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CASE REPORT OF TWO RARE TUMORS IN THE SPHENOID SINUS

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ABSTRACT

Sphenoid sinus lesions usually have a late clinical presentation due to the deep seated location of the sinus within the body of the sphenoid bone. Hence mostly there is invasion of the adjacent structures at the time of diagnosis. Here we report two rare tumors arising from sphenoid sinus with invasion of the surrounding structures.

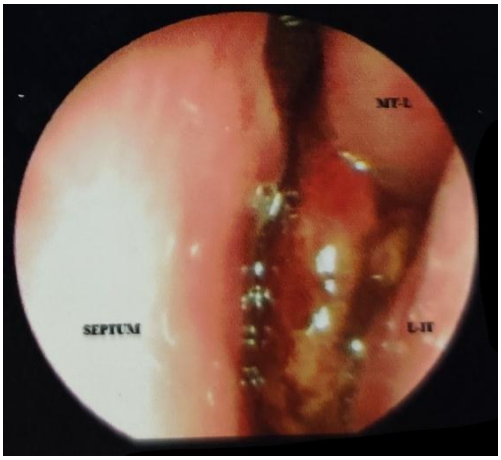
KEYWORDS: Sphenoid sinus, juvenile nasopharyngeal angiofibroma, plasmablastic lymphoma.

INTRODUCTION

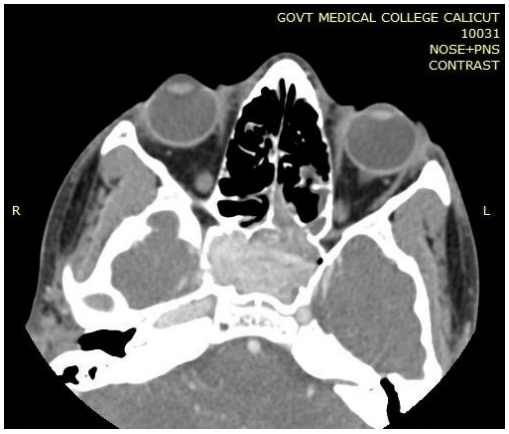
Isolated sphenoid sinus lesions are uncommon, accounting for 1%–2.7% of all paranasal sinuses lesions.[1] They usually present with nonspecific symptoms and are inaccessible to physical examination. The clinical presentation involves vague headaches and may be associated with purulent rhinorrhea, retropharyngeal drip, nasal obstruction, abnormal vision, and nerve deficits.[2] The close anatomical relationship of sphenoid sinus with brain, meninges, cavernous sinus , cavernous segment of internal carotid artery , the lower cranial nerves and other paranasal sinuses causes early spread of the disease process involving the sphenoid sinus. CT and MRI serve as the important investigation modalities for the imaging of sphenoid sinus which play complementary roles. Extent of the disease can be assessed in both CT and MRI, however MRI is better for the assesement of intracranial and intraorbital extensions as well as probable tissue characterisation of the lesion. Conventional radiography has its limitations in diagnosing sphenoid sinus pathologies.

CASE NO: 1

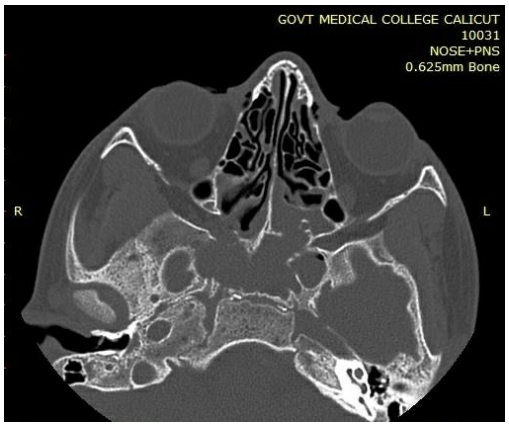
A 13 year old male presented with history of headache for a duration of 2 years and epistaxis for 5 months, with anterior rhinoscopy showing no significant abnormality. Nasal endoscopy revealed a fleshy mass/blood clot arising from left sphenoethmoidal recess extending to the nasal cavity [Fig. 1]. CECT sections showed an intensely enhancing soft tissue density lesion in the sphenoid sinus with extension into the posterior ethmoid sinus [Fig. 2] causing erosion of the walls of sphenoid sinus [Fig. 3]. There was no extension of the lesion into sphenopalatine foramen [Fig. 4]. The diagnosis of angiofibroma arising from the sphenoid sinus was made. The patient underwent transnasal endoscopic resection of the tumor after preoperative embolisation of the feeding vessels. Histopathologic examination revealed mixture of stellate and staghorn blood vessels with irregular fibrous stroma with stromal cells showing small vesicular to large pyknotic nuclei. Large and small vessels were noted in the tumor with smaller vessels showing plump endothelial cells. All these were confirmative of juvenile nasopharyngeal angiofibroma.



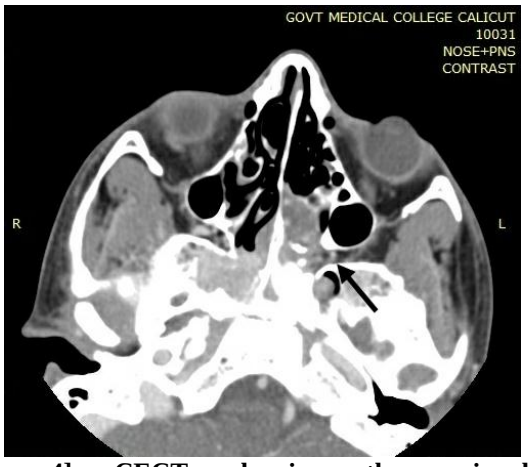
[Fig. 1] Nasal endoscopy showing the fleshy mass in the left sphenoethmoidal recess.



[Fig. 2] CECT section showing intensely enhancing lesion within the sphenoid sinus extending to the posterior ethmoid sinus.



[Fig. 3] CT bone window showing erosion of the walls of sphenoid sinus.

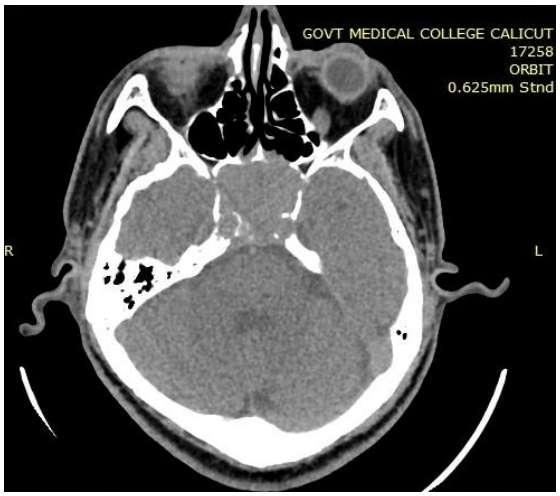


[Fig. 4] CECT showing the uninvolved sphenopalatine foramen with no widening of the pterygopalatine fossa

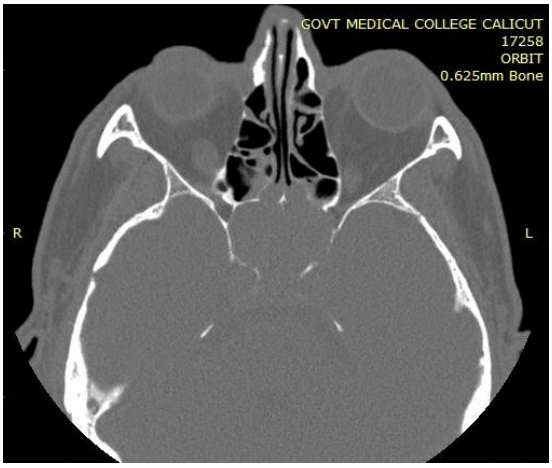
CASE NO: 2

A 64 year old male presented with history of epistaxis and headache for one month duration and drooping of left eyelid for a duration of 2 weeks. Clinical examination revealed left 3rd cranial nerve palsy. CECT sections showed an intensely enhancing hyperdense lesion of average density 50 HU in plain CT [Fig. 5] within the sphenoid sinus with erosion of the bony walls of sphenoid sinus [Fig. 6], extending inferiorly into the nasopharynx, superiorly into the sella and laterally into bilateral cavernous sinuses [Fig. 7] displacing cavernous segment of internal carotid artery. Differential diagnosis of lymphoma, myeloma and chordoma was made.

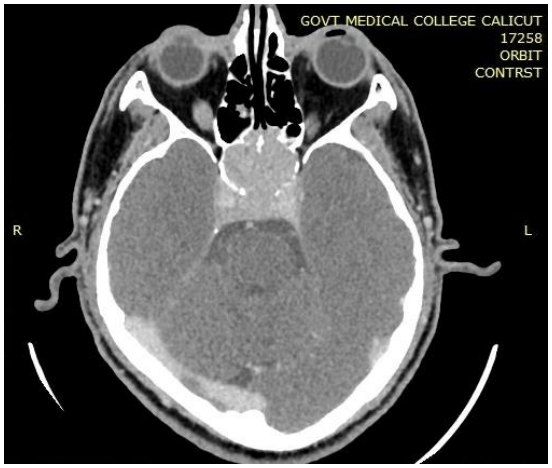
Multiple myeloma was ruled out by laboratory investigations and bone marrow biopsy .For further anatomical delineation and tissue characterisation a contrast MRI study was done, which showed a lobulated lesion involving the sphenoid sinus appearing isointense in T1WI, iso to hyperintense in T2WI with intense post contrast enhancement [Fig. 8 to 11]. Diffusion weighted sequence showed diffusion restriction [Fig. 12 & 13]. No blooming was noted in gradient echo sequence. The differential diagnosis of sinonasal malignancy involving sphenoid sinus and lymphoma was made, following which the patient underwent diagnostic nasal endoscopy and biopsy. The histopathologic examination revealed tumor cells positive for CD138, CD20 and LMP-1(EBV) and CD3 showed reactive pattern, which was suggestive of plasmablastic lymphoma. For the staging of the disease, patient underwent whole body PET- CT study which showed metabolically active lymphomatous lesions involving pelvis, ribs, mandible, vertebrae, sternum, clavicles, lower lobe of left lung and both infradiaphragmatic and supradiaphragmatic lymph nodes.



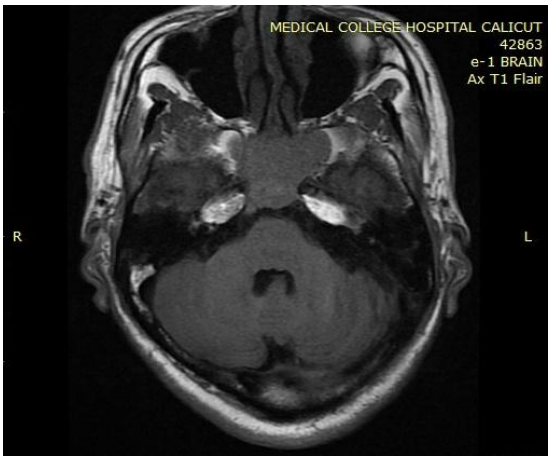
[Fig. 5] Plain CT section showing hyperdense lesion within the sphenoid sinus.



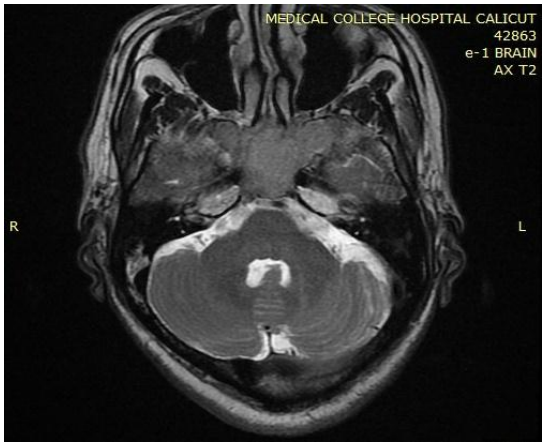
[Fig. 6] Showing the erosion of the walls of the sphenoid sinus.



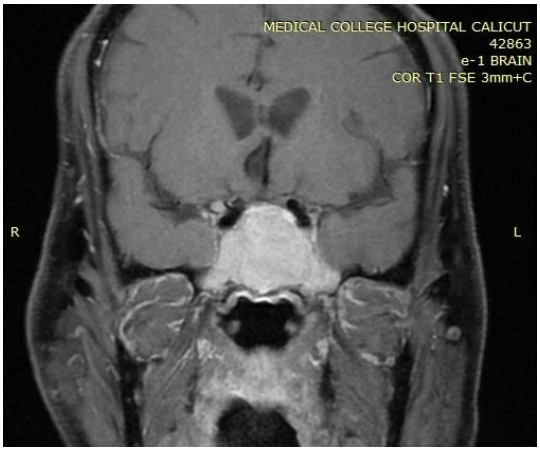
[Fig. 7] CECT section showing intensely enhancing lesion within the sphenoid sinus extending to bilateral cavernous sinuses displacing the cavernous segment of internal carotid artery.



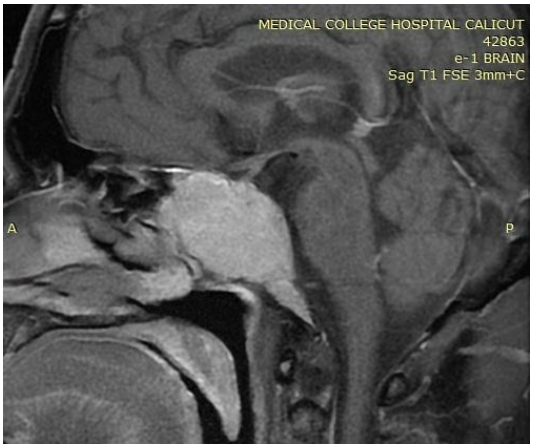
[Fig. 8] Axial T1W image showing the lesion.



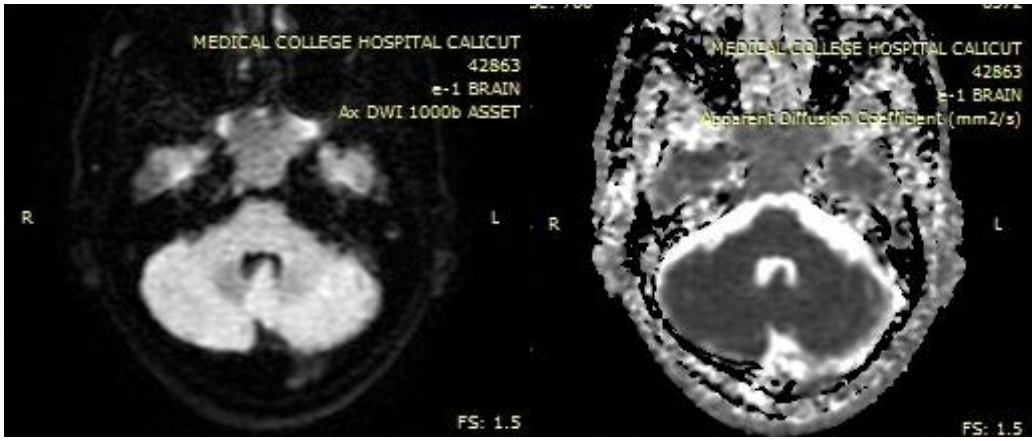
[Fig. 9] Axial T2W image showing the lesion.



[Fig. 10] coronal contrast sections showing the lesion extending to bilateral cavernous sinuses.



[Fig. 11] sagittal contrast sections showing the lesion extending superiorly into the sella and inferiorly upto the nasopharynx.



[Fig. 12& 13] Showing areas of diffusion restriction within the lesion with corresponding fall in ADC.

DISCUSSION

This case study is unique as both the cases reported here, that is a juvenile nasopharyngeal angiofibroma and plasmablastic lymphoma are rare lesions that are reported in the sphenoid sinus to the best of our knowledge.

Juvenile nasopharyngeal angiofibromas are benign tumors of vascular origin with a locally aggressive behaviour. They are exclusively found in adolescent males. They usually present with epistaxis and obstructive symptoms. Usually they arise in the region of the sphenopalatine foramen and tend to widen the pterygopalatine fossa, extend into the nasal cavity, orbit, paranasal sinuses, infratemporal fossa and intracranially. Extensive bony destruction is not a usual feature, rather they cause bone resorption / remodelling. Angiography may be done if planning for a pre-operative embolisation to identify the feeding vessels. The major differential diagnosis is angiomatous polyp.

Sinonasal malignancy is rare and a multitude of differentials may be considered. Approximately 80% are squamous cell carcinoma, the most of the remainder is contributed by adenocarcinoma, adenoid cystic carcinoma and lymphoma and the rarer ones include neuroblastoma, melanoma and sarcoma . Of these only less than 5% arise in the sphenoid sinus.[3] Clinically they present with facial pain, paresthesia, recurrent sinusitis, nasal discharge, eyelid swelling, cranial nerve palsies and proptosis.

Plain radiographs no longer play a role in diagnosing sphenoid sinus lesions. CT is done to assess the bony involvement and MRI for tissue characterisation and assesement intraorbital and intracranial extension. MRI may also be done to distinguish between retained secretions and mucosal thickening.

CONCLUSION

Majority of the sphenoid sinus lesions present late and a detailed pre-operative work up including contrast CT and MRI with histopathological examination is often essential for the confirmation of the diagnosis.

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