

Angiosarcoma of the nasal cavity

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Abstract

Angiosarcoma of the nasal cavity is extremely rare. We present a case of angiosarcoma of the nasal cavity in an eight-year-old boy. He was treated with medial maxillectomy via lateral rhinotomy. The histological diagnosis was confirmed by immunohistological stain with Factor VIII-like antigen. Magnetic resonance imaging (MRI) was useful in determining the extent of the tumour.

Case report

An eight-year-old boy first noticed left-sided epistaxis in December 1990. He was taken by his father to the ENT department at the Takaoka City Hospital on January 23, 1991. Examination of the left side of his nose showed an haemorrhagic mass, situated in the middle meatus. An intranasal biopsy showed suspicion of angiosarcoma. He was then transferred to our hospital on 12 February 1991. Physical findings were unremarkable with the exception of pallor of the face and skin. Local examination of the left side of the nose showed that the tumour was situated in the middle turbinate and middle meatus (Fig. 1). Laboratory studies revealed severe anaemia (haemoglobin 7.6 g/dl). All other blood tests were within normal limits. X-ray of the chest was normal. On computed tomography (CT), the tumour was not detected, but opacification of the left maxillary antrum, ethmoid cells and frontal sinus were observed. An MRI study was performed to differentiate whether the contents were due to an inflammation or not. The MRI study showed bright signals coming from the fluid-filled sinuses on T₂-weighted images (Fig. 2). The pathological specimen was found to be positively stained with Factor VIII-like antigen immunohistologically. Thus, the tumour was diagnosed as angiosarcoma.

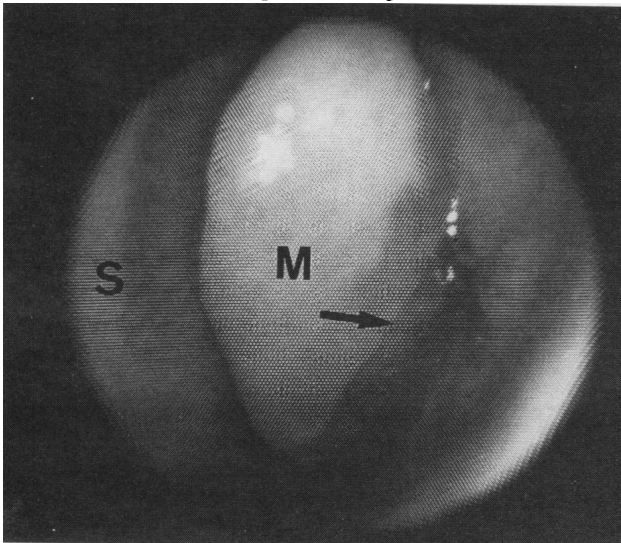


FIG. 1

Tumour of the left middle meatus (arrow). M = middle turbinate, S = nasal septum.

The patient was given a blood transfusion for three days, and the haemoglobin level returned to normal. A medial maxillectomy via a lateral rhinotomy was performed. The lacrimal sac was then divided anteriorly and posteriorly, and sutured to the underlying soft tissue. Bleeding amounted to 210 ml. Definitive histology of the operated specimen revealed complete excision and the tumour was localized in the middle turbinate. Eight months post-operatively there was no evidence of recurrence.

Pathology

Microscopically, the tumour showed both solid and papillary areas. The cells in the solid area were pleomorphic and displayed mitotic activity. An irregular arrangement of often poorly formed vascular channels was present, lined by pleomorphic and hyperchromatic cells (Fig. 3). Factor VIII-like antigen was tested for and found to be strongly positive (Fig. 4).

Discussion

Most head and neck angiosarcoma tend to affect the soft tis-

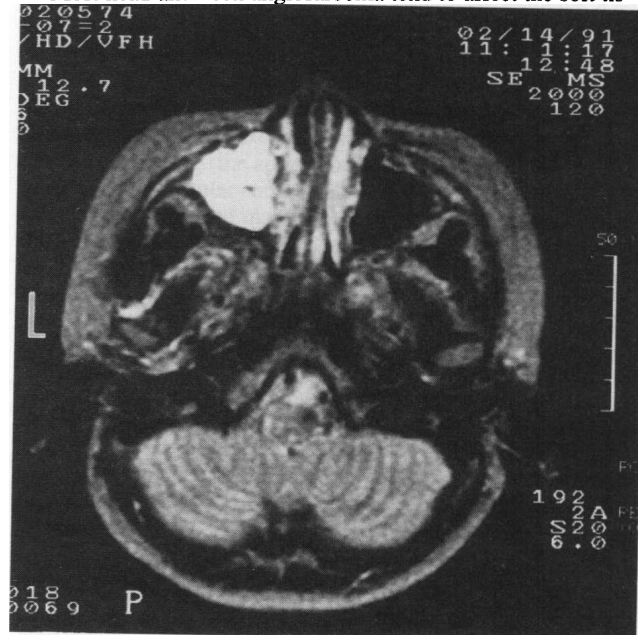


FIG. 2

MRI study shows bright signal coming from the fluid-filled left maxillary sinus on T₂-weighted image.

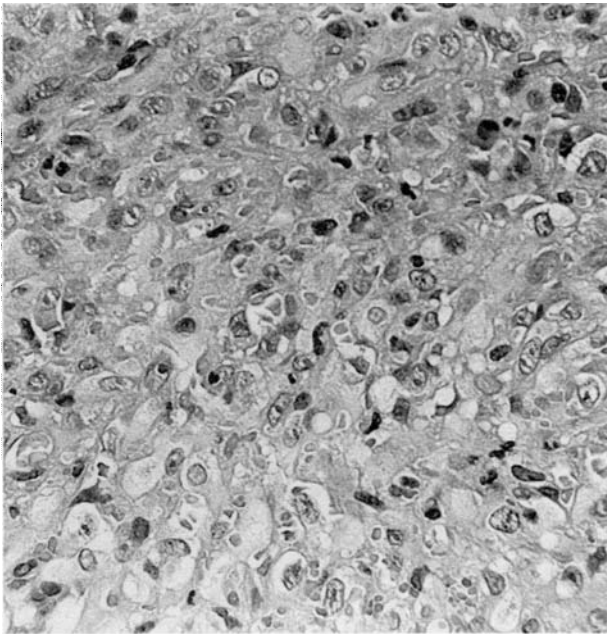


FIG. 3

Tumour shows an irregular arrangement of poorly-formed vascular channels (H-E stain, $\times 400$).

sues, especially the scalp (Enzinger and Weiss, 1988). Angiosarcoma of the nose and maxillary sinus are very rare (Bankaci *et al.*, 1979). Lanigan *et al.*, (1989) reviewed 13 cases of angiosarcoma of the maxilla and maxillary sinus. According to them, the maxillary sinus or maxilla was involved in all cases, but there were no cases where the tumour was localized in the nasal cavity. To the best of our knowledge, the present case is the first in which the tumour was localized in the nasal cavity.

In this case, there were two major problems; first, the initial diagnosis of angiosarcoma is usually made following a biopsy, although it may still be difficult to make a definitive diagnosis. However the availability of immunohistochemistry has made this easier (Du Boulay, 1985). Factor VIII related antigen is a reliable marker for vascular endothelial cells as the cells synthesize Factor VIII rag which is also present in small amounts in the plasma and in megakaryocytes (Mukai *et al.*, 1980). The diagnosis of this patient was confirmed by immunohistochemistry.

Second, it was difficult to determine the extent of the tumour. A CT study showed the contents of the left maxillary sinus, ethmoid cells and frontal sinus. If these contents were considered as those of invasion by the angiosarcoma, a more radical operation should have been undertaken. A MRI study, however, was available for this problem. These contents showed bright signals on T₂-weighted images, indicating the fluid-filled opacification due to the obstruction of the ostio-meatal lesion.

Surgery, radiotherapy and chemotherapy have been used in the treatment of head and neck angiosarcomas. The preferred treatment would appear to be a radical surgical resection of the tumour (Farr *et al.*, 1970; Kurien *et al.*, 1989; Solomons and Stearns, 1990). Although this tumour is radiosensitive, it is rarely curable by radiation alone. We did not include radiotherapy, as the patient was only eight-years-old. Only a few cases

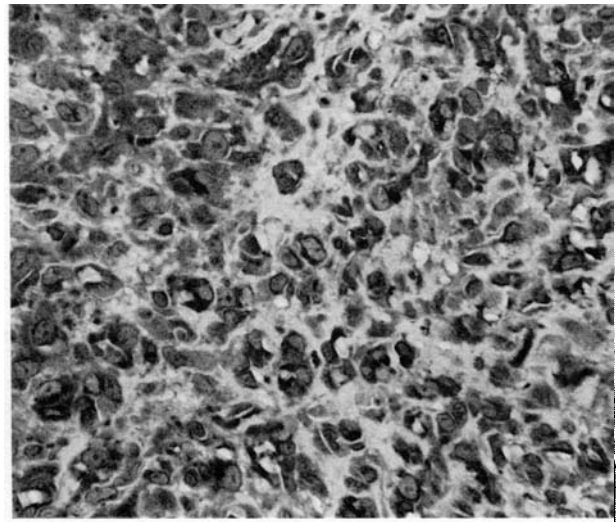


FIG. 4

Stained with Factor VIII-like antigen ($\times 400$).

of angiosarcomas of the nose and maxillary sinuses have been reported, and thus it is difficult to draw conclusions regarding the efficacy of various treatment regimens on the prognosis of the tumour at this site.

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