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View from beneath: Pathology in Focus
Synovial sarcoma of hypopharynx

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Abstract
Synovial sarcoma of the hypopharynx is a rare neoplasm. To date only 23 cases of synovial sarcoma of the hypopharynx have been reported in the literature. An additional case in an 18-year-old male is presented. This is the first case of synovial sarcoma in the hypopharynx to be reported in Singapore. The presentation was that of a mass in the hypopharynx; progressive dysphagia, intermittent hoarseness and gradual airway compromise. A CT scan was valuable in determining the site of origin and extent of the lesion. Histopathology was diagnostic. Treatment comprised of wide surgical excision of the tumour and post-operative radiotherapy.

Introduction

Synovial sarcoma constitutes only 8–10 per cent of all soft tissue sarcomas. Ninety per cent of synovial sarcomas occur in the extremities. Synovial sarcomas of the head and neck are rare; fewer than 80 cases have been reported in the literature. In the head and neck the neoplasm has been reported in the neck, cheek, nasopharynx, tongue, hypopharynx and the larynx. To date 23 cases of synovial sarcoma in the hypopharynx have been reported in the literature. We report one additional case in an 18-year-old Malay male. This is the first case of synovial sarcoma of the hypopharynx to be reported in Singapore.

Case report

An 18-year-old Malay male was referred to the Department of Otolaryngology, Singapore General Hospital in January 1991 with a two-month history of progressive dysphagia and intermittent hoarseness. He also complained of dyspnoea on exertion one month prior to presentation and had lost almost four kilogrammes of his body weight during the period. On examination, a large lobulated mass was visible behind the base of tongue. Fibreoptic examination of the pharynx showed the mass to be almost filling the entire hypopharynx obscuring the view of the larynx. No regional lymph nodes were palpable in the neck.

The patient was admitted to the hospital. Routine laboratory investigations were within normal limits. Chest radiograph excluded pulmonary metastasis. A CT scan revealed a large mass arising from the left lateral hypopharyngeal wall obstructing the laryngeal inlet (Fig. 1). The thyroid cartilage and vocal cords were uninvolved. As endotracheal intubation was not possible, a tracheostomy under local anaesthesia, was performed prior to the assessment. Subsequent direct laryngoscopic examination under general anaesthesia showed a large lobular mass arising by a flattened stalk from the lateral wall of the pyriform fossa. The mass was flopping onto the laryngeal inlet causing a 'ball valve' obstruction. The mass was excised trans-orally by transecting the stalk and was sent for histology. Biopsies were also taken from the tumour pedicle and base.

The main tumour mass was a piece of greyish soft translucent tissue measuring 4 × 4 × 1 cm. Foci of haemorrhage were seen. Histological examination showed a highly cellular tumour consisting of spindle cells with scant and indistinct cytoplasm. They were arranged in fascicles. The mitotic count was high and averaged 5 per high power field. A notable feature was the presence of cuboidal to columnar epithelial cells arranged in cords as well as around gland-like spaces.

The spaces contained PAS and mucin positive material. This epithelial component gave a biphasic histological pattern to the tumour and a diagnosis of synovial sarcoma was made (Fig. 2). The case was presented at the Head and Neck Tumour Board and

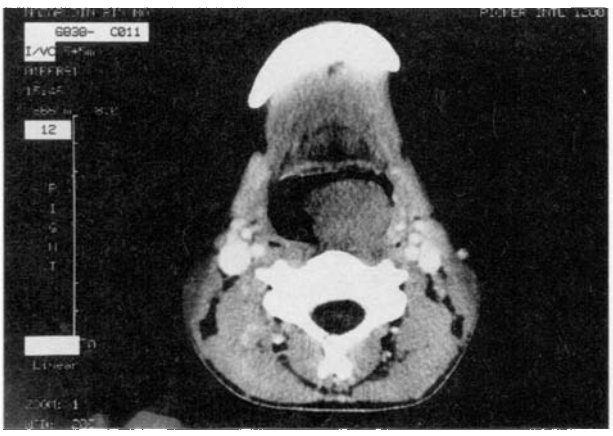

Fig. 1
CT scan demonstrating the tumour arising from the hypopharynx.

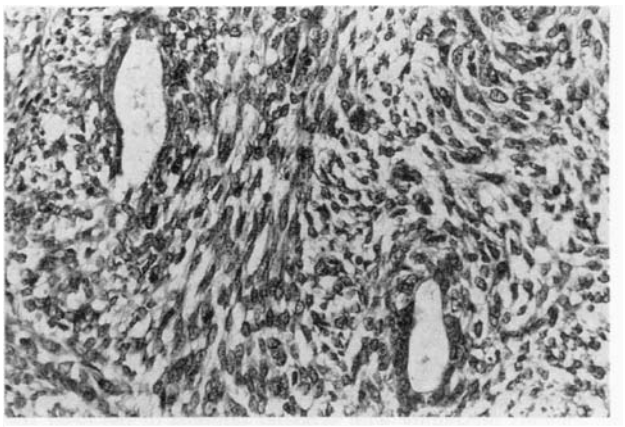

Fig. 2
Photomicrograph of the tumour showing the classical biphasic pattern of synovial sarcoma. H & E stain. Original magnification ×400.

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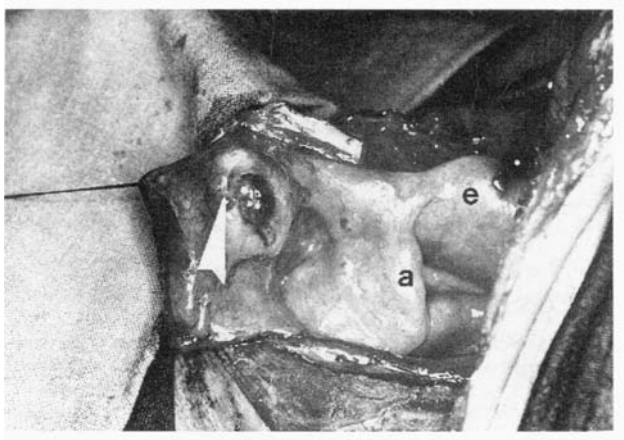

FIG. 3

Tumour as approached from a lateral pharyngotomy. White arrow head pointing to the tumour base. a = left aryepiglottic fold, e = epiglottis.

it was decided to treat the patient with combined therapy modality, comprising of wide excision of the tumour followed by post-operative radiotherapy. A wide excision of the tumour base using a left lateral pharyngotomy approach was carried out (Fig. 3). The surgical specimen consisted of 1 cm diameter tumour base with 2 cm margin around it (Fig. 4). The resected margins and base of the specimen were free of tumour both on frozen and paraffin sections. The pharyngeal defect was closed primarily. The patient's post-operative course was uneventful and he was discharged 12 days after the surgical procedure. The tracheostomy tube was removed before discharge. There was no hoarseness and the patient was able to swallow liquids and solids well. Radiotherapy was started six weeks post-operatively and administered using 10 mv linear accelerator. A total dose of 6000 cGy was administered over 50 days in 34 fractions. The treatment volume initially consisted of the entire hypopharynx and the regional lymph nodes. The field was gradually reduced to include only the tumour site. It has been six months since the patient completed treatment. He has been on monthly follow-up in the clinic and so far has remained free of recurrence.

Discussion

Synovial sarcomas account for 8–10 per cent of all soft tissue sarcomas (Pack and Ariel, 1950). Ninety per cent of synovial sarcomas primarily occur in the extremities (Cadman *et al.*, 1965). Synovial sarcomas in the head and neck are rare. It was first described in the head and neck when Jernstrom (1954) reported a case of synovial sarcoma of the pharynx. Since then there have been isolated reports of synovial sarcomas in the head and neck (Krugman *et al.*, 1975; Hirokawa *et al.*, 1980; Applebaum and Mantravadi, 1983; Holtz and Magieleski, 1985). A large series reporting follow-up of 24 cases of synovial sarcomas of the head and neck was reported by Roth *et al.* (1975). Their report was based on 24 surgical specimens removed from the region of the neck in the period 1949 to 1969. The first case reported by Jernstrom was also included in the series.

The hypopharynx has been described as a very unusual and rare primary site for synovial sarcomas (Gapany *et al.*, 1978). However, review of the literature shows that more cases of synovial sarcomas have been reported in the hypopharynx than any other single site in the head and neck. In the Roth *et al.* series of 24 cases of synovial sarcomas in the head and neck, 10 occurred in the hypopharynx. Of the synovial sarcomas reported in the head and neck, 23 cases have been reported in the hypopharynx.

Patients presenting with synovial sarcoma of the head and neck are usually young, aged between 20 to 35 years. When the tumour occurs in the hypopharynx the usual symptoms are pain, dysphagia and hoarseness. Metastasis to regional lymph nodes occurs in 12.5 per cent of cases in the head and neck (Pack and Ariel, 1950; Cadman *et al.*, 1965; Roth *et al.*, 1975). In Roth's series of 24 cases of synovial sarcoma in the head and neck, 10 patients died of pulmonary metastasis.

Synovial sarcomas do not necessarily arise from the synovial membrane. In fact, the majority of the cases reported in the head and neck arise at sites which have no apparent relation to synovial structures such as bursae or joints. A theory suggesting that these tumours arise from undifferentiated mesenchyme which retain the potential to synovioblastic differentiation was suggested by Jernstrom (1954) and later by Mackenzie who in 1966 wrote 'Synovial sarcomas are not so named because they arise from synovial membrane or from tendon sheaths or bursae. Most of them do not. They are so-named because they carry the hallmarks of a particular type of mesenchymal differentiation.'

Histologically the neoplasm is classically biphasic manifesting both spindle cells and epithelial cells. According to MacKenzie (1966) this cellular duality is the only histological criterion by which synovial sarcomas may be diagnosed with certainty. MacKenzie (1966) also described the monophasic variants where either cell type predominates. These variants present considerable diagnostic difficulties and may be mistaken for undifferentiated fibrosarcomas or other mesenchymal tumours.

Various treatment modalities have been used to treat these tumours in the head and neck; these include: surgical excision (Roth *et al.*, 1975; Gapany *et al.*, 1978; Quinn, 1984) combined modality comprising of surgical excision with pre-operative or post-operative radiotherapy (Roth *et al.*, 1975; Hirokawa *et al.*, 1980; Applebaum and Mantravadi, 1983) and adjuvant chemotherapy (Gatti *et al.*, 1975; Holtz and Magieleski, 1985; Moore and Berke, 1987).

There have been encouraging reports of increased survival rates in patients with soft tissue sarcomas who underwent combined modality treatment comprising of surgical excision and adjunctive radiotherapy (Pack and Ariel, 1950; Lindberg *et al.*, 1981). However the value of combined therapy for synovial sarcomas in the hypopharynx is not established because of the rarity of these tumours and the varied treatment policies adopted in the patients reported in the literature. There have been reports of use of adjunctive systemic chemotherapy (Gatti *et al.*, 1975; Holtz and Magieleski, 1985; Moore and Burke, 1987) but the experience and the data available is insufficient to permit an evaluation of the various chemotherapeutic combination.

Our patient conforms to case reports of synovial sarcoma of the hypopharynx reported by others (Gapany *et al.*, 1978; Hirokawa *et al.*, 1980; Applebaum and Mantravadi, 1983). In keeping with favourable results with combined modality of treatment, we treated our patient with wide local excision and post-operative radiotherapy. It has been six months since the patient completed treatment. The patient is on regular follow-up in the clinic and there has been no evidence of recurrence so far.

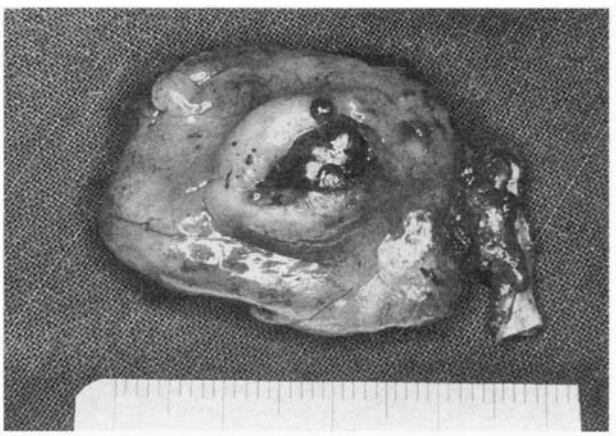

FIG. 4

The excised surgical specimen.

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