Schwannoma of the Sinonasal Tract Clinically Presented As a Nasopharyngeal Angiofibroma

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Abstract: Schwannomas are solitary benign and slowly growing tumors arising from nerve sheath. Approximately 25-40% of all schwannomas occur in the head and neck region [1]. The vestibular nerve is the most frequent site involved. According to literature, other locations include the scalp, oral cavity, pharynx, larynx, parotid gland, middle ear and sinonasal tract [1,2]. Schwannomas of the sinonasal tract represents less than 4% of the schwannomas of the head and neck [3]. Sinonasal schwannomas affects equally males and females and the age ranges from 12 to 76 years, with most cases occurring between ages 25 and 55 years.[4]

Keywords: Schwannoma, Angiofibroma, Nasopharynx, Nasal Obstruction, Epistaxis.

1. INTRODUCTION

Schwannomas are benign encapsulated tumors originally described by Stout in 1935[5]. Batsakis more accurately described these tumours as neurilemmomas, referring to the cells of origin [6]. Schwannomas almost always occurs as solitary lesions [7]. Macroscopically, schwannomas appear as gelatinous or cystic, well encapsulated masses. Microscopically, schwannomas shows two types of patterns: Antoni A which is characterized by areas of high cellularity with spindle shaped cells, often arranged in bundles, palisades, or whirls. Also,groups of compact parallel nuclei are seen and are known as "verocay bodies". Antoni B is loose myxoid stroma with spindle cells running in a haphazard manner [1].

The Sinonasal tract schwannomas are very rare representing less than 4% of all head and neck schwannomas. Most cases occur between ages 25 and 55 years and presented with the following symptoms; rhinorrhea, epistaxis, anosmia, and facial swelling.[8] The most common affected area is the ethmoid sinus, followed by the maxillary sinus, nasal pits, and sphenoid sinus.[8] This paper reports an unusual case of schwannoma that presented as nasopharyngeal mass clinically mimicking a nasopharyngeal angiofibroma.

2. CASE REPORT

A 28-year-old Saudi male presented with 4 years history of recurrent epistaxis and bilateral nasal obstruction. The bleeding was spontaneous, moderate in quantity, 5-6 attacks per year, and painless. Nasal obstruction started gradually mainly from right side then it progress over time to complete bilateral obstruction. The patient also reported a history of headache and anosmia.

On physical examination, the patient appeared well. Examination of his throat, ears and neck did not reveal any abnormalities. A neurologic examination was grossly intact.

Nose examination using rigid nasopharyngoscopy showed soft tissue mass arising from right lateral maxillary wall and obstructing choanae.

His laboratory workup was within normal limits Computed Tomography(CT) of paranasal sinuses showed a well-defined soft tissue mass in the nasopharynx extending into the left nasal cavity and part of the right posterior nasal cavity, measuring around 24X33mm. Contrast study showed mild enhancement with no bony remodeling. (**Figure1**)

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Figure 1. A: Non-Contrast Axial CT showed the mass in the nasopharynx and extending to left nasal cavity and posterior part of right nasal cavity . B: Contrast-enhanced Axial CT showed minimal enhancement with no bony remodeling . C: Contrast-enhanced Coronal CT showed mass occupying nasopharynx . D: Contrast-enhanced Sagittal CT showed the nasopharyngeal mass extending to nasal cavity Cerebral Angiogram showed hypervascular nasopharyngeal mass predominantly on the right side measuring 4X3.7cm mainly supplied by right internal maxillary artery. According to Radiologist report, the mass is most likely to be nasopharyngeal angiofibroma. (**Figure2**)

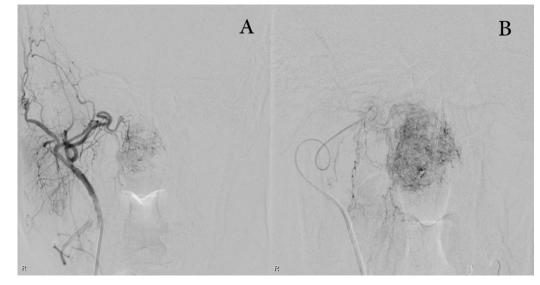


Figure 2. A:angiogram showing main supply of mass by right internal maxillary artery. B:angiogram pre embolization showed hypervascular right nasopharyngeal mass

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The patient was scheduled for surgery under general anesthesia. A preoperative biopsy was not performed because of the bleeding nature of the mass. Angioembolization of right internal maxillary artery branches successfully done 24 hours prior to surgery.(**Figure3**)



Figure 3. Post angioembolization of right internal maxillary artery showing disappearance of the mass.

An endoscopic transnasal approach was chosen for removal of the tumor. The tumor was removed successfully (**Figure4**) and sent for histopathologic examination.



Figure 4. Nasopharyngeal mass post endoscopic transnasal removal

Histopathological examination showed typical Antoni A and Antoni B patterns of a schwannoma. It's also strongly immunoreactive to S100 acidic protein and non-reactive CD34, GFAP, SMA and CD31(Figure5). So, histopathological diagnosis was nasopharyngeal schwannoma.

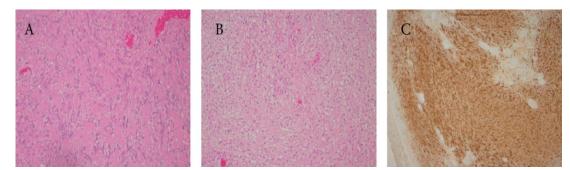


Figure 5. A: Hypercellular "Antoni A" pattern with palisading nuclei surrounding Verocay bodies, B: Hypocellular "Antoni B" pattern with a looser stroma and myxoid change. , C: S100 positive neural marker

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3. DISCUSSION

Schwannomas is a benign encapsulated neoplasm arising from the neural sheath cells of Schwann. It was first described as a pathologic entity by Verocay in 1908 and Antoni described the two histologic patterns in 1920. In the nose and paranasal sinuses, schwannomas arise from the intranasal nerves, from the ophthalmic and maxillary branches of the trigeminal nerve, and from branches of the autonomic nervous system.[$_6$]

Sinonasal schwannomas affects both sexes equally and the most common age ranges between ages 25 and 55 years.[4] Our patient was 28-year-old .

Since schwannomas grow slowly and are encapsulated, symptoms and signs are brought through the tumor expansion or the necrosis of surrounding tissues. Therefore a relatively long period is required before symptoms and signs appear. Nasal obstruction is the most common clinical symptom followed by epistaxis.[$_{9,10,11}$] literature review showed varied clinical presentations . Hasegawa et al reported 6 patients who had complaints of nasal obstruction and epistaxis.[$_{12}$] Alessandrini et al reported patient who had complaints of only nasal obstruction[$_{13}$]. Leakos and Brown reported a single patient with nasal obstruction and anosmia.[$_{14}$] Our patient complained of epistaxis, nasal obstruction, anosmia, and headache.

Computerized tomography is important imaging modality because it 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.[15]

Histopathologic examination is required to confirm a diagnosis of schwannoma. Schwannoma shows histologic pattern of Antoni A and Antoni B areas. The Antoni type A pattern consists of a swirls or palisades of spindle-like cells surrounding a central core of cytoplasm. The Antoni type B pattern consists of a loose cellular array without characteristic swirls or palisades. Although differentiation between Antoni type A and Type B cellular patterns has no prognostic significance, pattern identification aids in diagnosis and is traditional.[11] Higo *et al.* review 160 cases of schwannoma in nasal and paranasal sinuses and reported that Antoni type A comprised 62.5%, Antoni type B 11%, and Antoni type A and B 26.5% .[16]

The only treatment for schwannoma is wide local excision through an approach allowing adequate exposure [9,11,15] because schwannoma is radioresistant [17] 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.[10,18] in our case, endoscopic transnasal approach is chosen.

Schwannoma does not recur when completely excised, and malignant transformation is not reported in the literature.

4. CONCLUSION

We report a 28-year-old male with sinonasal schwannoma which is excised successfully through endoscopic transnasal approach. Although schwannoma of the sinonasal tract is rare, it should be included in the differential diagnosis of any nasal mass.

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