A RARE CASE OF RECURRENT NASAL SCHWANOMA

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ABSTRACT
Schwanoma are neurogenic tumors which are rarely found in nose and paranasal sinus. A 65 year old male presented with chronic rhinosinusitis affecting his right nose. He has transnasal surgery for the same complaint. Histopathological report suggestive of schwannoma which was positive for SOX-10 and S-100 on immunohistochemistry. The lesion was removed by endoscopic sinus surgery with debridment. The patient made good post-operative recovery and remained disease free at six months follow-up.

KEYWORDS
Recurrent nasal schwanna, S-100, SOX–10, benign tumor.

Introduction
Schwanoma are benign tumours originating from peripheral nerve sheaths. Previous reports indicate that 25–50% of schwanomas occur in the head and neck region, but tumours originating from the nasal cavity or paranasal sinuses are rare, with a reported rate of approximately 4% [1,2].

We present a case of recurrent nasal schwanna in a 65 year old male which was excised endoscopically.

Case Report
A 62 year old male was referred to V.S Hospital, Ahmedabad in June 2015 complaining of recurrent swelling near right eye, nasal bridge widening causing deviation of nose to left side, nasal blockade, nasal discharge, watering from right eye with anosmia. He had the prior surgical history for the same complaint in 2002 at different setup. The histopathological specimen of the excised specimen revealed the spindle cell neoplasm with moderate to marked nuclear atypia with SOX-10 and S-100 (Schwanoma marker) positive on immunohistochemistry.

External examination showed 3*3 cm non-tender firm swelling on medial canthus of right eye extending to nasal dorsum. [FIG.1]

Anterior rhinoscopy showed painless, pale, firm lobulated mass in the right nasal cavity with deviation of nasal septum towards left side.

On Ophthalmic evaluation, finger counting was more than three feet for both the eyes.

The computed tomography with contrast of nose and paranasal sinus showed 76*28 mm well-defined lobulated mass with heterogeneous post–contrast enhancement in the right nasal cavity. Superiorly, the lesion was extending into right and left anterior ethmoidal and bilateral frontal sinus. Laterally, it was extending into right maxillary sinus along with lateral bowing of right lamina papryacea. Posteriorly, it was reaching upto right posterior ethmoidal and bilateral sphenoidal sinus. Right cribiform plate and anterior skull base appeared destroyed. [FIG.2]

Surgical excision of the mass was planned under general anaesthesia. Informed consent was taken from the patient. Complete debridment of the lesion was done through trans nasal endoscopic approach. Exposed dura was covered by surgicel followed by abgel. Nasal packing was done with merocel.

Post-operatively the patient condition was uneventful.

The excised specimen was sent for microscopic examination which confirmed the diagnosis of schwannoma [same as previous report]. There was no CSF leak and recurrence at six months follow up.

Discussion
Unilateral tumours in the nasal cavity causing nasal obstruction, pain, fullness, and epistaxis are usually caused by benign disease processes such as polyps, cysts, and mucoceles. A unilateral tumour originating from the nasal cavity should also stimulate the consideration of the rare...
Schwannomas of the nose and paranasal sinuses present with similar findings but are very rare.

A schwannoma is a benign tumor only seen arising from the nerve sheath; thus, it is only seen in myelinated nerves. In the head and neck region, the most common site is the eighth cranial nerve (vestibulocochlear); other sites include the scalp, face, parotid gland, oral cavity, pharynx, larynx, and trachea [4]. The optic and olfactory cranial nerves are not potential sites of the origin, since they lack sheaths that contain schwann cells and since nasal schwannomas are sometimes removed without loss of original nerve functions, it is usually difficult to determine the neural origin. Nasal schwannomas are presumed to be arising from the sheath of the ophthalmic and maxillary branches of the trigeminal nerve and autonomic ganglia [5].

The literature mentions approximately only 70 cases of nose and paranasal sinus schwannomas that are mostly seen in adults aged 40-60 years and without gender or racial predilection [6]. The most commonly involved sinus is the ethmoid sinus, followed by the maxillary sinus, nasal cavity, and sphenoid [7].

Patient may complain of nasal obstruction, epistaxis, rhinorrhea, anosmia, facial swelling or pain [8].

Computerized tomography (CT) delineates an image of the soft tissue tumor and simultaneously outlines the skeletal margins well enough to rule out invasion and demonstrated central lucency and peripheral enhancement after contrast administration in case of schwannomas because peripheral neovascular areas of the tumor are enhanced in contrast with nonenhancing necrotic or cystic regions [9]. Although magnetic resonance imaging (MRI) is superior in defining soft tissue tumors, CT offers better resolution of bony invasion. However, as benign schwannoma can erode bone by pressure, bony erosion is not a criterion for malignancy [10]. With either approach, final diagnosis rests solely on the histologic examination.

Macroscopically schwannomas appear as gelatinous or cystic well encapsulated masses. Microscopically schwannomas are classified into three major histological types [11]. Hypercellular or Antony A areas comprised of spindle shaped cells arranged in interlacing fascicles and hypocellular or Antony B areas where the cells are present in a loose myxoid stroma. Parallel rows of palisading nuclei (Verocay body) can be seen in highly differentiated tissue [FIG. 5(a)]. The differential diagnosis includes inflammatory polyps, angiobroma, inverted papilloma, meningiomas, neurofibroma, melanoma and neuroblastoma [11,12]. Histopathology remains gold standard for the diagnosis. Schwannomas usually show intense immunostaining for S-100 which helps to distinguish peripheral nerve sheath tumour from other tumors. Typical microscopic examination shows spindle shaped cells arranged in short bundles and forming interlacing fascicles with nuclear palisading. Antony A and Antony B areas with vascularity bodies (H and E; 10X). (b) Positivity for S-100 thus confirming the diagnosis (10X).

Conclusion
Schwannomas are usually asymptomatic, non-recurrent benign tumor that can go undetected for years. Clinical history and radiographic findings on MRI are required for diagnosis. Surgical excision is the best treatment modality.

References