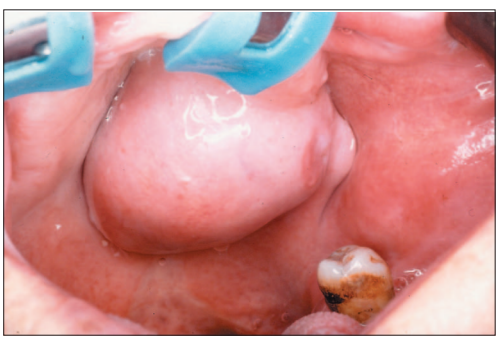
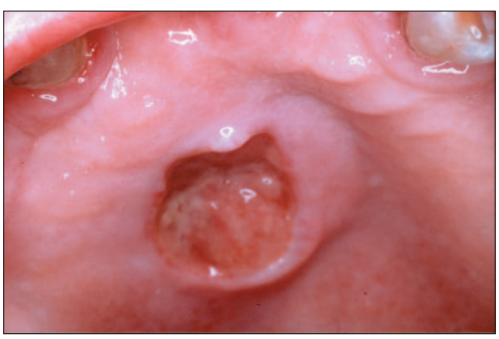


Figure 5. Ulcerated surface of oral lesion.



INVESTIGATIONS

There are some who advance the view that preoperative knowledge of the histological type of tumour is irrelevant to the surgical treatment subsequently provided and, therefore, this school of thought carries out no initial investigations (Spiro, 1986).

Others, on the other hand, attempt to determine the histological nature of the tumour before definitive surgery. They also attempt to define the location and extent of the lesion by radiological means in the hope that the acquisition of such knowledge will allow them to decide whether radical surgical treatment is appropriate.

For major salivary gland lesions, fine needle aspiration (FNA) has become the preferred method of determining histology and is totally dependent on the skill of the cytopathologist. In skilled hands, accuracy of over 90% may be expected.

In cases where there are clinical signs highly suggestive of malignancy, a 'tru-cut' needle biopsy may be performed, taking care to place the needle track in a position where it may be subsequently excised.

Both computed tomography and magnetic resonance scanning are extremely useful in delineating the extent of the tumour, and with a skilled radiologist, in many cases, it is possible to distinguish benign lesions from malignant ones.

For minor salivary gland tumours, one can, with varying degrees of ease and depending on location, perform an incisional biopsy with little or no risk of seeding of malignant cells.

Finally, if at the time of surgery the clinical appearance of the tumour gives rise to the suspicion of malignancy, it is appropriate to obtain a frozen section, providing one is confident of the ability of the pathologist to distinguish malignant disease from benign disease in this notoriously histologically difficult tissue.

TREATMENT

The current consensus in treating malignant salivary gland neoplasms is for primary surgery to eradicate the tumour locally in three dimensions, while maintaining function where at all possible. Depending on the histological type and the adequacy of surgery, postoperative radiotherapy may be required.

In the case of the parotid gland, this generally means a demonstration of the facial nerve and the removal of the entire superficial lobe of the parotid gland (*Figure 6*). If, at the time of surgery, there is obvious infiltration of

Figure 6. Preservation of facial nerve following superficial parotidectomy.

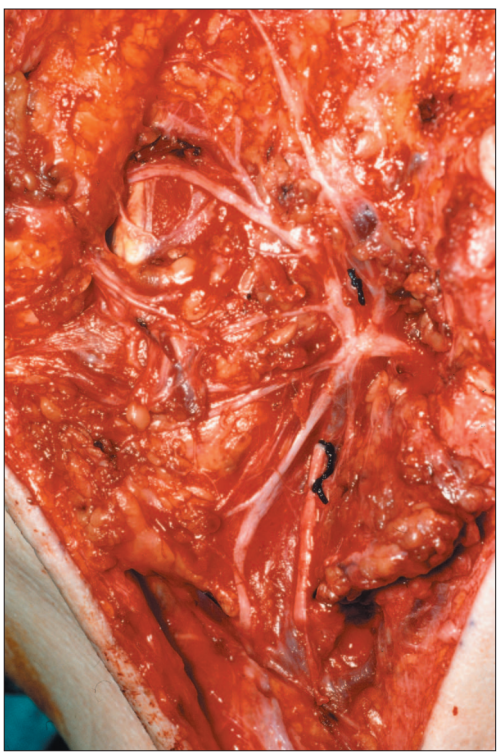
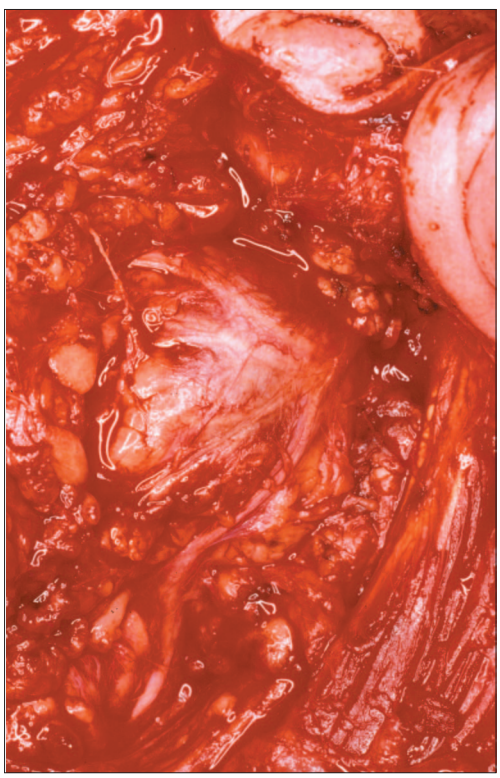


Figure 7. Cervical approach, tumour deep to branches of the facial nerve.



branches of the facial nerve, a decision has to be made whether or not it is appropriate to sacrifice these branches. The general view is that sacrifice of branches of the facial nerve is a manoeuvre of last resort, and in most cases, one should rely on postoperative radiotherapy to prevent local recurrence (Jackson et al, 1983).

Should the tumour involve the deep lobe of the parotid gland, having performed a superficial parotidectomy with preservation of the facial nerve, it may be necessary to gain access to the infra-temporal fossa by a variety of means:

Via a cervical approach
One usually combines this approach (*Figure 7*) with some form of neck clearing procedure, and access is obtained to the lower and posterior aspects of the infra-temporal fossa.

Figure 8. Access to antero-inferior aspect of infra-temporal fossa.

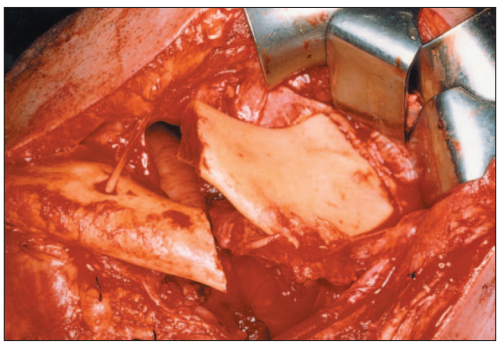


Figure 9. Mid-face approach to the infra-temporal fossa.

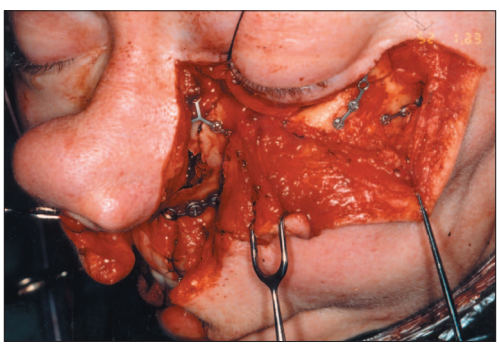
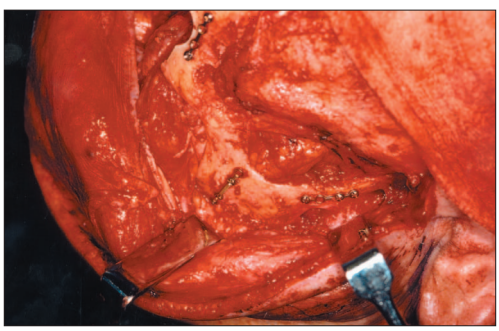


Figure 10. Subtemporal approach to the infra-temporal fossa.



Via a mandibulotomy
Access is gained (*Figure 8*) to the anterior aspect of the infra-temporal fossa.

Via a mid face approach (Altemir)
This approach (*Figure 9*) will allow access to the roof and anterior aspects of the infra-temporal fossa.

Via a subtemporal approach
In this approach (*Figure 10*), the zygomatic bone is removed and allows access to the roof of the infra-temporal fossa.

Via a mandibulectomy approach
Access is gained to virtually the entire infra-temporal fossa (*Figure 11*).

In high-grade tumours, where there is no possibility of preserving the facial nerve, it is appropriate to sacrifice the facial nerve and, if necessary, the overlying skin (*Figure 12*). The resultant facial nerve defect is reconstructed with either a vascularized or non-vascularized radial nerve graft harvested from the non-dominant forearm (*Figure 13*). The graft may be combined with a fascio cutaneous-free radial forearm flap (*Figure 14*). The results of such reconstructions are acceptable, even when postoperative radiotherapy has been employed (*Figures 15, 16* and *17*) (Jackson et al, 1983; Vaughan and Richardson, 1993).

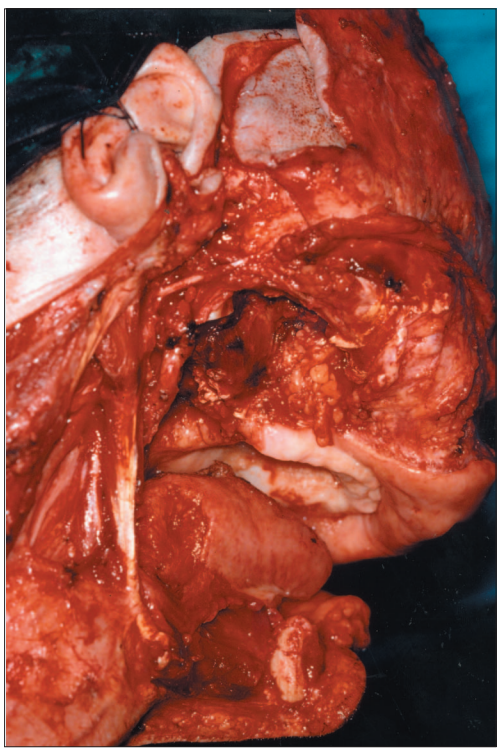


Figure 11. Mandibulectomy to access the infra-temporal fossa.

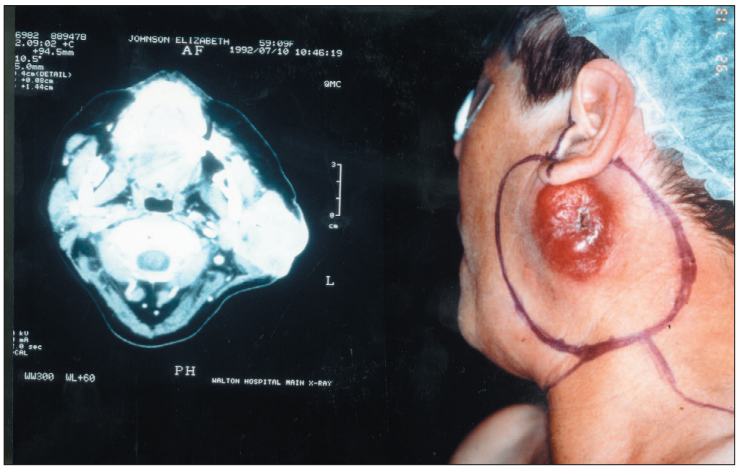


Figure 12. High-grade tumours may require sacrifice of the facial nerve and overlying skin.

Neck dissection

The role of neck dissection in the management of malignant salivary gland disease is not clear. In the case of an asymptomatic parotid swelling which subsequently proves to be malignant, the factor which will decide whether it is appropriate to perform a neck dissection will be the histological type. In the case of high-grade mucoepidermoid and adenoid cystic carcinomas, carcinoma ex pleomorphic adenoma and other adenocarcinomas, it would be reasonable to perform a selective function-preserving neck dissection, to include levels I–V secondarily.

Where there are obvious signs of malignancy and where there are involved neck nodes, again a selective neck dissection should be performed in continuity with the primary resection. The morbidity from this added procedure is slight. A similar attitude may be adopted for the submandibular gland.

Radical neck dissections are not normally performed for malignant salivary gland tumours unless the neck involvement is massive.

In the case of minor salivary glands, it is usual to adopt a wait-and-see policy, responding surgically if the neck subsequently becomes involved.

Malignant tumours of salivary gland origin in children are excessively rare. They generally appear in the second decade and roughly 50%

Figure 13. Reconstructing a facial nerve defect with a radial nerve graft.

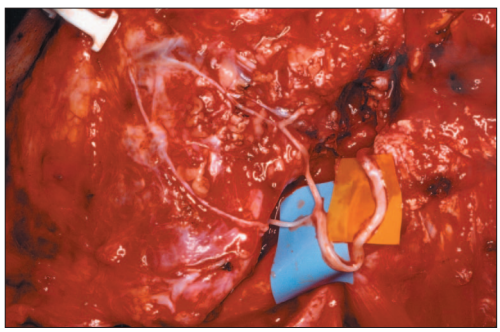


Figure 14. Graft combined with a fascio cutaneous-free radial forearm flap.

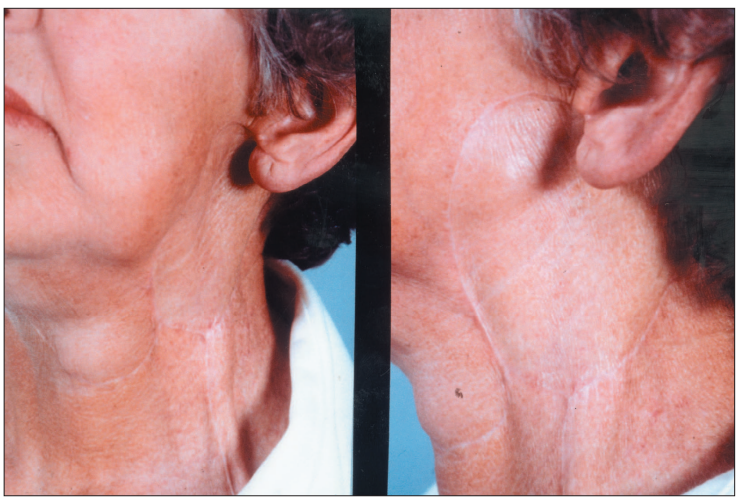
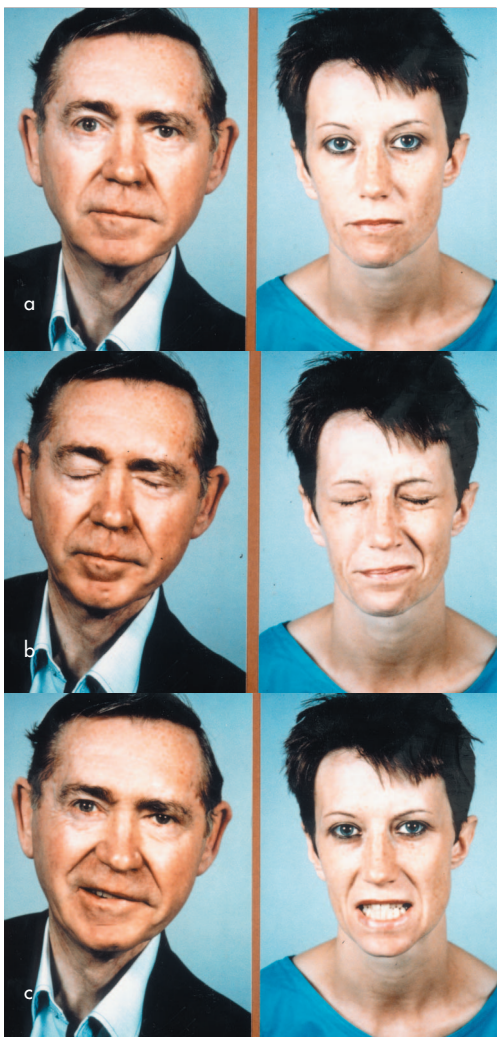


Figure 15. Facial nerve reconstruction in terms of (a) facial symmetry, (b) eye closure and (c) ability to smile.



are malignant with mucoepidermoid carcinomas predominating. It is wise to consider all childhood salivary gland tumours malignant until proved otherwise. However, preservation of the facial nerve is of paramount importance.

Radiotherapy

Malignant salivary gland tumours are moderately radiosensitive, and radiotherapy is employed in an adjunctive role, particularly when there are incomplete excision margins and when the tumour is high grade (Calman, 1995). Such combination therapy gives local control rates comparable to those when complete surgical clearance has been obtained.

Where the tumour is surgically unresectable, hyperfractionated radiotherapy to 70+ Gy has been reported to give very good control rates (Wang and Goodman, 1991), but there is no evidence of an improved survival benefit.

Fast neutron therapy has been advocated by Caterall and Errington (1987) as a means of treating unresectable malignant salivary gland tumours. However, the long-term morbidity of this treatment modality has now rendered it unacceptable according to RD Errington (personal communication, 1995).

OUTCOME

A variety of factors play a role in the prognosis of patients with malignant salivary gland tumours, chief among them being the grade of the tumour, with high-grade tumours having a worse prognosis. As with all malignant tumours, the degree of local development influences the outcome, with small tumours invariably having a better prognosis than large ones.

Carcinoma ex pleomorphic adenoma and poorly differentiated adenocarcinomas fare least well, while the indolent nature of adenoid cystic carcinomas is well recognized. Unfortunately, the long-term survival of patients suffering from adenoid cystic carcinoma is poor, with few survivors at 20 years.

It is well recognized that patients with malignant tumours of the submandibular gland do less well than patients who have involvement of the parotid gland.

In the case of malignant minor salivary gland tumours, especially those of the oral cavity, there is an impression that survival is better than similar type tumours involving the major salivary glands. Unfortunately, it is difficult to confirm this impression, because of the rarity of these tumours. HM

Conflict of interest: none.

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KEY POINTS

- Malignant salivary gland tumours are extremely rare.
- The aetiology is unknown.
- The major salivary glands are most commonly affected, with the parotid gland being most frequently involved.
- Mucoepidermoid carcinomas are most common in the parotid.
- Adenoidcystic carcinomas are commonest in the submandibular gland.
- Investigation of these tumours is dependent on the site of origin and includes fine needle aspiration, tru-cut needle biopsy and frozen section.
- Radiological investigation involves both computed tomography and magnetic resonance scanning.
- The primary modality of treatment is surgery, with every effort made to preserve the facial nerve.
- Where tumour excision is incomplete or inadequate, adjunctive radiotherapy is indicated.
- Outcome is dependent on site, tumour type and histological grading.