A Rare Presentation of Sinonasal Hemangiopericytoma-like Tumour in Frontoethmoidal Sinus: A Diagnostic Challenge

ABSTRACT

Sinosonal hemangiopericytoma-like tumour (SHPCL) are rare vascular neoplasms derived from Zimmerman’s capillary pericytes. They originate in a paranasal sinus and extend into the nasal cavity secondarily. Hemangiopericytomas of soft tissue usually occur in the retroperitoneum or the thigh and are an uncommon finding in the nasal and paranasal sinuses. They occur most commonly in adults in the sixth and seventh decades of life and clinically mimic allergic polyps. These patients most commonly present with symptoms of epistaxis and nasal obstruction. Microscopically, these tumours demonstrate a vascular architecture, are composed predominantly of spindle cells, and lack nuclear or cytoplasmic pleomorphism, mitotic activity, haemorrhage or necrosis. These criteria include the presence or absence of mitotic figures, necrosis, anaplasia, and haemorrhage. The present case with early onset in fourth decade with predominantly orbital symptoms, minimal findings on nasal endoscopy and biopsy gives a diagnostic challenge. Treated with complete surgical excision and diagnosis was confirmed by immunohistochemistry reports as a rare SHPCL. Being a locally invasive tumour with very less propensity for metastasis or recurrence, two year of recurrence free endoscopic follow up is sufficiently justified it to be cured.

KEYWORDS sinusonal hemangiopericytoma-like tumour, hemangiopericytomas, glomangiopericytoma

INTRODUCTION

Sinosonal hemangiopericytoma-like tumour (SHPCL) is a rare vascular neoplasm of the nasal cavity and paranasal sinuses characterised by a pattern of prominent perivascular growth. The incidence is less than 0.5% of all sinosonal neoplasia of which ethmoid and sphenoid sinuses are the most commonly involved ones. Synonyms include sinusonal-type hemangiopericytoma (HPC) and hemangiopericytoma of sinonasal origin. The HPCs are derived from Zimmerman’s capillary pericytes, which surround all capillaries, and which account for 1% of all vascular tumours. Initially described by Stout and Murray, they are found most frequently in retroperitoneum/pelvis and lower extremities and are known to have malignant behaviour. The etiology remains unknown; though past trauma, hypertension, pregnancy, and use of corticosteroids are considered predisposing factors. Due to variation in histological and clinical features of sinusonal type HPCs, they are also referred to as glomangiopericytoma, due to its similarity to glomus tumours. Its effective management requires wide surgical excision with clear resection of margins, as these tumours are relatively radioresistant. Recent advances in endoscopic surgery have led to the development of techniques that nowadays permits successful endoscopic resection as treatment of choice.

The present case with early onset of disease and without any significant sinonasal symptoms is characterised as a limited disease on radiological evaluation. Its minimal findings at endoscopy with inconclusive biopsy give a diagnostic challenge to the surgeon till the final diagnosis was provided by immunohistochemistry reports.
CASE REPORT

A 37-year-old female patient attended the OPD of ENT department with complaints of protrusion of left eye since 1 year, insidious onset, gradually progressive; associated with progressive diplopia since 6 months. There is occasional history of epistaxis since last 2 months. No associated history of pain, nasal obstruction, rhinorrhea, sneezing episodes, headaches or excessive lacrimation were there with non-contributory past medical or surgical history. On examination, left eye shows proptosis with inferolateral displacement of eyeball (Fig. 1), with fullness between superior rim of the orbit, eye movement was restricted in superior direction; visual acuity was normal, no significant findings found on clinical evaluation of nose, ear and throat. No neck swelling or enlarged node palpable. Provisional clinical diagnosis of frontoethmoid mucocele was made. A friable reddish tissue filling the left frontoethmoid region of nasal cavity was observed on endoscopic examination and biopsy was taken and sent for histopathological examination. The histopathological report comes to be inconclusive.

Radiological evaluation was done. The plain radiograph shows diffuse opacity in left frontal sinuses and the adjacent orbital area. Non-enhanced computed tomogram (NECT) in axial view image shows a homogeneous hypo-attenuating mass in the left ethmoidal sinuses extending to anterior nasal cavity. Its coronal view shows a homogeneous iso-attenuating mass in left frontal sinus (Fig. 2). The radiological diagnosis of benign/cystic lesion of frontal sinus extending to adjacent ethmoidal sinuses was made.

External left frontoethmoidectomy (Fig. 3) was done through Lynch-Howarth incision. Frontal sinus exposed by drilling, the growth was followed inferiorly, superorly and laterally, extending to the left infratemporal fossa, inner table of the skull base was found eroded, underlying dura was found exposed. The growth was followed up to right frontal sinus and removed en block. Wound sutured and a corrugated rubber drain was kept for 48 hours. The amount of bleeding was about 50 ml (measured in the suction pump) at the end of complete removal. No complication or CSF leak was observed at the end of the operation as well as in the post-operative period. Intranasal tamponing with antibiotic ointment was applied; removed on second post-operative day. Excised specimen was sent for histopathological examination. No recurrence has been observed on her 2-year follow-up by nasal endoscopy.

Staining with haematoxylin and eosin (H&E) the specimen showed a cellular neoplasm covered with respiratory epithelium. The tumour cells were oval to spindle with short spindle nucleus, inconspicuous nucleoli and eosinophilic cytoplasm. Significant pleomorphism, atypia and mitoses were not seen. The cells were arranged around ramifying thin-walled vascular channels with staghorn-like appearance. An immuno-histochemical study was performed using the Dako Envision method. The tumour cells were strongly positive for vimentin, α-smooth muscle actin (Fig. 4), and...
In World Health Organization classification (2005) of head and neck tumours, it was recommended that sinonasal hemangiopericytoma like tumour (SHPCL) be referred to as glomangiopericytoma (GPC), in light of the similarity with glomus tumour. SHPCL, a rare mesenchymal tumour arises almost exclusively from the nasal cavity or paranasal sinuses and is characterised by a pattern of prominent perivascular growth. Out of all sinonasal neoplasia, SHPCL comprises less than 0.5%. The peak incidence is during the sixth or seventh decade with a slight female predominance.

Epistaxis and/or nasal obstruction are the most common symptoms. Although past trauma, hypertension, pregnancy, and steroids intake are considered as predisposing factors; even then etiology remains unclear.

The diagnosis depends primarily on thorough endoscopic evaluation and secondly on imaging techniques. Radiological examinations often reveal opacification caused by polypoid mass, rarely with any bone invasion. The typical findings on computed tomogram (CT) can include a soft tissue mass with enhancement following the administration of intravenous contrast, and gadolinium contrast in case of MRI scanning.

In light of above in the present case there was a pattern of prominent perivascular growth. The histological grading of HPC has been addressed as benign, borderline, low grade malignant, overtly malignant by several authors.

The histological features of typical HPC of soft tissue as compared to SHPCL consists of uniform, spindle-shaped cells with indistinct cytoplasm and large nuclei, distributed around vascular channels, typically exhibiting a stag horn branching pattern. The vascular channels are frequently more readily defined following reticulin staining. Though, in its initial description, the distinctive histological features in HPCs have been established to be non-specific and found in up to 15% of unrelated soft tissue neoplasms. The histologic grading of HPC has been addressed as benign, borderline, low grade malignant, overtly malignant by several authors. On immunohistochemistry, the HPC cells were invariably positive for vimentin, SMA but very infrequently and only focally for smooth muscle markers such as desmin. A few tumour cells were also immunoreactive for factor XIIIa and histocompatibility antigen HLA-DR. CD34 usually stains only endothelial cell component, though sometimes there is a diffusion effect in the immediate vicinity. The SHPCL tumour cells composed of closely packed exhibiting storiform, whorled or palisaded pattern, interspersed with numerous thin-walled, branching staghorn vessels. Their neoplastic cells show uniform round to spindle-shaped nuclei. Immunohistochemical study of SHPCL tumour shows that the tumour cells are strongly positive to vimentin, α-smooth muscle actin and muscle specific actin and negative to CD34.

Other differential diagnosis comprised solitary fibrous tumour, glomus tumour, synovial sarcoma and myopericytoma. Solitary fibrous tumour is rich in collagen; tumour cells are positive to bcl-2 and negative to CD34 and actin. The tumour cells of myopericytoma have a myxoidstroma with a solid growth pattern. It is distinguished by its morphologic character as concentric perivascular proliferation of spindle cells. Glomus tumour is different morphologically, as it does not have staghorn-like vessel proliferation and it is frequently localised in distal extremities. Synovial sarcoma is a biphasic tumour as it has both epithelial and mesenchymal components.

The treatment is a complete surgical resection. Because the tumour is highly vascular, several authors advocate preoperative embolisation. The practice of endoscopic resection techniques for SHPCL has been increased because of its slow expanse growth which produces smooth instead of infiltrative tumour borders. Endoscope allows a magnified view and therefore correct assessment of the site of origin and its relation to surrounding structures can be assessed. Furthermore, it preserves the nasal physiologic function and avoids a large external incision. Factors that are not favourable include septal deviation, tumours which are large or highly vascular, intracranial extension, orbit or the pterygopalatine fossa involvement. Thus, patients have to be strictly assessed and selected.

Although it is considered as a borderline low malignancy tumour by WHO classification, a minority can recur or can be fatal. It is understood that recurrence might be a consequence of incomplete initial excision. The reported recurrence rate of SHPCL is relatively varied, ranging from 7% to 20%, with an average time of recurrence of 6–7 years. While an incomplete primary excision has been identified as the prime factor in recurrent disease, severe nuclear pleomorphism, osseous invasion, large tumour size (more than 5 cm) and a high mitotic to proliferation rate also appear to significantly increase the risk of recurrence. In the current case, the tumour was completely resected, so we consider that the patient is now completely cured and the potential for recurrence is low; however long-term follow-up with systemic examination is needed.
REFERENCES