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CASE REPORT

Sinonasal Hemangiopericytoma of Inferior Turbinate presenting as Chronic Dacryocystitis: A Rare Presentation

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ABSTRACT

Sinonasal hemangiopericytoma are unusual tumors that develop from pericytes. We present a unique case of sinonasal hemangiopericytoma of inferior turbinate which presented with chronic dacryocystitis. Patient usually present with nasal obstruction, epistaxis, mass, headache, pain, discharge. But, chronic dacryocystitis is very unusual presentation.

Keywords: Hemangiopericytoma, Inferior turbinate, Dacryocystitis.

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INTRODUCTION

Sinonasal hemangiopericytoma (HPC) are rare neoplasms that develop from pericytes within the outer capillary wall. They account for less than 5% of all sarcomas. Within the head and neck, 20% of all hemangiopericytomas develop in the nasal cavity or sinuses.[1] They are characterized as benign or malignant, round to spindle cell tumors with numerous 'staghorn' branching vascular channels.[2] The majority of sinonasal-type hemangiopericytomas behave in a benign manner with excellent long-term prognosis following surgery alone. We are presenting a case of sinonasal HPC with rare presentation as chronic dacryocystitis.

CASE REPORT

A 7-year-old female patient presented to the outpatient clinic of Department of Otorhinolaryngology, Sardar Patel Medical College, Bikaner, with unilateral watering from left eye since 3 to 4 years. She was diagnosed as left chronic dacryocystitis. Anterior rhinoscopy showed mucoid secretions and congested mucosa and deviated nasal septum toward left. On pressing over sac, there was regurgitation of mucopurulent discharge from both puncta. Patient was taking treatment from ophthalmologist since 3 to 4 years and even after multiple session of syringing by ophthalmologist, patient showed little improvement. There was no history of nasal obstruction, epistaxis, rhinorrhea.

Diagnostic nasal endoscopy showed a well localized, small, globular mass arising from middle one-third of inferior turbinate in inferior meatus. CECT of nose and paranasal sinus showed a small soft tissue density causing obliteration of left inferior meatus and nasolacrimal duct (Figs 1A and B).

Patient was posted for endonasal surgical excision biopsy of mass under GA. Biopsy report showed sinonasal hemangiopericytoma. After excision of mass, patient showed excellent improvement in chronic dacryocystitis.

This patient was kept on regular follow-up for 1 year and nasal endoscopy was performed repeatedly. There was no recurrence and no further intervention was required.

DISCUSSION

Hemangiopericytoma is an unusual vascular tumor derived from pericytes. Compagno et al[3] described this entity as 'hemangiopericytoma like tumor' as early as 1976, and Fletcher[4] recently pointed out that sinonasal HPCs are tumors with true pericytic myloid differentiation distinct from soft tissue hemangiopericytomas. Macroscopically tumors are beefy red to grayish pink, soft, edematous, fleshy to friable masses, often demonstrating hemorrhage. Microscopically, HP is characterized by a proliferation of spindle-shaped to ovoid cells between vascular channels. Sinonasal hemangiopericytoma are basically of three types. These histological subtypes are soft tissue type HPC, true HPC and sinonasal glomus tumor. Soft tissue type HPC tumors are frequently aggressive while true HPC variant have far better prognosis and recurrence is rare. Care must be taken with differential diagnosis between sinonasal HP and other soft tissue tumors, such as glomus tumors or vascular leiomyoma.[5] With regard to histopathologic staging, HPC is generally divided into three groups: low, intermediate and high grade. Staging features include tumor cellularity, mitotic figures, nuclear atypia, and the presence of hemorrhage and necrosis.

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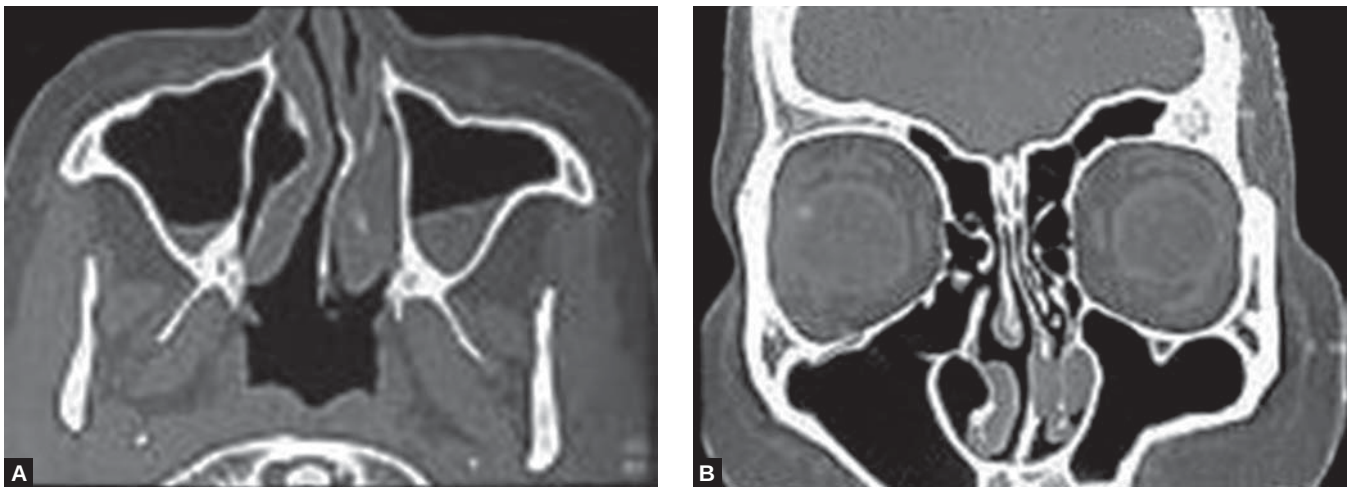

Figs 1A and B: Computed tomography scan (axial and coronal) showing soft tissue density in left inferior meatus

Thompson[7] et al in their study of 104 cases found that the tumors most frequently arose in the nasal cavity alone (n = 47) paranasal sinuses in a fair number of cases (n = 26). Specific anatomic locations were described, but because this occurred in a limited number of cases (turbinate, n = 10; septum, n = 8), the ethmoid sinus (n = 7) or maxillary sinus (n = 5). In our case tumor was arising from middle one, third of inferior turbinate.

Electron microscopy (EM) and immunohistochemical staining facilitate the histologic diagnosis of HPC as the pericytes in these tumors usually retain features sufficient to make them recognizable. Immunohistochemically, cases of sinonasal HP express mostly vimentin and factor XIIIa.[6] In comparison with other localizations, sinonasal HP is usually of low malignant potential.[7]

The majority of patients present clinically with symptoms of nasal obstruction, sometimes with epistaxis. A wide range of other nonspecific findings were identified, including a mass, polyps, difficulty breathing, sinusitis, headaches, congestion, pain, discharge and changes in smell. But, in our case, patient came with history of chronic dacryocystitis which is very unusual presentation of sinonasal HPC.

Computed tomography, and Magnetic resonance imaging (MRI) scans are helpful in determining anatomic location, extension of tumor and underlying bone destruction.

Treatment recommendations for sinonasal HPC centred around wide local excision, the specifics of which are dependent on tumor size and location. In our case, we used transnasal endoscopic approach used due to its limited extension. Recurrence rates for sinonasal HPC vary in the literature from 7 to 50%. High recurrence rates are usually correlated with incomplete tumor removal. Adjuvant radiotherapy is also recommended in highly aggressive disease.

CONCLUSION

Sinonasal HPC are usually benign. Transnasal endoscopic excision is sufficient if disease have limited extension. But, a long-term follow-up is mandatory. Every patient of chronic dacryocystitis should undergo detailed nasal examination to rule out any nasal pathology which could be a cause of nasolacrimal duct (NLD) obstruction. And, it can be relieved by treating the underlying pathology without the need of surgery like DCR.

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