Mallory (1908) first coined the term hemangioendothelioma to designate neoplasia of vascular endothelium, including both benign and malignant varieties. Weiss and Enzinger (1986) used the term Hemangioendothelioma for the vascular tumours which histologically are intermediate in appearance between a cavernous hemangioma and conventional angiosarcoma and identified three subgroups of Hemangioendotheliomas. These are (1) Epitheloid hemangioendothelioma (2) Spindle cell hemangioendothelioma (3) Malignant endovascular papillary hemangioendothelioma.

This report presents a rare case of spindle cell hemangioendothelioma of the maxillary sinus.

**Case Report**

A 21 year old male consulted an ENT specialist in March 1990 for the complaints of right nasal obstruction, epistaxis and frontal headache for two and a half months. X-ray paranasal sinuses showed haziness of the right maxillary sinus and right nasal cavity. A proof puncture was done following which the patient had severe epistaxis and hence was referred to our hospital. Anterior rhinoscopy revealed a bulge in the right inferior and middle meatus with blood clots in the nasal cavity. Posterior rhinoscopy showed a pinkish mass in the right posterior choana. The nasopharynx and oral cavity were normal. X-ray of the paranasal sinuses revealed complete haziness of the right maxillary sinus and right nasal cavity. CT scan with contrast revealed an enhancing soft tissue mass in the right naso-maxillary region extending to the right ethmoid region (Fig 1 & 2).

The tumour was approached through a Weber-Fergusson incision. A well circumscribed vascular mass (Fig. 3) was found in the right maxillary sinus extending to the right nasal cavity with destruction of the intervening partition wall and thinning of the antero-lateral wall of the right maxillary sinus. An initial impression of a benign tumour was formed due to the tumour being firm and well circumscribed and a partial maxillectomy was therefore performed. The histopathological report showed spindle cell hemangioendothelioma. A one year follow up showed no tumour recurrence.

**Histopathological Findings**

Histopathology revealed a naso-formative circumscribed neoplasm...
Spindle Cell Hemangioendothelioma of the maxillary sinus – Seth et al

Fig. 3. Photograph showing gross specimen

Fig. 4. Photomicrograph of spindle cell Hemangioendothelioma

growing within the submucosa and covered with stretched denuded sinus mucosa. It consisted of narrow vascular channels and cords, and papillae lined by endothelium with wide separation due to diffuse hemorrhage. Scattered between the gaping vessels was a population of spindle cells, some of which lined slit like vascular spaces. These were well differentiated and there was minimal nuclear atypia. An eosinophilic infiltrate was seen in some areas. It was hence diagnosed as a spindle-cell hemangioendothelioma (Fig 4).

Discussion

Spindle cell hemangioendothelioma is a newly recognized form of neoplasm of vascular endothelium first described by Weiss and Enzinger in 1986. It is a low grade angiosarcoma that combines the features of both a cavernous hemangioma and Kaposi’s sarcoma. In a review of literature, thirty cases have been so far reported (Weiss and Enzinger 1989). They occur at any age with male predominance and develop preferentially in the dermis and subcutaneous tissues of the distal extremities. Involvement of the maxillary sinus has not been reported so far. However, Luniqen et al (1989) have reported 13 cases of conventional angiosarcomas of the maxillary sinus.

But because of their different histological features and biological behaviour, spindle cell hemangioendotheliomas are grouped as a separate entity. Clinically, they present with spontaneous bleeding from trivial trauma, and swelling or growth as the principal complaints. Other presentation could be headaches, toothaches, tooth mobility, nasal obstruction, epistaxis or hemorrhagic rhinorhoea, facial asymmetry or paraesthesia and lymphadenopathy.

Grossly, the tumours are small, circumscribed reddish nodules which may be solitary or multifocal. Histologically, the hallmark of these tumours are the peculiar juxtaposition of cavernous blood spaces and a spindle cell stroma. The two components vary quantitatively from lesion to lesion and account for the range of diagnosis (e.g. hemangioma, arterio-venous hemangioma, Kaposi’s sarcoma).

Hence, accurate pathological scrutiny must be done of all the biopsy material to establish the correct diagnosis. Weiss and Enzinger could not predict the biologic behaviour of these tumours. They appear to be borderline with a clinical course between that of hemangioma and that of conventional angiosarcoma. Unfortunately, on histological ground it is neither possible to predict which benign lesions will metastasize nor to predict which one may evolve histologically over a period of time to a malignant lesion. Therefore, a close follow-up is essential to detect and treat the frequent recurrences that are known.

Because of the low grade nature of these tumours, complete and wide local excision without adjuvant radiotherapy or chemotherapy is the preferred mode of treatment as opposed to conventional angiosarcoma. For histologically malignant forms, a radical neck dissection should be considered for metastasis to lymph nodes. Radiotherapy is reserved for multifocal and recurrent disease as sufficient data is not available to judge the radio responsiveness of the tumour as seen in conventional angiosarcoma.

Summary

Spindle cell hemangioendothelioma is a newly recognized vascular neoplasm. It is intermediate between a hemangioma and conventional angiosarcoma, histologically and in biological behaviour, and therefore should be grouped as a separate entity. A high index of suspicion and accurate pathological scrutiny is essential for correct diagnosis. Because of the low grade nature of these tumours, wide