CASE REPORT

Sino-Nasal Hemangiopericytoma

Aniece Chowdhary, Parmod Kalsotra, Des Raj Bhagat, Subash Bhardwaj*, Parvez Katoch

Abstract

Hemangiopericytoma is an uncommon vascular tumor derived from pericytes of Zimmermann, representing only a small portion of head and neck tumors, mostly occurring in the sino-nasal tract. A rare case of sino-nasal hemangiopericytoma in a 52 year female presenting with symptoms of nasal abstruction and epistaxis and its treatment modalities are discussed in the present case report.

Key words

Sino-nasal hemangiopericytoma, Epistaxis.

Introduction

Sino-nasal hemangiopericytoma is an uncommon vascular tumour believed to be derived from the pericytes of Zimmermann, cells that lie external to the reticulum sheath of capillaries with a grossly fleshy or spongy consistency and a thin walled branching ("staghorn") vascular pattern microscopically. Most commonly arises as slowly growing mass in the pelvic retroperitoneum or the lower extremities (especially thigh) of middle aged women, reaching 5-15 cm in maximum diameter.

Overall, 25 per cent of the hemangiopericytomas arise in head and neck. However, they represent only a small portion of head and neck tumours. The nasal cavity and paranasal sinuses are the most common locations in the head and neck; less frequent being the orbit, parotid and neck spaces. The tumour typically presents as a painless mass in all age groups, predominantly in 6th and 7th decade of life with no sex predilection. The etiology of hemangiopericytoma is unknown, although these lesions have been linked to trauma, prolonged steroid use and hormonal disturbances (1).

Many soft tissue tumours focally display "hemangiopericytoma-like" pattern. These include solitary fibrous tumour, synovial sarcoma, infantile myofibromatosis, low-grade endothelial stromal sarcoma, mesenchymal chondrosarcoma, infantile fibrosarcoma and phosphaturic mesenchymal tumour (2). Hemangiopericytoma is divided into two groups *adult :* Occurs in elderly patients and is usually located in deep soft tissue, lower extremities, pelvis and retroperitoneum. *Infantile or congenital hemangiopericytoma :* Usually present at birth or noted soon after. This tumour has a benign course. Sino-nasal hemangiopericytomas usually present with symptoms of nasal obstruction and epistaxis. The diagnosis is made at biopsy and CT scan (1).

We report a case of hemangiopericytoma of the sino-nasal tract in a 52 year old female who presented with symptoms of nasal obstruction and epistaxis.

Case Report

A 52 year old female patient presented in the Department of ENT & Head Neck Surgery, SMGS Hospital, Government Medical College, Jammu with the complaints of right-sided nasal obstruction for the last 1½ years with occasional history of epistaxis. On examination, a bleeding polypoidal mass was seen on anterior rhinoscopy filling the right nostril completely. No mass was seen on posterior rhinoscopy and rest of the

From the Postgraduate Departments of ENT & *Pathology, Govt. Medical College, Jammu (J&K)
Correspondence to : Dr. Des Raj Bhagat, H. No. 35, W. No. 1, Arnia, Teh. Bishnah, Distt. Jammu (J&K)

ENT examination was within normal limits. Biopsy of the mass showed hemangiopericytoma (Fig. 1a & 1b). CT scan of the nose, nasopharynx and paranasal sinuses (Fig. 2) revealed a heterogeneous hyperdense mass filling the right nasal cavity with expansion and evidence of extension into the right maxillary, frontal, ethmoid and sphenoid sinuses. The mass was also extending to right infra-orbital margin. Bone window settings revealed destruction of medial wall of maxillary sinus, orbital floor, lacrimal and ethmoidal bone.

Fig. 1a. Photomicrograph showing a vascular tumor. The cells are plump and spindle shoped and lying among from the endotheium of the blood vessels. (Arrow) : (H&E = 10X)

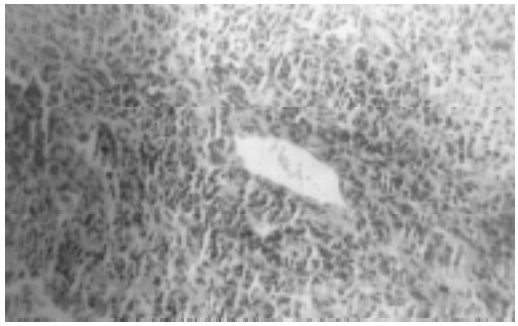

Fig. 1b. Same lesion under high power. (H&E=40X)

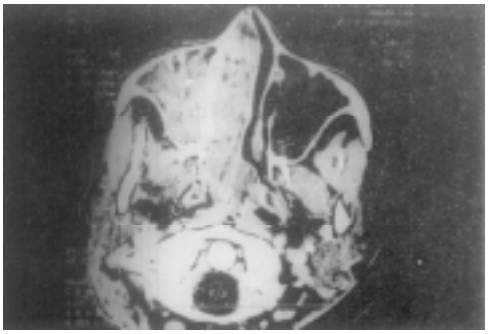

Fig. 2. CECT.

All other investigations were within the normal limits. Surgery was planned as the modality of treatment but the patient did not agree for operation due to her personal beliefs and left the department against the medical advice.

Discussion

Hemangiopericytoma was first described by Stout and Murray in 1942, as a less form of glomus tumour arising from pericytes. Grossly, it is nearly always solitary and solid, with a smooth surface and a colour ranging from greyish white to reddish brown. In about three-fourth of cases, the tumour is well circumscribed or encapsulated with areas of haemorrhage, necrosis and cystic degeneration. The metastasis is seen in about 10-15 per cent of cases, lungs and skeleton being the most common sites of metastasis. The recurrence can occur five or more years after excision (2).

Immunohistochemically, the hemangiopericytoma cells are invariably positive for vimentin but very rarely and only focally for smooth muscle markers such as actin and desmin. A sub-population of tumour cells is also immunoreactive for factor XIIIa and histocompatibility antigen HLA-DR. CD34 usually stains only endothelial cell component, although sometimes there is a diffusion effect in the immediate vicinity. Microscopically, the features noted are cytoplasmic filaments and processes, pinocytotic vesicles, basal lamina and poorly formed intercellular junctions. The aberrations of $12_{q13\text{-}15}$ have been detected in a subgroup of hemangiopericytomas (3).

Sino-nasal hemangiopericytoma is an uncommon upper aerodigestive tract tumour of uncertain cellular differentiation, with majority of them behaving in a benign manner with excellent long-term prognosis (88% - 5 year survival) following surgery alone. These have a characteristic light microscopic appearance with an immunophenotypic profile resembling that of glomus tumours. Many series on hemangiopericytoma in the head and neck literature involve the sino-nasal tract as primary site (4). In the CNS, the hemangiopericytoma is a highly cellular and vascular tumour which is indistinguishable histologically from the similar lesions occuring in the peripheral soft tissues. Aggressive growth, tendency to

recurrence and relatively frequent metastasis are the clinical features of these tumors in CNS. The local recurrence rate of intra-cranial hemangiopericytomas is almost inevitable upto 91 per cent of cases, while extracranial metastasis occur in 68 per cent of cases at 15 years (5).

Sino-nasal hemangiopericytoma have been subtyped as (4) :

(a) *Soft tissue type hemangiopericytoma :* An aggressive tumour, locally destructive and may metastize to regional lymphnodes and lungs. It does not show myoid differentiation. Smooth muscle actin and other myoid markers are negative.

(b) *True hemangiopericytoma :* Has benign behaviour and do not metastize. It has a myogenic appearance and shows convincing pericytic differentiation. The smooth muscle actin is positive. Some of these tumours recur locally.

The differential diagnosis of sinonasal hemangiopericytoma includes - glomus tumour, angiofibroma, fibrohistiocytic tumour, solitary fibrous tumour and lobular capillary hemangioma (5).

The treatment of choice in hemangiopericytomas is wide surgical excision which are known to recur insidiously even years later (57%). Well differentiated tumours rarely metastize, however given their propensity to recur locally, these lesions are considered malignant. Similar to angiosarcomas, these lesions are graded as high or low grade tumours with survival rates improved in the latter group. Radiation therapy is usually reserved for recurrent lesions not amenable to surgical excision and those with a more active histology. These tumours are poorly chemosensitive, however a recent report documented a good response to alpha-interferon in two patients. Stereotactic radiosurgery of hemangiopericytomas can result in increased tumour control and should be considered as a treatment option for patients in whom the diagnosis has been established and in whom residual tumour is demonstrated post-operatively. Close clinical and radiological follow-up evaluation is necessary in these patients because of the high rate of local recurrence and distant metastasis (6). The features indicating poor prognosis include increased cellularity, necrosis, haemorrhage and more than 4 mitotic figures per 10 high power field (2).

References

1. Harrison DFN. Tumors of the nose and sinuses. In : Ballantyne J, Groves J (eds), Scott-Brown's Diseases of Ear, Nose and Throat, Butterworths and Co. Publishers, Toronto : Forth Edition 1979; Vol. 3, Chapter 17 : pp. 364.
2. Stout AP, Murray MR. Hemangiopericytoma : A vascular tumour featuring Zimmermann's pericytes. *Ann. Surg.* 1942; 116 : 23-33.
3. Nielsen GP, Dickersin GR, Provenzal JM, Rosenberg AE. Lipomatous hemangiopericytoma. A histologic, ultractural and immunohistochemical study of a unique variant of hemangiopericytoma. *Am J Surg Pathol* 1995; 19 : 748-56.
4. Thompson LD, Miettinen M, Wenig BM. Sino-nasal type hemangiopericytoma : A clinicopathological and immunophenotypic analysis of 104 cases showing perivascular myoid differentiation. *Am J Surg Pathol* 2003 ; 27 (6) : 737-49.
5. Tzuu-Yuan H. Intracranial hemangiopericytoma : Diagnosis, treatment and outcome. *Chinese Med J* 2002 ; 65 (7) : 305-06.
6. Chang SD, Sakamoto GT. The role of radiosurgery for hemangiopericytomas. *Neurosurg Focus* 2003 ; 14 (5) : 14-15.