The Journal of Laryngology & Otology
May 2000, Vol. **114,** pp. 381–384

Angiosarcoma of the maxillary sinus

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Abstract

Angiosarcomas are rapidly growing malignant neoplasms arising from the vascular endothelial cells. Most common sites are the extremities and the retroperitoneal space, with only four per cent of angiosarcomas arising in the head and neck area, whilst the paranasal sinuses are one of the rarest locations.

We report the case of a maxillary sinus angiosarcoma in a 72-year-old male patient. The first biopsy was inconclusive, whereas the second revealed an angiosarcoma. Medial maxillectomy was performed with subsequent external irradiation.

Key words: Haemangiosarcoma; Maxillary sinus, surgery

Introduction

Angiosarcoma is a rare malignant tumour of the vascular endothelium characterized by the formation of irregular vascular channels lined by one or more layers of atypical endothelial cells, often of immature appearance, and accompanied by solid masses of poorly differentiated anaplastic tissue.[1] Angiosarcomas comprise less than one per cent of all sarcomas.[2]

Although angiosarcomas may occur at any location in the body, they have a predilection for skin and superficial soft tissue.[2] They often arise in the liver, spleen and heart. In addition, the tumour's aggressive behaviour and poor prognosis, with a five-year survival rate of only 10–20 per cent, have been documented in the literature.[3]

Angiosarcomas of the head and neck are extremely rare with the scalp being the most common site.[1] The angiosarcoma of the maxillary antrum is one of the rarest locations. Only 19 cases have been reported in the literature. In this report, we present the case of an angiosarcoma arising from the right maxillary sinus, we review the literature and we discuss the diagnosis and management.

Case report

A 72-year-old man presented with severe epistaxis of the right nasal cavity. Four months before, he had been treated at another hospital for a mass of the right maxillary sinus, and had undergone a Caldwell-Luc operation. The mass had been discovered because of epistaxis. The histological examination of the mass showed no evidence of malignancy.

The clinical examination of the patient's nose showed an irregularly shaped haemorrhagic mass with a polypoid configuration occupying the right nasal cavity. No other abnormality was detected. A computer tomography (CT) scan showed that the lesion was filling most of the right maxillary sinus (Figure 1) and was penetrating through the bony margins of the medial wall. An intranasal biopsy was obtained and reported as angiosarcoma. Ten days later, a right medial maxillectomy was performed with a standard Weber-Ferguson approach. Multiple tissue fragments of reddish colour and friable consistency were removed from the right maxillary antrum and the ethmoid sinuses. Histology showed most of these tissues to be angiosarcoma. The tumour consisted partly of vascular spaces lined by atypical cells, whereas elsewhere it had a more compact appearance. Very few mitoses were seen. Many neoplastic cells were immunopositive with monoclonal antibodies against CD31 (endothelial marker), CD34 and actin, whereas no immunopositivity was seen with a broad specificity anti-keratin antibody (KL1) or with the monoclonal antibody against the melanoma antigen HMB45. Bone spicules with an osteoblastic rim, as well as many osteoclast-type giant cells were seen (Figures 2 and 3). The initial biopsy slides were reviewed in view of the final diagnosis, nevertheless, the lack of malignancy was upheld,

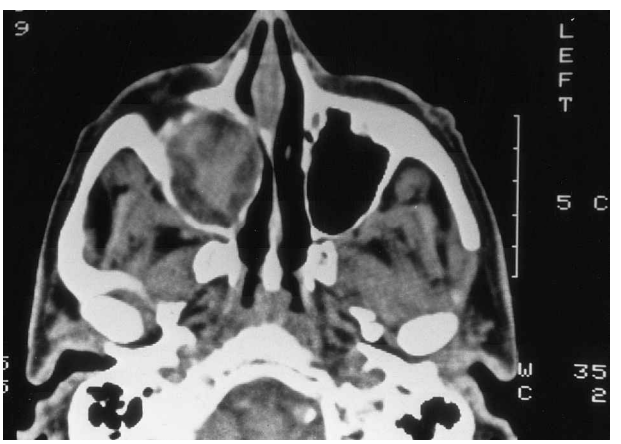

Fig. 1

CT scan showing the lesion almost filling the right maxillary sinus and extending through the bony margins of the medial wall.

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Accepted for publication: 7 January 2000.

https://doi.org/10.1258/0022215001905634 Published online by Cambridge University Press

TABLE I
REPORTED ANGIOSARCOMAS OF MAXILLARY SINUS AND MAXILLA

Source	Sex and age	Site	Treatment	Prognosis
Henny[14]	Male, 3 months	Right maxilla	Wide excision	No recurrence after 27 months
McCarthy and Pack[13]	Male, 13 years	Maxillary sinus, nares, orbit	Radiotherapy	Lost to follow-up
Arena[15]	Female, 5 months	Maxilla	Maxillectomy and radiotherapy	No recurrence after 9 years
Maesaka and Konishi[16]	Male, 27 years	Left maxillary sinus	Maxillectomy, radiotherapy and chemotherapy	Distant metastasis, still under therapy
Goudaert *et al.*[17]	Female, 62 years	Left maxilla	Radiotherapy	Death after 4 months
Bardwill *et al.*[18]	Male, 54 years	Right maxilla, ethmoid sinus	Excision	No recurrence after 5 years
Bomer and Arnold[19]	Female, 47 years	Right maxillary sinus	Maxillectomy and radiotherapy	Died after 3 months
Fu and Perzin[20]	Male, 63 years	Right maxillary sinus	Hemimaxillectomy, sphenoethmoidectomy and radiotherapy	No recurrence after 14 months
McClatchey *et al.*[21]	Female, 26 years	Left maxillary sinus	Radiotherapy	No recurrence after 7 months
Sharma and Nawalkha[22]	Male, 10 years	Left maxillary sinus	Maxillectomy and radiotherapy	Lost to follow-up after 1 month
Bankaci *et al.*[23]	Male, 68 years	Right maxillary sinus	Maxillectomy and radiotherapy	No recurrence after 22 months
Goldenberg[24]	Female, 33 years	Right maxillary sinus	Partial maxillectomy	No recurrence after 14 months
Zakrzewska[9]	Male, 58 years	Right maxillary sinus	Maxillectomy and radiotherapy	Death after 3 months
Carr and Green[10]	Male, 66 years	Right maxillary sinus	Excision	Death after 9 days
Williamson and Ramsden[12]	Male, 48 years	Right maxillary sinus, orbit	Hemimaxillectomy and radiotherapy	Death after 27 months
Lanigan *et al.*[1]	Male, 73 years	Right maxilla, maxillary sinus	Maxillectomy with orbital exenteration and radiotherapy	No recurrence after 33 months
Kurien *et al.*[25]	Male, 38 years	Left maxillary sinus	Medial maxillectomy and palato-alveolar resection	No recurrence after 7 months
Solomons and Stearn[2]	Male, 33 years	Left maxillary sinus	Caldwell-Luc and radiotherapy	No recurrence after 27 months
Kimura *et al.*[26]	Male, 8 years	Nasal cavity	Medial maxillectomy	No recurrence after 2 years
Present case	Male, 72 years	Right maxilla, maxillary sinus	Medial maxillectomy and radiotherapy	No recurrence after 1 year

the specimen consisting mostly of granulation tissue or fragments of respiratory epithelium with no underlying tissue. Since the tumour was incompletely excised and the posterior surgical margins were not free of tumour, the patient was in addition treated with post-operative irradiation. During the two-year follow-up after the operation, no recurrence has been detected.

Discussion

Angiosarcomas are among the most rare sarcomas, deriving from the vascular endothelial cells. The most commonly affected sites are the extremities and the retroperitoneal area.

Less than five per cent of all soft tissue tumours occurring in the head and neck area are sarcomas and only about 10 per cent of them are angiosarcomas. Most of the head and neck angiosarcomas affect the scalp. The earliest report of an upper respiratory tract angiosarcoma is from McCoomb and Martin.[4] This is one of the most unusual tumours for that area[5] The reverse is also true, the nose and paranasal sinuses being one of the rarest locations for angiosarcomas: 13 out of 366 (four per cent) slide files of the Armed Forces Institute of Pathology.[6] For the majority of these tumours, the neoplastic cells are immunopositive for endothelial markers, whereas the cells of the remaining, poorly differentiated tumours are not.[5] Angiosarcomas have also been reported to involve the nasopharynx, parotid gland, lips, cheek, palate, floor of the mouth, gingiva, orbit, temporal bone and larynx.[1] Angiosarcomas of the maxilla and the maxillary sinus are very rare neoplasms. Only 19 cases have been reported in the English literature (Table I) with the youngest patient three months old and the oldest 73 years old.

The incidence of angiosarcomas is generally reported to be almost the same for males and females. Bundens and Brighton[7] have reported that the incidence of this lesion in the bones is almost twice as much as in males as in females. Of the reported cases involving the maxilla and maxillary sinus, 14 cases have been in males and five in females. In contrast, in Zachariades' review[8] of oral angiosarcoma, females were affected almost twice as much as males.

Most patients with angiosarcomas in the head and neck region had signs and symptoms present for only four to eight weeks.[9] Angiosarcomas are rapidly growing neoplasms with an insidious onset and minimal symptoms. In the maxillary antrum these symptoms are related to the invasion by the tumour of the surrounding structures (eye,

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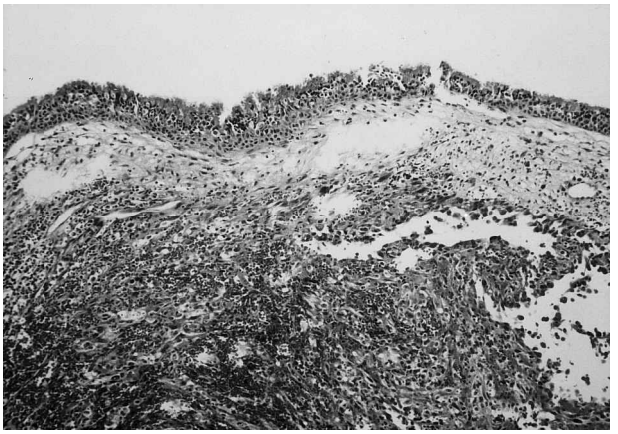

FIG. 2
Tumour fragment covered by respiratory-type epithelium (top). (H & E; ×10).

orbit, alveolus, lateral wall of the nose, anterior wall of the antrum and intra-orbital nerve). Because of the vascularity of the tumour, the most common presenting symptom is epistaxis. Other symptoms include headaches, toothaches, tooth mobility, nasal obstruction, facial asymmetry displacement of the eye, diplopia, anaesthesia or paraesthesia and lymphadenopathy.[1]

The radiographic appearance of angiosarcomas involving the facial bones is generally that of a destructive osteolytic lesion.

The initial diagnosis of angiosarcoma is usually established by a biopsy, although it may be difficult to make a definite diagnosis. Angiosarcoma of bones is histologically similar to lesions of the soft tissues. Angiosarcomas are extremely vascular tumours so that even a diagnostic biopsy, and most certainly a surgical excision, may be an exsanguinating event.[10] Batsakis and Rice[11] divided their cases into two histological grades, low grade and high grade. The diagnostic histological features of the low-grade lesion are of diffuse proliferation of inter-anastomosing atypical capillaries lined by large endothelial cells, with papillary fronds into the lumen and hyperchromatic nuclei. The high-grade lesions are diffusely cellular with areas of undifferentiated cells with pleomorphism and multiple mitoses.

The suggested aetiological factors for the development of angiosarcoma in other parts of the body (e.g. skin, liver and spleen) include exposure to X-rays, thorium and most important vinyl chloride monomer.[12] Reports in the literature point out that angiosarcomas can arise from benign angiomas that have been irradiated.[13] Pregnancy has also been reported to stimulate their growth.[8] McCarthy and Pack[13] hypothesized that angiosarcomas arise from the capillaries of granulation tissue in traumatized areas. Other authors consider a history of trauma to be of doubtful significance.[3]

Angiosarcomas spread by local infiltration and by early haematogenous metastases. Lymphatic spread may also occur.[8,10]

Surgery, radiotherapy and chemotherapy have been used in the treatment of head and neck angiosarcomas. The preferred treatment is wide surgical excision of the tumour followed by irradiation. Radiotherapy may also be helpful pre-operatively to decrease the vascularity of primary bone tumours. Chemotherapy has not been found to be effective in the cure of angiosarcomas.[9]

The prognosis for angiosarcomas is generally poor, although there is significant variation among different anatomical sites and ages, making prediction of the outcome somewhat uncertain. These tumours have a strong

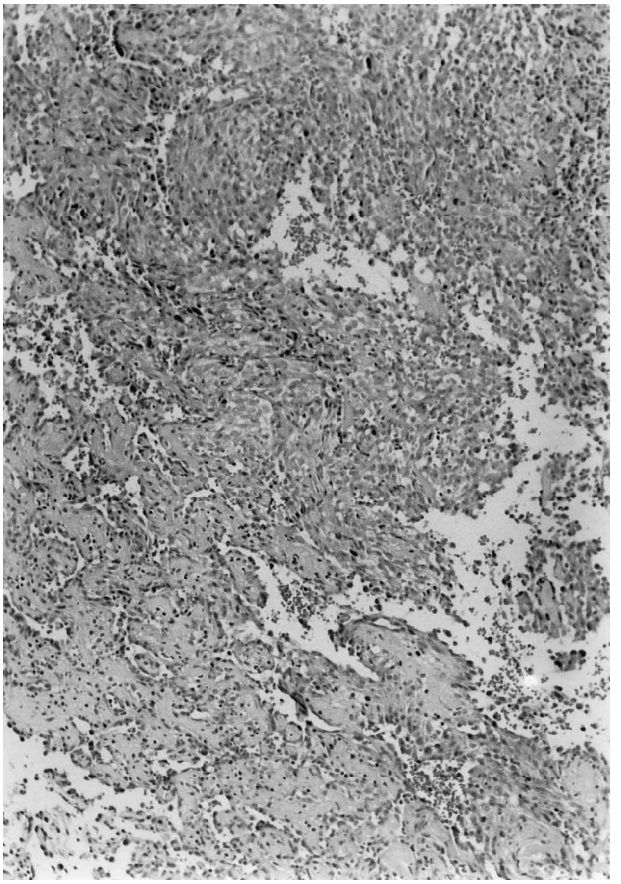

FIG. 3
Closer view of the tumour with both vascular channels lined by neoplastic cells and more compact areas. (H & E; ×10).

tendency to recur locally and to metastasize. Patients with metastases have the poorest prognosis.[8] The main cause of death is pulmonary metastases. McCarthy and Pack (1950)[13] reported a three-year survival rate of 17 per cent and a five-year survival rate of nine per cent.

Because so far only a few cases of angiosarcomas of the maxilla and maxillary sinuses have been reported, it is difficult to draw firm conclusions regarding the efficacy of various treatment regimens on the prognosis of the tumour at this site.

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Mr G. Velegrakis takes responsibility for the integrity of the content of the paper.
Competing interests: None declared.

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