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Adenoid cystic carcinoma of the sublingual salivary gland in a 16-year-old female – report of a case and review of the literature

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Abstract
Tumours of the sublingual salivary gland are exceptionally rare. The present case report describes an adenoid cystic carcinoma of the sublingual salivary gland occurring in a 16-year-old girl, in itself an uncommon event. In addition, an interesting feature of the presentation was obstruction of the ipsilateral submandibular gland due to involvement of Wharton's duct.

Key words: Sublingual gland; Carcinoma

Introduction

Adenoid cystic carcinoma is an uncommon malignancy of the seromucinous glands of the head and neck, accounting for only four per cent of all major salivary gland neoplasms (Eveson and Cawson, 1985) and 11 per cent of malignant neoplasms (Spiro *et al.*, 1974). It is characterized by extensive, insidious invasion along fascial planes, bony cancellous spaces and perineural sheaths. It has a persistent recurrence pattern and a propensity for distant metastases, (Conley and Dingman, 1974; Seaver and Keuhn, 1979; Andersen *et al.*, 1991). Tumours of the sublingual salivary glands are rare and comprise only 0.3 to one per cent of all salivary neoplasms (Eveson and Cawson, 1985; Spiro *et al.*, 1989; Batsakis, 1991) although up to 80–90 per cent of these are malignant. Adenoid cystic carcinoma may account for 36 per cent of sublingual salivary gland carcinomas (Nishima *et al.*, 1984). The peak age incidence of salivary gland tumours is in the sixth and seventh decades and as few as 0.6 per cent of malignant tumours occur before the age of 20 years (Nishima, 1984; Spiro *et al.*, 1989).

Most patients with adenoid cystic carcinoma present with a mass which may have been present for months or years. Pain is a feature in a minority of patients and is more often associated with advanced or recurrent tumours (Andersen *et al.*, 1991). Large tumours may eventually interfere with function of the oral and nasal cavities, the orbit or the ear and parotid lesions can present as facial nerve paresis. Ulceration is rare in major gland adenoid cystic carcinoma and Conley and Dingman (1974) have suggested that any non-ulcerating submucosal tumour causing mild discomfort should be considered adenoid cystic carcinoma until proved otherwise by biopsy. Only one case of sublingual salivary gland adenoid cystic carcinoma obstructing the submandibular duct appears to have been previously reported (Whear and Addy, 1993). The rarity of this lesion at this site may lead to diagnostic error or delay in definitive management, especially when the presentation is of relatively common obstructive symptoms. A case of adenoid cystic carcinoma of the sublingual salivary gland is presented to illustrate this pitfall.

Case report

A 16-year-old female was referred with mild obstructive symptoms of the right submandibular gland and a swelling of the right floor of her mouth of one month duration. She had experienced discomfort in the floor of her mouth which was referred to the right perauricular region. Examination showed a firm mass measuring 2.5 cm × 1 cm in the region of the right sublingual salivary gland. There was fullness and tenderness of the right submandibular gland with absence of salivary outflow. There was no palpable lymphadenopathy and no neurological deficit. Plain radiography failed to demonstrate a salivary calculus. A magnetic resonance image (MRI) scan showed an enhancing tumour mass arising in the right sublingual space, supero-medial to the mylohyoid muscle. The tumour extended postero-laterally where it was closely related to the superior aspect of the right submandibular gland and infiltrated the right medial pterygoid muscle (Figure 1). There was no evidence of more distal spread or cervical lymphadenopathy.

Fine needle aspiration of the mass revealed a cellular specimen consisting of sheets of bland-appearing epithelial cells arranged around magenta-coloured globules of basement membrane material, some of which was fibrillary and some sharply defined. The appearances were suggestive of either adenoid cystic carcinoma or pleomorphic adenoma. Incisional biopsy confirmed infiltrating adenoid cystic carcinoma.

The patient underwent a right supraomohyoid neck dissection via a U-shaped incision with in-continuity resection of the tumour mass and floor of mouth. The right hypoglossal nerve and the right lingual nerve were sacrificed. Multiple frozen sections from the excision

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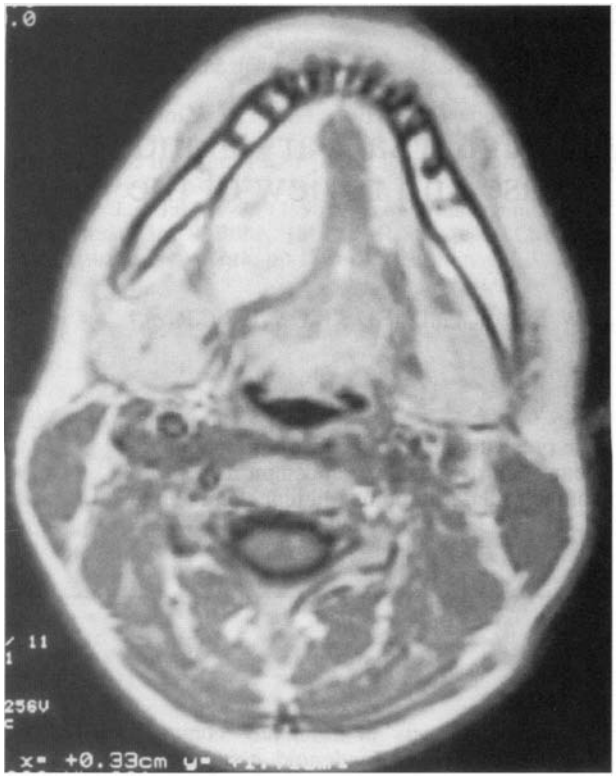

FIG. 1

Magnetic resonance image scan showing tumour mass arising in the right submandibular space.

margins were negative. The surgical defect was reconstructed with a superiorly-based platysma myocutaneous flap. Post-operative tongue function and speech were excellent despite the sacrifice of the hypoglossal nerve.

Histological sections showed a cribiform and tubular adenoid cystic carcinoma arising from the sublingual salivary gland with evidence of perineural and vascular invasion (Figures 2 and 3). Excision was judged to be complete. The patient remains free of tumour after 18 months follow-up.

Discussion

Microscopically adenoid cystic carcinoma is composed of small round cells with hyperchromatic nuclei and eosinophilic cytoplasm. The cells are typically arranged in cords or sheets fenestrated by pseudo-glandular spaces filled with mucoid or hyaline material creating the classic cribriform (cylindroma) pattern seen in all tumours (Nascimento *et al.*, 1986). The tubular variant of this basic pattern is considered by some to be the best differentiated form and has cells arranged in duct-like cords but evidence is conflicting as to whether there is any significant difference in behaviour between tubular (grade I) types and cribiform (grade II) types (Perzin *et al.*, 1978; Nascimento *et al.*, 1986). Occasionally, there are areas of solid cellular growth with a basaloid appearance and few if any fenestrations (histological grade III). These tumours may show cellular atypia and are associated with a more aggressive course than tubular-cribriform types, especially where the solid component makes up more than 30 per cent of the tumour mass (Szanto *et al.*, 1984).

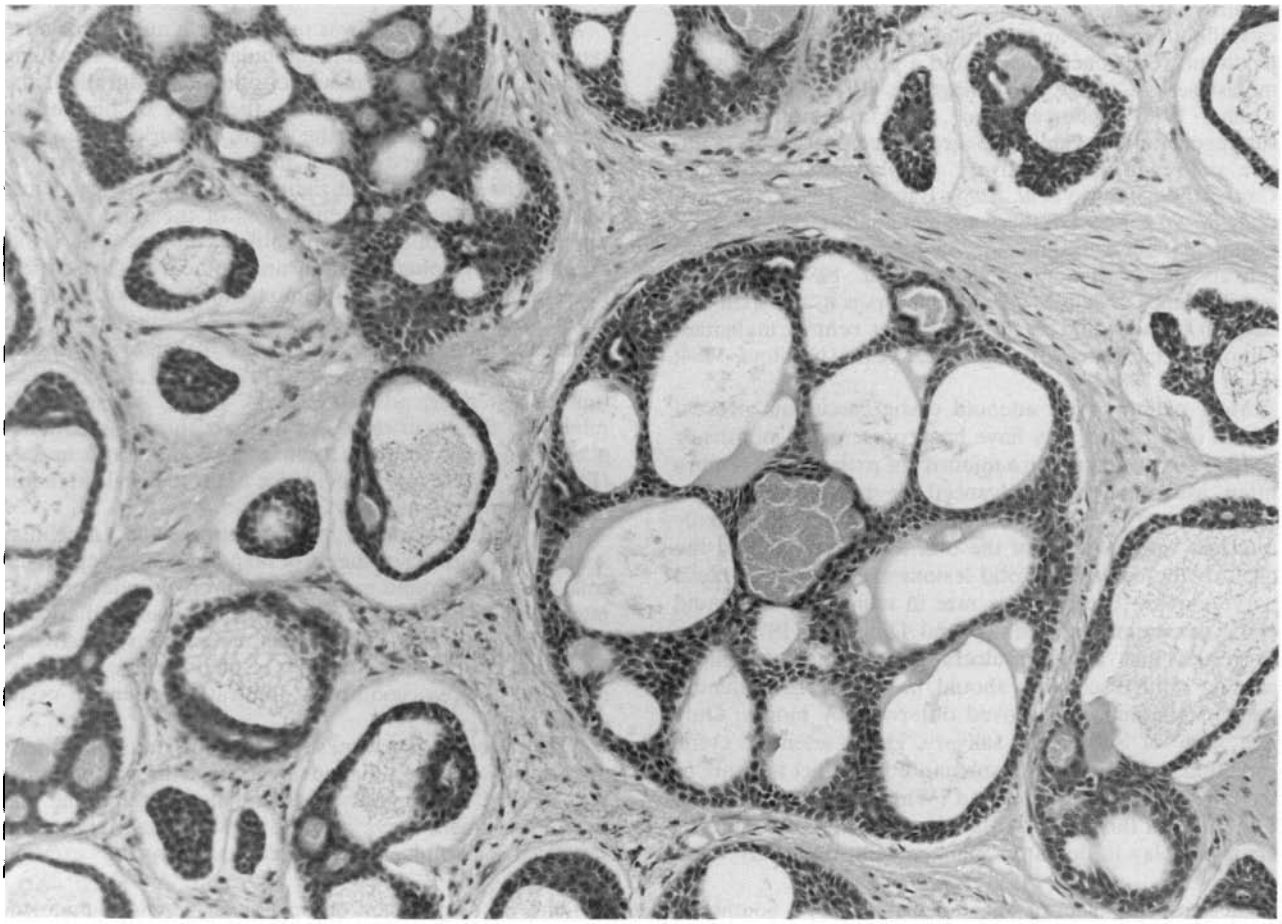

FIG. 2

Adenoid cystic carcinoma showing typical cribriform (cylindromatous) configurations (H & E; × 150).

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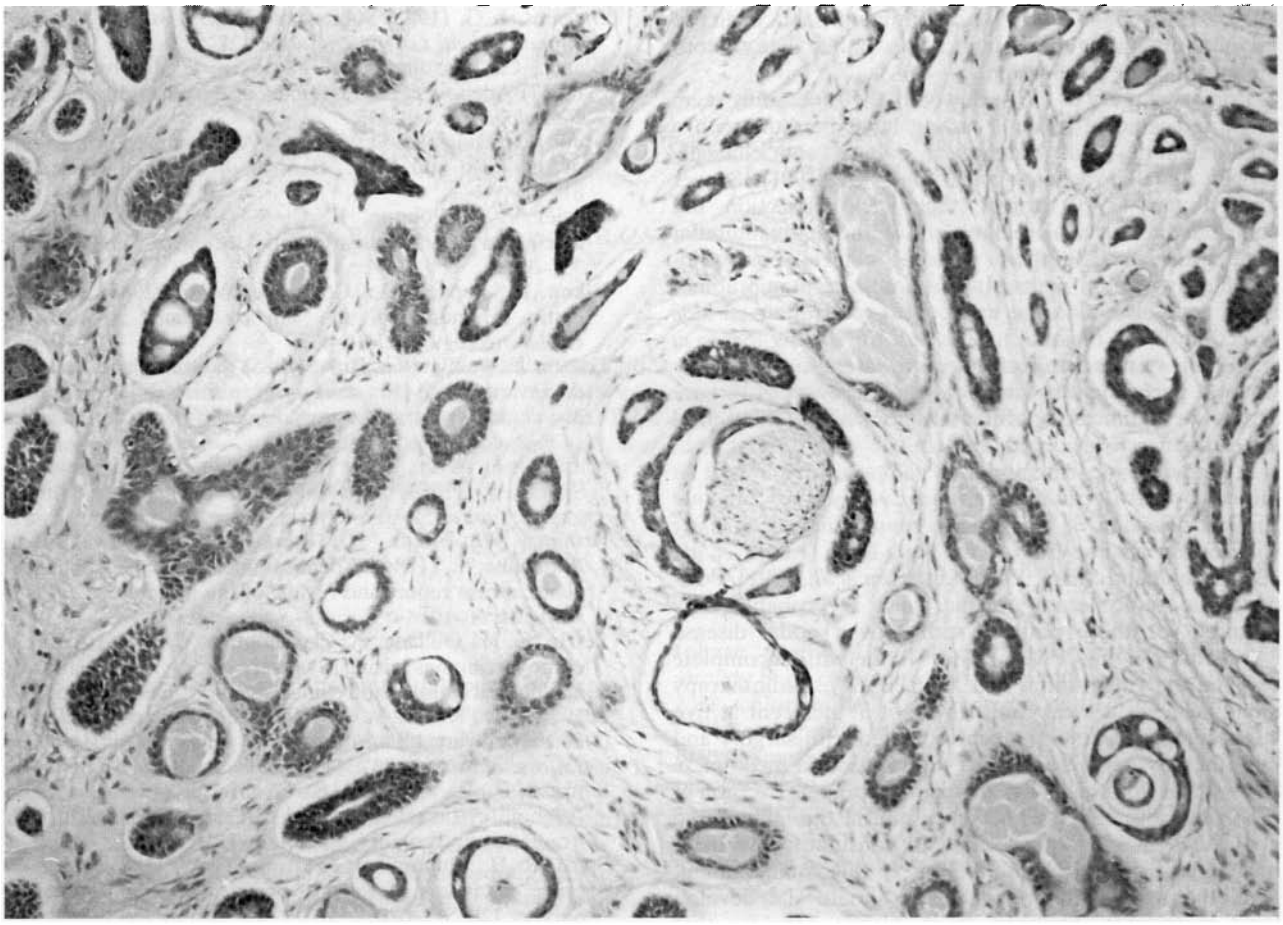

FIG. 3
Adenoid cystic carcinoma showing predominantly tubular variant and perineural invasion (H & E; × 150).

It is generally reported that patients with adenoid cystic carcinoma have a prolonged survival albeit with persistent disease. Local failures and metastases may not become evident for 10 years or more after initial treatment and the clinical course is characterized by repeated surgical interventions (Seaver and Keuhn, 1979). Survival rates differ considerably between reported series with rates at five, 10 and 15 years of 37–75 per cent, nine to 28 per cent and three to 20 per cent respectively (Conley and Dingman, 1974; Seaver and Keuhn, 1979; Nascimento *et al.*, 1986; Ampil and Misra, 1987). The prognosis of adenoid cystic carcinoma is influenced by a variety of factors including the histological type, clinical stage, site and adequacy of primary treatment. Some authors report that younger patients and patients with symptoms of shorter than one year duration may have better survival rates (Nascimento *et al.*, 1986; Spiro *et al.*, 1989). Those with tumours of the palatal or parotid glands may fare better than those with tumour in the submandibular gland or minor glands (Szanto *et al.*, 1984). The UICC staging system, based on size of the primary tumour and presence of lymph node and distant metastases has been shown to correlate well with survival and is probably the single most important prognostic factor (Spiro *et al.*, 1989; Andersen *et al.*, 1991). However, the issue is clouded by the fact that tumour size is also related to histological grade (Nascimento *et al.*, 1986).

Perineural spread is seen in up to 76 per cent of adenoid cystic carcinomas and has been implicated in the propensity of these tumours for local and distant recurrence (Blanck *et al.*, 1967). Vrielinck *et al.*, (1988) found that five-year survival for those with perineural invasion was 36.9 per cent compared to 93.8 per cent for those without nerve invasion. Other investigators have failed to demonstrate any prognostic significance of perineural invasion but found that adenoid cystic carcinoma may progress through bone without destruction of the basic trabecular pattern and that bone involvement by tumour carries a poor prognosis (Perzin *et al.*, 1978; Szanto *et al.*, 1984; Nascimento *et al.*, 1986). Metastatic disease may be apparent in only five per cent of cases at presentation but up to 50 per cent will subsequently develop metastasis in the lung (41 per cent), brain (22 per cent), cervical lymph nodes (16 per cent), bone (13 per cent) and other organs e.g. skin, pleura and liver (four per cent), (Conley and Dingman, 1974; Simpson *et al.*, 1984; Andersen *et al.*, 1991). Patients with metastatic disease may survive for up to seven years without treatment. (Andersen *et al.*, 1991) Ampil and Misra (1987) consider that almost all patients eventually have recurrence.

There is little doubt that surgical resection is the treatment of choice although resection is often inadequate. Conley and Dingman (1974) advocate the 'biggest operation that can be reasonably developed' and consider that primary surgery is the best opportunity for cure. Despite this, 44–55 per cent of patients undergoing surgery will develop local recurrence especially if resection margins are not free of tumour (Elkon *et al.*, 1980; Simpson *et al.*, 1984). Palliation may be indicated when patients present with clinical signs of nerve infiltration as radical surgery may not increase chances of cure but significantly increases morbidity (Stell *et al.*, 1985). Regional lymph node involvement by adenoid cystic carcinoma is uncommon, occurring in only four to 13 per cent of cases (Spiro *et al.*,

1974; Armstrong *et al.*, 1991) and in many cases is due to invasion of continguous lymph nodes rather than tumour embolization or permeation.

Primary surgery of the neck would, therefore, only seem to be indicated in patients having large tumours with a predominantly solid histological pattern and clinically proven nodal invasion (Armstrong *et al.*, 1991). Seaver and Keuhn (1979) found that neck dissection did contribute to survival in tumours of the submandibular region. In our case there were no clinically or radiographically evident nodes. In this case a right supraomohyoid neck dissection was carried out to facilitate *en-bloc* resection of the regional nerves and for staging (Spiro *et al.*, 1989). A more radical or contra-lateral neck surgery was not performed as there was no extension of disease across the midline and perineural spread across this neural watershed was considered unlikely.

Adenoid cystic carcinoma is radiosensitive but it is difficult to justify radiotherapy in cases where complete surgical resection of tumour has been achieved as many will remain disease-free in the long-term (Elkon *et al.*, 1980; Ampil and Misra, 1987; Andersen *et al.*, 1991). Postoperative radiotherapy to the neck is probably only required for those patients with proven nodal disease (Armstrong *et al.*, 1991) or for those with incomplete excision margins (Ampil and Misra, 1987). Radiotherapy alone achieves a local control rate of 37 per cent at five years compared to 86 per cent obtained with surgery and radiotherapy combined where the excision is incomplete (Cowie and Pointon, 1984). Given that compete excision of the primary tumour was achieved and in the light of the above evidence it was decided in consultation with an oncologist not to give radiotherapy to this young patient. Radiotherapy will be reconsidered should she develop local recurrence. Simpson *et al.* (1984) consider that patients in whom local recurrence develops after surgery should be treated vigorously with radiotherapy as disease control in these cases may be as good as those who were treated with radiotherapy at the time of diagnosis. In the long-term the major threat to this patient's survival is likely to be metastatic disease (Spiro *et al.*, 1974; Ampil and Misra, 1987).

In conclusion, adenoid cystic carcinoma of the salivary glands is unpredictable and 'cure' cannot be assumed even after many disease-free years. A high index of clinical suspicion and life-long follow-up must be adopted for all patients with adenoid cystic carcinoma.

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