CASE REPORT

A case of expansile ancient schwannoma of the nose and paranasal sinuses

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Abstract A rare case of a unilateral nasal mass involving the left nasal cavity and the paranasal sinuses in a 54-year old man with progressive left nasal obstruction is presented. A polypoid mass in the left nasal cavity was evident from anterior rhinoscopy. Computed tomography scan revealed soft tissue attenuation in the left nasal cavity and paranasal sinuses with an erosion of the nasal septum and the compression of the lamina papyracea of left orbit with an extension to its extra-conal space, which suggested the expansile nature of the mass. It was excised endoscopically and found to be an ancient schwannoma, known from histopathological and immunohistochemical studies, which was a rare occurrence in nose and paranasal sinuses.

1. Introduction

Schwannomas or neurilemmomas are neurogenic benign tumours, originating from myelinated schwann cells of the nerve sheath.1 These are encapsulated tumours usually seen in peripheral nerves of limbs, head and neck;2 approximately 25–40% of all neurilemmomas occur in the head and neck region.3 Sinonasal schwannomas are rare, representing less than 4% of head and neck schwannomas.4 Sinonasal schwannomas occur from 12 to 76 years of ages, predominating from 25 to 55 years. Males and females are affected equally.5 Ancient schwannoma is a rare variant of a neurilemmoma, which is a slow growing, benign tumour. A case of ancient schwannoma of the nasal cavity with nasal obstruction had been reported.6 Cases of ancient schwannomas had also been encountered to arise in the retroperitoneal7 and the scrotal region.8

This is a case of an ancient schwannoma of nose and paranasal sinuses with radiological features of erosion of the nasal septum and compression of the lamina papyracea of left orbit suggesting the expansile nature of the tumour.

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2. Case report

A 54-year-old man presented to the ear, nose and throat outpatient department of the hospital, complaining of progressive left nasal obstruction and nasal discharge since 18 months. He had developed bilateral nasal obstruction and anosmia for last 4 months. There was no history of allergy, asthma, diplopia or loss of vision. Clinical examination, full blood counts with differential white cell count, sweat chloride test results and serum immunoglobulin levels were normal. On external examination, there was no proptosis, external nasal deformity or tenderness of paranasal sinuses. Anterior rhinoscopy showed a polypoid mass in the left nasal cavity having an irregular surface. On probing, it was found to be non-tender, soft and mobile. Computed tomography (CT) scan of the paranasal sinuses revealed soft tissue attenuation in the left nasal cavity extending to the left maxillary, ethmoid and frontal sinuses. The nasal septum was partially eroded and pushed to the opposite side. There were signs of thinning and remodelling of the lamina papyracea with bowing into the extra-conal space and compression of the infero-medial wall of the orbit. There was no radiological evidence of any intracranial extension (Fig. 1). Complete endoscopic excision of the nasal mass was performed under the general anaesthesia (Fig. 2). Intravenous ceftriaxone and amikacin were administered to the patient throughout his hospital stay. The postoperative period was uneventful, and the patient was discharged 5 days after surgery. Histopathological examination of the excised specimen revealed a spindle-cell tumour with areas of high cellularity, Antoni A interspaced with areas of low cellularity, Antoni B, along with haemorrhage, necrosis, myxoid degeneration, perivascular hyalinization and cellular atypia without any mitotic figures. ‘S100 immunohistochemistry’ was strongly positive. The final diagnosis was ancient schwannoma (Fig. 3). The patient was given a course of cefixime and anti-histamine tablets during the postoperative period and was prescribed alkaline nasal douching for 1 month and the follow-up endoscopic examination showed healthy nasal and paranasal sinuses (Fig. 4). He had no clinical or radiological recurrence of the nasal mass during the 2 year postoperative follow-up period.

3. Discussion

Schwannomas originate from the myelinated Schwann cells of the nerve sheath and are recognised by the presence of spindle cells with Antoni A and Antoni B regions; and those are designated as ancient, when either cystic or fatty degeneration, focal accumulation of hyaline material, thick capsule, infiltration of histiocytes, siderophages and hyperchromatism are encountered. These histopathological features are attributed to the long duration of the schwannoma.\textsuperscript{9,10}

Figure 1  CT scan of the nose and paranasal sinuses showing soft tissue attenuation in the left nasal cavity and paranasal sinuses with erosion of nasal septum and compression of the orbital wall.
Misdiagnosis of these lesions as sarcomas might arise due to the presence of hypercellularity and atypia, as in a report of 6 cases out of 11 being misdiagnosed as sarcomas. Tumours such as, hemangiopericytomas, leiomyomas, leiomyosarcomas and malignant melanomas, often resemble schwannomas. Moreover, the key feature of differentiating a benign ancient schwannoma from malignant schwannoma is the absence of any mitotic activity. Further, the presence of a capsule, thick-walled vascular structures and areas representing degenerative changes also would suggest the benign nature. But, malignant changes in a benign schwannoma are rarely encountered. The diagnosis of malignant change of a benign

Figure 2 Intra-operative endoscopic photograph showing the mass in the left nasal cavity.

Figure 3 Histopathological picture of the excised specimen showing a spindle-cell tumour with Antoni A and Antoni B regions, myxoid degeneration, perivascular hyalinization and cellular atypia.

Figure 4 Post-operative endoscopic photograph, after one month showing healthy nasal cavity and paranasal sinuses.
schwannoma is based on the criteria signposted, areas of increased cellularity, numerous mitoses, anaplastic cells and invasiveness, along with the absence of clinical evidence of neurofibromatosis-1. Indeed, the treatment of choice for schwannoma is surgical excision, with preservation of the function and integrity of the involved nerve, along with grafting of defective nerve segments in cases where preservation of the nerve is not possible. The nerves, being small in size are rarely encountered during surgical resection of sinonasal schwannomas. In the present case, the tumour most likely originated from branches of the trigeminal nerve of the sinonasal mucosa.

In conclusion, the finding of a schwannoma with degenerative changes observed in histopathological evaluation, along with S100 positivity in immunohistochemical study of the nasal mass suggested the diagnosis as ancient schwannoma. Its expansile nature was evident due to compression of the surrounding structures, nasal septum and orbital wall.

4. Conflict of interest

The authors have no conflict of interest to declare.

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