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Nasopharyngeal hamartoma: importance of routine complete nasal examination

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Abstract
The authors report clinical experience in managing an 82-year-old female presenting with long-standing bilateral nasal obstruction resulting from a nasopharyngeal mass. The patient had undergone a number of treatments including surgery. The previous examinations, investigations and treatment had all been performed within the previous 10 years and although examination had been documented there was no evidence on review of the notes that the nasopharynx had been inspected either by nasendoscopy or indirectly. The mass was removed via a combined nasal and oral approach. Histopathological examination of the specimen was consistent with mesenchymal hamartoma. In addition to describing a rare presentation the authors believe this case highlights the importance of complete examination in all patients with nasal symptoms.

Key words: Nasopharynx; Neoplasms; Diagnostic Techniques and Procedures

Introduction

Hamartomas are non-malignant malformations or inborn errors of tissue development characterized by haphazard arrangement and growth of tissue indigenous to the specific part of the body where they present. Although common in the lung, kidney and intestines, they are rare in the nasal cavity and nasopharynx. Of these rare lesions adenomatoid lesions tend to be more common; here the authors describe the presentation of a mesenchymal hamartoma.

Case report

An 82-year-old female presented with a 10-year history of bilateral nasal obstruction, and frontal headache. The lady had previously been seen in ENT clinic where she had been diagnosed with septal deviation and enlarged inferior turbinates on nasal speculum examination; these had been treated with septoplasty and subsequent inferior turbinectomy with no improvement in symptoms. A revision septoplasty had recently been recommended to her. During her previous attendances there had been no evidence or documentation of nasopharyngeal examination.

Examination in clinic demonstrated markedly reduced airflow in both nostrils and anterior rhinoscopy showed no evidence of persistent septal deviation. Examination with a 30-degree rigid nasoendoscope revealed a large posterior nasal space mass obstructing both posterior choanae; this mass was also evident per orally using a 70-degree endoscope in the oropharynx. Computed tomography (CT) was obtained and showed an irregular, lobulated soft tissue mass with organized central calcification present in the nasopharynx (Figures 1 and 2). The lesion appeared closely related to the posterior bony nasal septum and hard palate and extended superiorly to the skull base. No associated destruction of bony structures was seen. CT measurements of the mass were approximately 4 × 2.3 cm transverse diameter. Based on these findings the radiologists suggested a provisional diagnosis of osteosarcoma or chondrosarcoma.

At examination under anaesthesia a bony hard mass was found, biopsies were attempted but failed due to the consistency of the tissue. At this time it was seen that the mass had started to separate from the surrounding tissue leaving healthy tissue, the mass was then removed *en bloc*

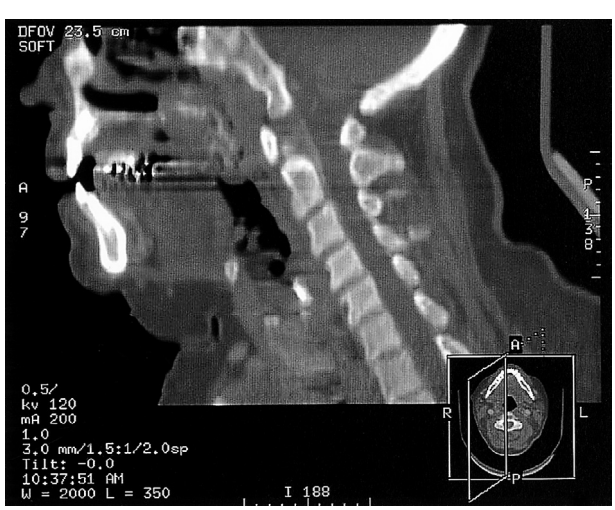

Fig. 1

Sagittal view CT scan displaying nasopharyngeal mass. The mass lies in the area between the posterior choanae and the soft palate.

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via the mouth. Further examination showed only normal tissue remaining in the nasopharynx. There was no significant bleeding and packing was not required.

- **This paper presents an elderly lady who had nasal obstruction for 10 years**
- **She had been diagnosed as having a deviated septum and had even undergone septal surgery without benefit before a post-nasal mass was discovered**
- **The nasopharynx was not routinely examined and, when it was, a large mass that was subsequently diagnosed as a hamartoma was found**
- **The paper stresses the need for a full examination in every case**

The resected mass was hard, cream in colour, 4.5 × 4.0 × 2.3 cm in size, polypoid in shape and weighed 30.7 g. Histological examination revealed that it was covered in respiratory-type epithelium, and was composed of moderately cellular connective tissue containing complex bony structures of woven and lamellar bone. There was a heavily vascularized, cellular, fibroblastic stroma with islands of cartilage and multicentric ossification, which had an organoid appearance (Figures 3 and 4). The diagnosis of harmartoma was made and confirmed by the regional reference laboratory. The lack of immature elements suggested mesenchymal hamartoma rather than benign mesenchymoma.

Post-operative recovery was uneventful. Follow-up at 12 months has not shown evidence of disease recurrence.

Discussion

From this case there are two points to discuss. Firstly hamartomas: these are non- malignant malformations consisting of proliferation of at least one cellular component of a given tissue and are considered an inborn error of tissue development. Although not inflammatory in nature, it is suggested that they may be the result of an inflammatory process .[1] Harmartomas unlike neoplastic lesions are incapable of unimpeded growth and so are self limiting; however, they do not have a tendency to spontaneously regress. The lesion in this case is unlike those previously described being composed of a fibroblastic stroma with multicentric ossification rather than the more typical adenomatous type lesions.[2] They are capable of bony erosion and can affect the skull base[2,3] this is due to mass effect and chronic inflammation rather than infiltration.

Discussion of developmental tumours surrounds the three most common types teratomas, dermoids and hamartomas. Differentiation of the lesions rests on histopathalogical examination: teratomas are composed of all three germ layers and commonly contain tissue foreign to the site of presentation, whereas dermoids contain ectodermal and mesodermal elements, and hamartomas are mesodermal tumours and contain only tissue that is normally found in the specific region of presentation. Teratomas, depending on maturity of elements, have the highest risk of malignant change; hamartomas as stated previously have no/negligible malignant potential.

Symptoms of masses within this area are generally of nasal obstruction and epistaxis, often the patient has described a number of years of difficulties before the diagnosis was made .[3]

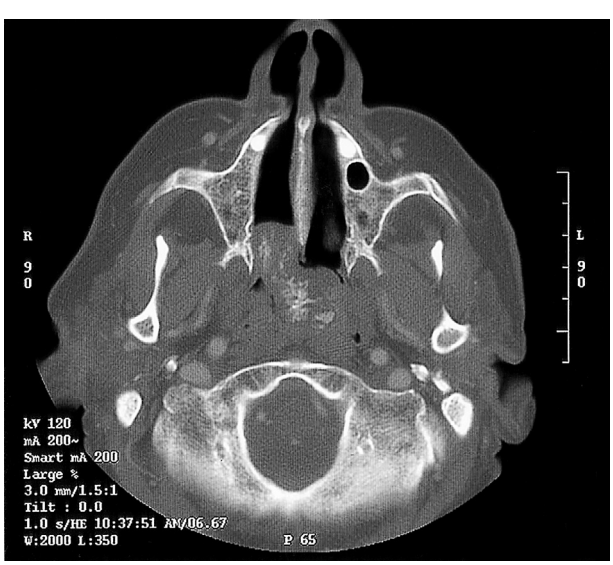

FIG. 2

Horizontal plane CT scan of nasal cavity and skull base showing lesion in nasopharynx obstructing both posterior choanae and extending into right nostril.

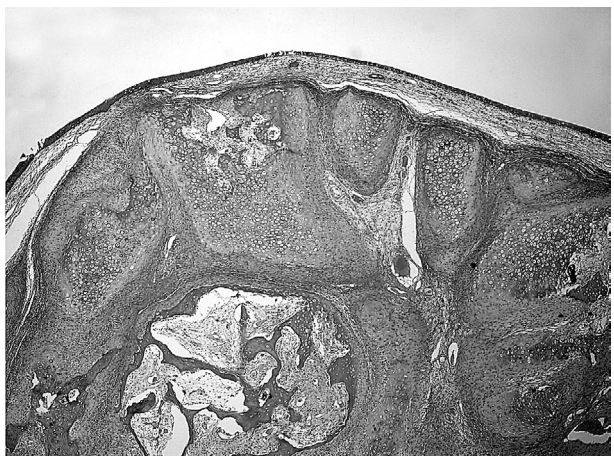

FIG. 3

Mesenchymal hamartoma of nasopharynx: A polypoid lesion with a normal surface respiratory epithelial cover and a core of visualised fibroblastic stroma with islands of cartilage showing ossification.

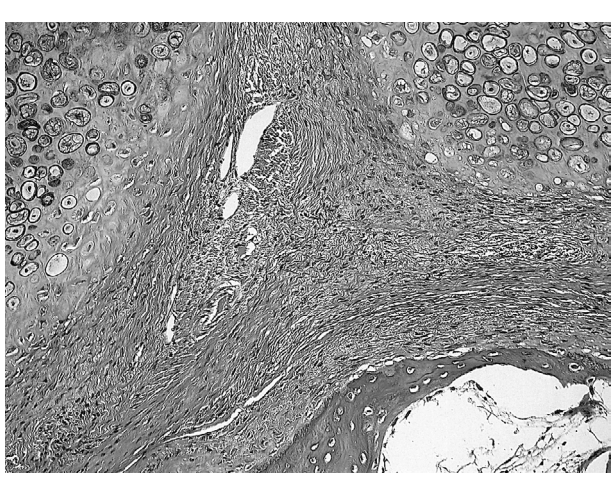

FIG. 4

Mesenchymal hamartoma of nasopharynx: detail of fibroblastic stroma, hyaline cartilage and a focus of ossification.

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Treatment of these lesions is by complete excision as incomplete treatment leaves the risk of re-occurrence. In this case the mass was excised without the necessity for wide margins, due to its nature and the normality of underlying tissue; however wider excision involving craniofacial approaches and lateral rhinotomy have been used[1] and these appear to be more appropriate when the mass encroaches on the paranasal sinuses.

The second point for discussion relates to the diagnosis of these lesions. Given the non-specific symptoms and slow progression, hamartomas of the head and neck are often diagnosed late. In this case, not only was there a delay in diagnosis, but the patient had undergone surgical intervention without benefit and further surgery had been planned. Other authors have noted a supplementary role for rigid nasendoscopy in fine-tuning the diagnosis of chronic rhinosinusitis.[4,5] This case shows that complete examination including nasendoscopy of the nasopharynx should also be performed to exclude the many rare conditions that may occur in this area.

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