Sino-Nasal Haemangiopericytoma

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Abstract

Haemangiopericytoma is a mysterious and rare vascular tumour derived from Pericytes. Benign or malignant nature of tumour is to be determined clinically as histological characteristics are inconsistent with tumour behaviour. HPC most commonly occur in skeletal muscles where it behaves aggressively. Sino-nasal presentation is rare and indolent, may be because of early presentation and lesser tendency of invasion. Thus aggressive tumour resection is avoided and de-bulking of tumour with long term follow up is recommended.

A case of sino-nasal haemangiopericytoma with intracranial extension is presented here.

Introduction

Haemangiopericytoma is uncommon tumour formed by proliferation of pericytes of “Zimmerman”. Term was first used by Stout and Murray in 1942. Pericytes are immature smooth muscle cells derived from mesenchyme and are found surrounding capillaries and post capillary venules, Enzinger with spongy consistency and a thin walled branching vascular pattern (staghorn appearance).

Malignant behaviour of tumour is related to location and anatomic dimension of tumour, in the para-nasal sinuses and nasal cavity HPC have relatively benign course.

The tumour can occur in person of any age and sex, though mostly affecting people older than 20 years of age. HPC can arise in virtually any part of body, most common in extremities especially lower limbs and retroperitoneum. Head and neck involvement is 15-30%, which is only a small per centage of all head and neck tumours, with predilection for soft tissues of scalp, face, neck, parotid gland and orbit. Fifty per cent of those occurring in head and neck involve nasal cavity and paranasal sinuses.

Diagnosis of HPC is not possible clinically and histopathology shows variable pictures, careful histological examination and reticulin staining is required for proper diagnosis.

Optimal treatment of HPC is wide excision with free margins which is difficult for sino-nasal HPC. More conservative approach with debulking of tumour followed by serial endoscopies and imaging studies is suggested for Sino-nasal HPC.

Case Report

A 42 year old female presented to our institute in June 1999 with complaints of headache, right nasal block, muco-purulent nasal discharge and bulging of right lateral wall of nose.

Anterior rhinoscopy showed reddish, firm, polypoidal mass with irregular surface and bleeding on probing. On posterior rhinoscopy mass was extending to nasopharynx (Fig. 1).

CT scan with contrast revealed heterogeneous mass in right nasal cavity with expansion and extension into the right maxillary sinus and ethmoid sinuses, mass was obscuring anatomical landmarks with destruction of bony walls. Tumour showed extension to intracranial space (Figs. 2 and 3).

A biopsy of mass was done, following which patient showed spontaneous expulsion of pieces from mouth in the ward.

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Histopathological study showed pseudostratified squamous epithelium and submucosal tissue consist of spindle cells arranged in interlacing bundles. Hyalinized areas of innumerable proliferation of capillaries and dilated blood spaces were seen with cytological features of malignancy (Fig. 4).

Considering patients general condition and intracranial extension surgery was deferred and
The patient was referred to the higher institute. Unfortunately, we could not follow up with the patient afterwards.

**Discussion**

Haemangio-pericytoma represents 5% of all soft tissue tumours and 1% of all vascular tumours. It is aggressive at other sites but sino nasal HPC is a low grade tumour and has a typical benign course, low metastasis and lesser mortality. Compagno and Hyams confirmed this fact in a study of 23 cases and reported it as a distinct HPC-like entity. But subsequent studies with electron-microscopy and immunohistochemical techniques and review by Hughes and Bard confirmed it to be true low grade HPC. Recurrence rate varies from 8-50% in literature. Tumour involving cribriform plate tends to be more destructive and metastasize. Five year survival rate is 88-92%

Under light microscopy HPC is characterized by vascular channels ranging from compressed structures to capillaries and sinusoids. Pericytes proliferate in varying degrees of differentiation accounting for variation in histological appearance. Thus differential diagnosis should include glomus tumour, schwannoma, angiosarcoma, haemangioendothelioma, and fibrous histiocytoma. Electron microscopy and immunohistochemical staining facilitate diagnosis.

Treatment recommended for haemangio-pericytoma is wide local excision, endoscopically or via lateral rhinotomy. Few reports of chemotherapy and radiotherapy trials showed limited success.

Because of indolent and non-destructive nature of tumour, a conservative approach with debulking of tumour to relieve obstruction and careful follow up with endoscopic examination and imaging studies is recommended. More aggressive resection is reserved for local invasion and extensive tumour, a view supported by recent studies.

**References**