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SINONASAL SCHWANNOMA - A CASE REPORT

Abstract

Schwannomas are benign, encapsulated and slowly growing neoplasms arising from Schwann cells of the sheath of the peripheral nerves. According to the literature, half of the schwannoma cases occur in the head and neck areas and only less than 4% occur in the sinonasal tract. We report a case of a 54-years-old man with a schwannoma involving left nasal cavity, paranasal sinuses engulfing the middle turbinate. We discuss the clinical presentation, differential diagnosis, imaging characteristics and treatment of this rarely encountered lesion.

Keywords: Endoscopic excision, Nerve sheath tumor, Sinonasal Schwannoma.

Introduction

Scwannomas are benign, encapsulated tumours arising from the Schwann cells of the nerve sheath. Approximately 25-40% of all neurilemmomas occur in the head and neck region. The acoustic nerve is the most frequent site involved. Other locations described in the literature include the scalp, oral cavity, pharynx, larynx, parotid gland, middle ear and sinonasal tract.^{1,2}

Sinonasal tract schwannomas are very rare, representing less than 4% of head and neck schwannomas. Patients with sinonasal schwannomas range from 12 to 76 years, with most cases occurring between ages 25 and 55 years. Males and females are affected equally.³

Symptoms and signs associated with sinonasal schwannomas include rhinorrhea, epistaxis, anosmia and facial swelling. Because these tumours are located in a cavity, they are able to grow silently to a substantial size before diagnosis. The most common affected area is the ethmoid sinus, followed by the maxillary sinus, nasal pits, and sphenoid sinus. Localization to the turbinates is exceedingly rare.⁴

This paper reports an unusual case of an sinonasal schwannoma involving left nasal cavity, paranasal sinuses engulfing the middle turbinate that presented as long standing unilateral nasal obstruction.

Case Report

The patient was a 54-years-old male. He visited us

because of a progressive left side nasal obstruction with intermittent purulent rhinorrhea for more than a year. No epistaxis, anosmia or any other nasal symptom was mentioned. He denied any systemic disease and had never undergone any surgery. During his visit, on anterior rhinoscopy, a large polypoid greyish white firm to hard mass was noted occupying whole of the left nasal cavity. Posterior rhinoscopy was normal.

CT scan and MRI scans were done to evaluate the extent of the lesion which showed a well defined soft tissue mass with mild enhancement involving the left nasal cavity, left maxillary and anterior ethmoid sinuses. Middle turbinate was not seen separately. Mass was abutting septum and hard palate. There was no intracranial or intra orbital extension. (Figure I).

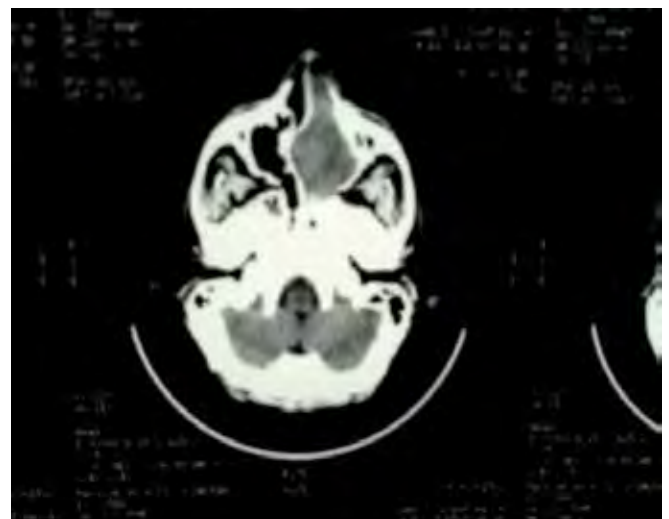


Fig I: CT scan of Nose and Paranasal Sinuses

Biopsy of nasal mass was taken under local anaesthesia and it was reported as schwannoma. The patient was operated under general anaesthesia. Endoscopic trans nasal route was employed for the surgery. Mass was found to be involving left middle turbinate extending into maxillary sinus and anterior ethmoids sinuses. The excised mass (Figure II) was sent for histopathology.

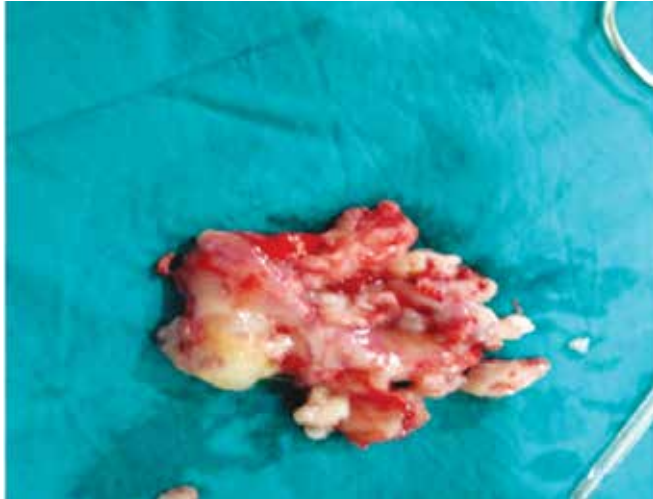


Fig II: Excised Nasal mass

Histopathological examination showed both spindle shaped schwann cell rich area with nuclear palisading (Antoni A) and schwann cell poor loose myxoid areas (Antoni B). Verocay bodies were present with chronic lymphoplasmocytic infiltrates without any evidence of malignant changes. The histopathological diagnosis was benign schwannoma (Figure III). The patient is under follow-up with no signs of recurrence so far.

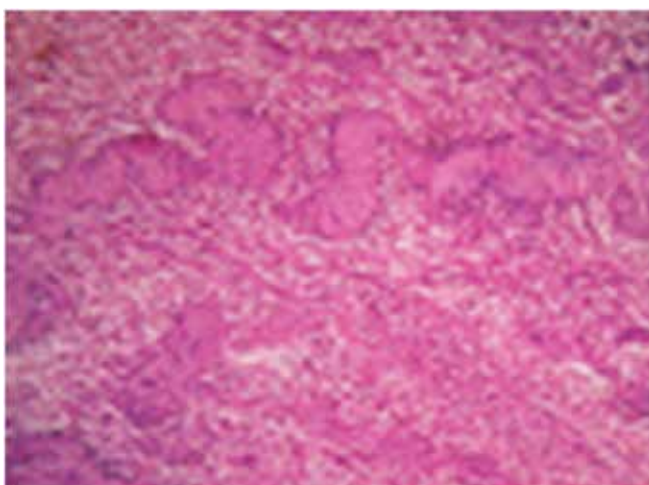


Fig III: Histopathology picture of schwannoma showing characteristic Verocay bodies (H&E x400)

Discussion

Schwannoma is a benign tumor of nerve sheath origin that can arise on any myelinated nerve. The most frequent site affected in the head and neck is the eighth cranial nerve (vestibular nerve). Other observed locations include the scalp, face, oral cavity, pharynx, larynx, trachea, parotid gland, middle ear and external auditory meatus. Whilst schwannomas almost always occur as solitary lesions with no associated genetic syndromes, in some instances they are multiple and occur in association with neurofibromatosis type 2.⁵ Malignant transformation in schwannoma is very rare. Schwannomas of the Sinonasal tract are very infrequent, representing less than 4% of the schwannomas of the head and neck.⁶ In this location they have been reported in patients between the ages of 6 years and 78 years. There is no sex or racial predilection. The ethmoidal sinus is most commonly involved, followed by the maxillary sinus, nasal fossa and sphenoid sinus. The clinical presentation of Sino nasal schwannoma is often varied and non-specific. Patients may complain of nasal obstruction, epistaxis, rhinorrhea, anosmia, or facial swelling and pain. There are no distinctive features to be noted on examination. Consequently, the diagnosis is only likely to be made once histology results are available. The differential diagnosis of a nasal tumor includes a wide variety of pathology including inflammatory polyps, juvenile angiofibroma, inverted papilloma, meningioma, neurofibroma, melanoma and olfactory neuroblastoma.⁷ Macroscopically, schwannomas appear as gelatinous or cystic, well encapsulated masses. Cystic degeneration, necrosis, lipidization and formation of angiomatous clusters of blood vessels with focal thrombi are degenerative processes that can occur. Microscopically, schwannomas are traditionally classified into two major histological types. Antoni A is characterized by a compact arrangement of spindle cells. Antoni B is typified by loose myxoid stroma with spindle cells running in a haphazard manner.⁸ The distinction is considered to have only academic interest. Securing the diagnosis on the basis of high-resolution imaging is difficult. In general, the appearances on CT are not specific enough to enable it to be distinguished confidently from other tumors in this region.

On CT, paranasal schwannoma usually have mottled central hypodense foci with peripheral enhancement after injection of contrast medium. The heterogeneous appearance is related to areas of increased vascularity with adjacent nonenhancing cystic or necrotic regions. This is important in distinguishing it from inflammatory polyps. The imaging characteristics of sinonasal schwannoma on MRI are similar to those of schwannoma observed elsewhere in the body. An intermediate signal is observed on T1 weighted images, whereas on T2 weighted images the signal varies from intermediate to high. A more uniform enhancement pattern after gadolinium administration has been observed.⁸ The only treatment for schwannoma is wide local excision through an approach allowing adequate exposure as schwannoma are generally radioresistant. But in treating benign schwannoma, functional and cosmetic considerations should be taken into account. Recently, the technique of endoscopic nasal surgery has rapidly developed and transnasal endoscopic excision of benign tumors of the nose, paranasal sinuses and nasal septum has been successful. The tumor mass in our case was found to be engulfing middle turbinate which is very rare and involving nasal cavity, maxillary and anterior ethmoid sinuses with erosion of medial wall of maxillary sinus and successfully excised by transnasal endoscopic approach. A single schwannoma does not recur when completely excised, but intracranial extension of a nasal schwannoma has been reported.⁹

Conclusion

Schwannoma is not a common tumor in the sinonasal tract. Though rarely seen, schwannomas should be considered in the differential diagnosis. Even though recurrences are not seen after total excision of the mass, histopathological diagnosis should be made, and since malignant transformation has been reported in the literature, the patient should be followed up for a long time.

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