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Case Report

Schwannoma Presenting as a Recurrent Nasal Mass: A Case Report

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Abstract

Schwannoma is a benign peripheral nerve sheath tumor arising from schwann cells. Schwannoma commonly occurs in the head and neck region but nasal schwannoma is extremely rare. Very few cases were reported till date. Sinonasal schwannoma has been postulated to be arising from branches of trigeminal nerve, and autonomic nerve. Nasal schwannoma usually presents as progressive nasal obstruction, anosmia, headache and in long standing cases it may cause nasal deformity, proptosis etc. Recurrence after total excision is quite unusual phenomenon. We report a case of an 18 year old female with schwannoma, presenting as a recurrent solitary mass from the nasal cavity recurring after nine years at the same site, with an emphasis on clinical presentation, differential diagnosis, imaging and treatment of this rarely found lesion.

Key-words: Neurilemmoma; Neurinoma; Neuroectodermal Tumors; Nerve Sheath Neoplasms; Schwann Cells; Schwannoma;

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Introduction

Schwannoma commonly occur in head and neck region and extremities[1] but nasal schwannoma is extremely rare. It accounts for less than 4% of benign solitary schwannoma of head and neck region.[2] Few cases and very few case series were reported till date. Origin of Sinonasal schwannoma from ophthalmic and maxillary branches of trigeminal nerve and branches of autonomic nerve has been postulated.[3]

Case History

An 18 years old female presented with progressive nasal obstruction, polypoidal mass in right nasal cavity, swelling at the right side of the nose near inner canthus of right eye, and headache for last eight years. There was past history of similar type of swelling at the same site at the age of nine years. Surgical removal was carried out at that time in our hospital and diagnosis of nasal neurilemmoma was made. After one year of operation, she again developed mass in the right nasal cavity which was gradually increasing in size and at the age of 18 years she again came to this hospital. In between the period, no intervention could be done as the patient did not attend the hospital.

Schwannoma usually presents as progressive nasal obstruction with or without epistaxis, rhinorrhoea, anosmia, and headache. Long standing cases may cause nasal deformity, proptosis and bony erosion leading to intracranial extension.[4-7] Sinonasal schwannoma are treated with conservative surgical excision and recurrence is very unusual following total resection. Histopathologically, nasal schwannoma is usually not encapsulated unlike other areas[4,6] and need to be differentiated from neurofibroma. We report this case for its rarity.

On examination, there was right sided proptosis and a large mass filling the right nasal cavity. Radiograph of paranasal sinus view revealed a huge soft tissue opacity involving the upper part of the nasal cavity displacing the right lamina papyracaea and medial wall of the right maxillary antrum laterally. Clouding of right frontal sinus was also seen. (Fig 1a) Contrast enhanced axial computed tomography scan of the nose showing irregularly enhancing soft tissue mass in right nasal cavity displacing the medial wall of the right maxillary sinus and lamina was displaced into the opposite nasal cavity. Splaying of the nasal bone and disjunction from the ascending process of maxilla resulted in bringing the mass into the subcutaneous soft tissue place in paranasal area. (Fig 1b & c) Chest X-Ray and other haematological examinations were within normal limit.

Excision of the mass was done and sent for histopathological examination. On gross examination, the specimen consisted of multiple greyish brown tissue pieces with irregular nodular papillary surface. (Fig 1d) On microscopic examination, tumour mass showed respiratory epithelium overlying an uncapsulated tumour composed of spindle shaped cells with nuclear palisading showing both antony A and Antony B areas. (Fig 2a, b & c) Verocay body (Fig 2d) was also noted. The histological features were consistent with schwannoma.

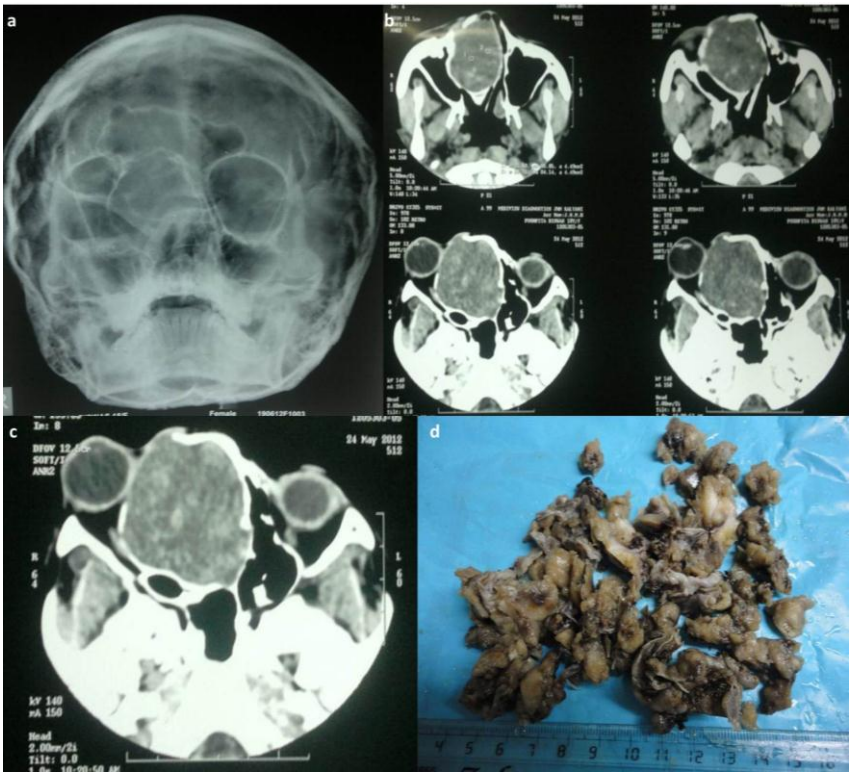

Figure 1: The radiograph of paranasal sinus view (a), contrast enhanced axial CT scan of nose showing soft tissue opacity in the right nasal cavity (b) and conforming the same (c). Gross specimen showing nodular tissue with greyish white areas (d).

Discussion

Sinonasal schwannoma is a rare benign peripheral nerve sheath tumor of nose and paranasal sinuses. A study conducted over five years by Shrinivas S et al,[8] showed nasal schwannoma accounting for 2.4% of benign nasal masses. According to ten years study conducted by Vaideeswar P et al[9] schwannoma formed 2.5% (three cases) of all Sinonasal tumors. Mey KH et al diagnosed and reported five cases of Sinonasal schwannoma over a period of 25 years.[6] There is no age and sex predilection, but in paediatric age group it is extremely rare. Josephson et al,[10] reported one case of paediatric nasal schwannoma. In our case the patient first presented at the age of nine years.

Sinonasal schwannoma is a slowly growing mass. Symptoms depend on the location of the tumor and usually it presents with progressive nasal obstruction with or without epistaxis, anosmia / hyposmia, facial swelling, headache and in long standing cases it may cause proptosis or bony erosion due to pressure effect and may lead to intracranial extension. Epistaxis is common in tumors of the ethmoid sinus and nasal fossae, while pain was related to lesions of the maxillary sinus.[11] In our case, it was a long standing nasal schwannoma which presented as large nasal mass with facial swelling and proptosis.

Histopathologically, most schwannoma are tumour masses surrounded by a fibrous capsule consisting of epineurium and residual nerve fibres. The hallmark of a schwannoma is the biphasic pattern of alternating Antoni A and B areas. Antoni A areas are composed of compact spindle cells arranged in short bundles or interlacing fascicles showing nuclear palisading and Verocay bodies, formed by two compact rows of well-aligned nuclei separated by

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fibrillary cell processes. Antoni B areas are hypocellular areas showing spindle or oval cells arranged haphazardly in the loosely textured matrix.

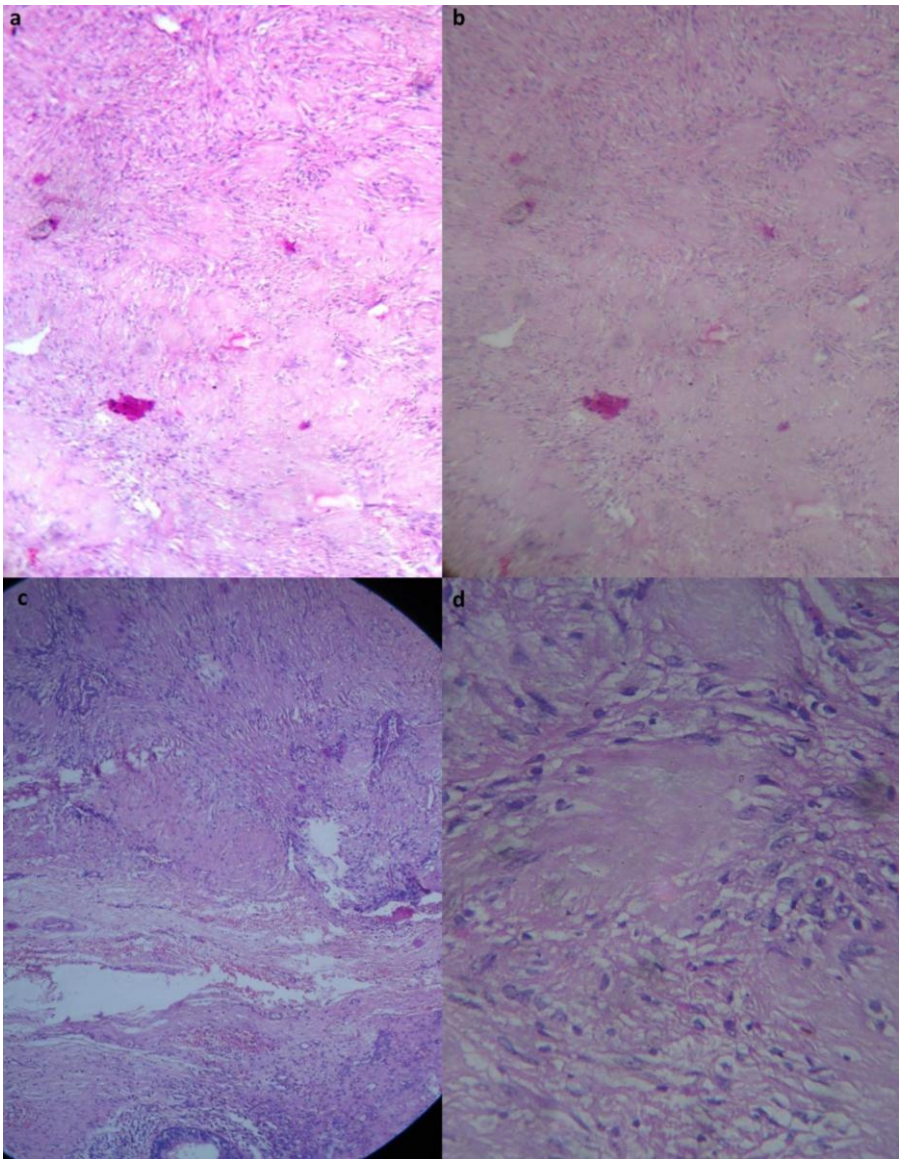

Figure 2: The photomicrograph showing Antoni A & Antoni B areas with nuclear pallisading (a), nuclear palisading with cellular spindle cell areas (b), schwannoma with nasal mucosal glands (H & E stain, x100) (c) and Verocay body in schwannoma (H & E stain, x400) (d).

On occasion, schwannomas develop cystic spaces lined by Schwann cells that assume a round or epithelioid appearance. S-100 protein is strongly expressed by most cells in a schwannoma, in contrast to the cells of neurofibroma, which variably express the antigen. Leu-7 and occasionally glial fibrillary acidic protein (GFAP) are present in these tumors[1]. Unlike other areas nasal schwannoma is usually not encapsulated. Mey KH et al in a case series of five nasal schwannoma[6] and another series of nasal schwannoma cases by Buob et al showed that none of the cases were encapsulated.[4] According to these authors this peculiarity could be explained by the development of these tumours from Sinonasal mucosal autonomic nervous system fibres, which are devoid of perineural cells similar to the case of gastric Schwannoma.[4,6,12] Our case was also not encapsulated probably supporting this theory. The treatment of choice is complete removal of the tumor and recurrence is extremely rare. Malignant schwannoma usually recur but itself is very uncommon.[13,14] Our case showed recurrence but the histological features of malignancy were not there and so the recurrence may be due to incomplete removal. Some authors preferred nerve resection with grafting reconstruction to prevent recurrence but conservative surgery

is still the best treatment of choice in extracranial schwannoma.[15]

In conclusion, solitary nasal masses have a variable differential diagnosis including inflammatory lesions, traumatic deformities, benign neoplasms, malignancies and congenital masses. The most common in the pediatric population are congenital growths such as dermal sinus cysts, encephaloceles, and gliomas. In this population, neurilemmomas rank low on the differential diagnosis for a nasal mass. However, they always should be kept in mind as they can occur at any age and location.

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