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NASAL SCHWANNOMA CAUSING COSMETIC DEFORMITY: EASY TO MISDIAGNOSE AND AN EASY-TO-MISS DIAGNOSIS?

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ABSTRACT

Schwannoma is a benign neurogenic tumor arising from the sheath of myelinated nerves. The most common site in head and neck region is vestibulocochlear nerve. 4% of Head and Neck schwannomas involve the sinonasal tract, and out of which origin from the nasal septum are rare. We report a case of a 17-year old female presented with a gradually progressive mass in right nasal cavity since 10 months causing progressive bilateral nasal obstruction and external nasal deformity. Biopsy helped in the final diagnosis of nasal septal schwannoma.

KEYWORDS

Schwannoma, Nerve Sheath Tumor, Nasal Septum

INTRODUCTION

Schwannoma is a benign, slow to grow nerve sheath tumour that can arise in any peripheral nerves covered with Schwann cells. Head and neck schwannomas comprise of 25-45% of all cases. Only 4% involve the sinonasal tract (ethmoid> maxillary sinus> nasal fossa> sphenoid sinus)(1). We report a case of a 17-year-old female who presented with bilateral nasal obstruction. Radiological and clinical examination suggested a benign tumor before the biopsy from the tumor confirmed the diagnosis. Whenever confront a mass in the nasal cavity, schwannoma should be borne in mind in the differentials. We discuss the clinical presentation, histologic features, differential diagnosis, and treatment for such a rare presentation.

Case Report

A 17- year old female patient presented with 10 months history of swelling in the right Nasal cavity, which was gradually progressive. She complained of progressive bilateral nasal obstruction (right> left) and external nasal deformity. There was no history of nasal bleed, facial numbness, anosmia. There was gross broadening of nasal dorsum and obliteration of right nasal groove. On examination there appeared to be a large firm, right-sided nasal polypoidal mass, which seemed to be occluding most of the right nasal airway. The mucosa was ulcerated and crusted; the nasal septum was deviated significantly to the left side of the nose (Fig A). Diagnostic nasal endoscopy could not be done on either nasal cavities because of mass on one side and mass effect on the other side. Endoscopic examination of her pharynx, larynx was otherwise normal. There was no cervical lymphadeno pathy, and the rest of the ear nose and throat and physical examinations were normal. Screening USG neck & abdomen revealed no abnormality.

Non-contrast CT evaluation of the sinonasal region was performed which revealed a 5.5x3.1x5cm size enhancing mass lesion in right nasal cavity causing bony remodelling arising from nasal septum in vestibule confined to right nasal cavity compromising lumen of left nasal cavity & causing left maxillary sinusitis; no bony destruction/ intraorbital/ intracranial destruction seen. (Fig C, D) The proposed differentials were Fungiform papilloma, Malignant mass lesion, Schwannoma.

Biopsy of the nasal lesion was done and tissue on initial histologic examination revealed Pleomorphic spindle cells with hypocellular and hypercellular areas i.e. mixed Antoni A and B substructure and when stained with S100, had a strong positivity, confirming a diagnosis of schwannoma of the nasal septum.

Endoscopic excisional biopsy with septoplasty was done (Fig B). Mass was found to be arising from anterosuperior part of nasal septum, possible origin from nasociliary nerve. Final histopathology also suggestive of Spindle cell neoplasm consistent with Nasal Schwannoma.

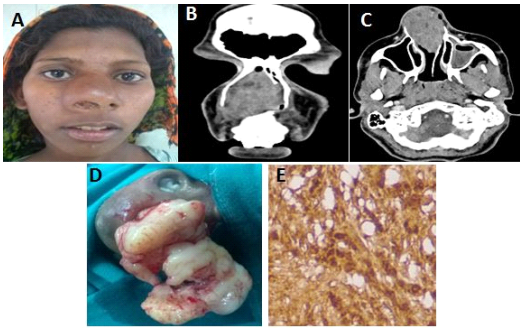

Fig- A. Pre-operative picture of nasal mass causing nasal obstruction, broadening of dorsum of nose and obliteration of nasal groove (Permission for the picture taken from the patient), **B& C.** Coronal and axial images of computerized tomography scan showing a large soft tissue mass in the right nasal cavity arising from nasal septum causing bony remodelling. **D.** Intraoperative picture, **E.** Spindle cells with hypocellular and hypercellular areas with S-100 positivity, and Verocay bodies.

DISCUSSION

To date, only 65 case reports of nasal septal schwannoma have been published in western literature. Nasal septum is an extremely rare site of origin; Vaideeswar et al (2) reported only 3 cases (2.5% of all sinonasal tumours) in a 10- year- long study while Mey et al (3) described 5 cases in 25 years with one case of melanotic schwannoma showing fatal malignant transformation 13 years after initial diagnosis while two others presented with intracranial extension.

A case of Neurilemmoma of Nasal Septum was first described by Bogdasanian and Stout (4), schwannoma is benign encapsulated nerve sheath neoplasm composed of Schwann cells first described by verocay in 1908, Stout (1935) coined the term neurilemmoma

believing that this tumor arises from Schwann cells which may also develop in any part of the body.

Mostly seen in 30- 60 years old with a female preponderance (2- 4:1) and usually present like other benign nasal masses. They are painless and progressive masses causing nasal obstruction, deformities of nasal pyramid, nasal discharge, epistaxis, headache, facial pain occasionally (5) (6) (7). Signs and symptoms depend on the size and location of tumor, and are due to mass effect. Nerve of origin may be difficult to identify during excision. In this case, the origin looks most likely from the somatosensory nerves of the nasal septum (nasopalatine nerve or nasociliary nerve branches).

Radiology is not consistent due to variable cellular pattern- ranging from variably enhancing homogenous ovoid mass to primarily cystic lesion. Usually ovoid, non-infiltrating, bone-remodelling lesions. Majority of sinonasal tumours have intermediate signal intensity on T2W images on MRI, schwannomas may appear with high signal intensity. Magnetic resonance imaging is helpful to evaluate intracranial extension.

Diagnosis is mostly made on biopsy or complete excision. Histologically they comprises two areas- Antoni A (compact arrangement of elongated spindle cells) and Antoni B (loose, myxoid stroma with few elongated cells) with strong S-100 positivity on IHC (vs S-100 negativity for neurofibromas) (8).

Nasal polyps (22.2%), Antrochoanal polyps (19%), Chronic rhinosinusitis (12.7%), concha bullosa (11.1%), retention cysts (6.3%), mucocele (3.2%), schwannomas(1.6%) and Neurofibroma, malignant peripheral nerve sheath tumors (MPNST), myxoma & fibromyxoma, Lobular Capillary hemangioma (LCH), sarcoma form important differentials(8).

Surgery is the treatment of choice; traditional techniques are now totally replaced by endoscopic excisional biopsy and are highly curative for these patients and no reports of recurrence have been published till date(6) (5) (7).

Radiation therapy is booked for cases of nerve sheath malignant tumors. Patients with tumors limited to the paranasal sinuses have outstanding prognosis (9). Endoscopic surgery introduces less morbidity, no surgical scars, and lesser hospital stay. (10)

CONCLUSION

This case is presented to always speculate and include schwannoma in the clinical differentials in patients who present with a nasal mass. Although they require a high index of suspicion for diagnosis as they are rarely encountered and has a non-specific endoscopic and radiological appearance. Even though these are slowly progressive benign lesions, their early detection and complete surgical removal is a must in view of potentially destructive nature and documented malignant transformation.

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