Sinonasal schwannoma with secondary changes

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Abstract Schwannomas of the Sinonasal tract are very rare, representing less than 4% of schwannomas arising in the head and neck [1]. The most frequent site is the acoustic nerve; other locations include the scalp, oral cavity, pharynx, larynx, trachea, parotid gland, and middle ear. Secondary changes in a Schwannoma are a very rare entity. We report a case of Sinonasal schwannoma with pseudoangiomatous changes. The purpose of this article is to highlight the possibility of unusual presentation of a sinonasal schwannoma

Keywords Nasal · Schwannoma · Pseudoangiomatous

Introduction

Schwannomas are benign encapsulated tumours of nerve sheath origin. Histologically Antony A and Antony B growth patterns are described. Neural elements may also be seen in the tumour. In schwannoma of the nose and paranasal sinuses, due to slow growth and vague complaints, the tumour is noticed at a very late stage. Most of the times, the nerve of origin is compressed and engulfed in the tumour at the time of diagnosis, so is not usually seen.

Case report

A 28-years-old female patient presented to us with a history of progressive nasal obstruction of 4 yrs duration, intermittent epistaxis from the left nasal cavity and gradually developing deformity of the external nose. Bilateral nasal obstruction was observed for 1 year. Hyponasal speech and headache were associated symptoms. There was no weight loss or decreased appetite.

On examination, a firm mass with smooth surface was seen in the left nasal cavity. There was gross deviation of nasal septum to right. The mass bled on touch. A reddish mass was visualized in both posterior choana on posterior rhinoscopy. The ear, throat and neck examinations were without any abnormality. Neurological examination was normal. Haemoglobin percentage, total WBC count, ESR, urine examinations were normal. Plain radiograph of the paranasal sinuses showed haziness of the left nasal cavity, left maxillary sinus and partially the right nasal cavity. The septum was deviated to right.

A CT scan of Paranasal sinuses was done employing 3 mm and 5 mm sections. A large expansile soft tissue density mass was seen in the left nasal cavity with inhomogeneous tissue density and enhancement pattern. There was no calcification. The mass showed increased vascularity. The mass extended from the nasal vestibule anteriorly up to the posterior choana and almost the entire vertical left nasal

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vestibular space. Superiorly the mass was seen abutting the cribriform plate, but there was no involvement or erosion of the cribriform plate. It was seen to displace the adjacent medial maxillary sinus wall. The septum was grossly displaced to right side.

In view of benign clinical course and the radiological findings, a provisional diagnosis of inverted papilloma was made. Biopsy was not done due to increased vascularity of the tumour. Under general anaesthesia, a lateral rhinotomy was done and the mass completely excised. The mass was very friable and there was lot of bleeding. Two units of blood were transfused intraoperatively. Inj. Ciprofloxacin and Inj. Metonidazole were administered intravenously. Analgesics were administered. Patient was observed in ICU and extubated after 24 hrs. The specimen was sent for histopathological examination.

Fig. 1 CT scan showing the mass occupying the left nasal cavity and the grossly deviated septum to right

Fig. 2 Microphotograph showing pseudoangiomatous changes (haematoxylin and eosin $\times 10\times$)

Gross specimen showed multiple grey brown soft tissue bits, largest measuring 3×3 cms, and smallest measuring 0.5 cms. Cut surface had friable grey white areas. Microscopic examination showed thinned out respiratory mucosal epithelium. A spindle cell lesion showing loose and cellular areas occupying the submucosa was seen. Cellular areas showed palisading cell nests, forming Verocay bodies at places only. Much of the lesion was edematous and secondary changes like cystic degeneration, hyalinization and focal pseudoangiomatous changes were seen. There was no evidence of malignancy. A diagnosis of schwannoma with secondary changes was made. Patient was followed up for 3 years. There has been no recurrence so far.

Discussion

Sinonasal tract tumors are usually of epithelial origin, such as papilloma, salivary gland tumors, or carcinoma. Soft tissue tumors of the sinonasal tract are rare and are predominantly vascular and fibrohistiocytic lesions. Neural tumors are unusual in this location. 25% of the schwannomas occur in head and neck regions. Among them, schwannomas of the sinonasal tract are very infrequent, representing less than 4% of the schwannomas of the head and neck [1]. Fewer than 100 cases, mainly isolated tumors, have been described in the literature to date, most cases occurring in patients aged 25 and 55 years. There is no sex predilection [2]. Neurogenic tumours form 6.7% of all sinonasal tumours [3]. The clinical symptomatology is varied and nonspecific, often in relation with signs of chronic nasal obstruction, such as rhinorrhea, epistaxis, anosmia, and facial swelling. The nerve fibers from which the tumor is thought to originate would belong to the branches of the trigeminal nerve. CT scan helps to know the extent of tumour especially the cranial extent and the involvement of vital structures such as the orbit.

The lesions are usually discrete, firm masses often showing cystic degeneration. Although neurilemmomas are frequently attached to a nerve, this is not usually seen in the nasal and sinus neurilemmomas [4]. The small size of the nerves of origin would explain why it is rarely observed during surgical resection. Furthermore, because schwannomas of the paranasal sinuses are located in a cavity, they are able to grow silently over a long time, allowing them to reach a substantial size and thereby surrounding the nerve of origin. Histologically, the diagnosis is made by the appearance of Antoni A and Antoni B areas. Type A tissue is compact and solid and its cells are set in orderly arrangement. Type B tissue is loosely textured, reticulate, patternless and oedematous [5]. Criteria in favor of malignancy are marked cytonuclear atypia, tumor necrosis, and a high proliferation index (>5 mitoses/20 HPF) with abnormal mitoses. In our case these features were not noted. The decreased expression of S100 protein by tumor cells is another argument for

malignancy [6]. Malignant degeneration and intracranial extension may complicate the course of a nasal schwannoma. Complete excision is the preferred therapy [7].

In our case, the presentation was unusual. Symptom wise, there were excessive bleeding episodes and the tissue was friable in nature and it bled on touch. On imaging, it was a mass showing increased vascularity with lack of typical encapsulation. This led authors to make a differential diagnosis of sclerosing haemangioma, inverting papilloma and even malignancy. On histopathological examination features of schwannoma showing Antony A, Antony B areas Verocay bodies were there at few places only. Most of the areas showed angiomatous changes, edematous stroma and hyalinization.

Conclusion

Schwannoma of the nose and paranasal sinuses are usually noticed very late due to their vague symptoms. Those with pseudoangiomatous changes will further add to the confusion by their symptoms resembling that of inverting papilloma and malignancy like epistaxis, friability. A detailed history, examination, CT scan to know the extent of spread and high index of suspicion is essential. Histological exami-

nation is confirmatory. Complete excision is the treatment of choice.

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