Angiofibroma do septo nasal: Uma apresentação extremamente rara

Angiofibroma of the nasal septum: An extremely unusual presentation

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ABSTRACT

Angiofibromas are well-documented tumours occurring almost exclusively in adolescent boys. They are morphologically benign but can be locally aggressive and destructives. Although most angiofibromas originate in the nasopharynx, they are not limited to this region. They can involve the nose, paranasal sinuses and cranium, although primary extranasopharyngeal sites are rare. We report a septal angiofibroma, an extremely unusual presentation, in a 54-year old patient. To date only seven reports have been described in the literature. We discuss the differential diagnosis, and treatment.

Key words: Angiofibroma; nasal septum; extranasopharyngeal.

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INTRODUCTION

Angiofibromas are vascular tumours morphologically benign but locally aggressive. They account for less than 0,5% of all head and neck neoplasms, although they are the most common benign tumour originating in the nasopharynx and are exceptional in patients older than 25 years[1]. Although most of angiofibromas originate in the nasopharynx they are not limited to this region. They can expand to the nose, paranasal sinuses and cranium. However, primary extranasopharyngeal sites are rare. They are most commonly located at maxillary (35%) and ethmoidal (12%) sinus[1]. Angiofibromas arising in the nasal cavity are infrequent and the nasal septum represents an extremely rare localization. To date only seven reports have been described in the literature[2-5]. Etiology of this tumours remains unknown, nevertheless appears to arise from the characteristic fibrovascular stroma normally seen in the nasopharynx. Usually arise from the posterolateral wall of the nasal cavity, at the sphenopalatine foramen, being easy to understand how it can spread to involve the sphenoid sinus and pterygomaxillary fossa. The natural history of the tumour is usually a slowly progressive enlargement. Rarely has a reduction in tumour size been documented as the patient has grown out of adolescence[6].

CASE REPORT

A 54-year-old male was referred to our clinic with a long history of recurrent minor epistaxis from right nasal cavity. A posterior plugging and blood transfusion was necessary for massive epistaxis in one occasion. Examination revealed a grey-red lesion in the area of nasal septum. The nasopharynx was normal and threre was no cervical lymphadenopathy. The suspicion of a vascular neoplasm excluded the need for an endoscopic biopsy. Computed tomography (CT) and magnetic resonance (MRI) demonstrated a 3 cm mass (figures I and 2). Due to the size of the tumour and the potential haemorrhage we elected to use a lateral rhinotomy approach. This approach allowed en bloc

FIGURE 1
Saggital MRI plane showing vascular neoplasm occupying almost the half posterior of nasal cavity.

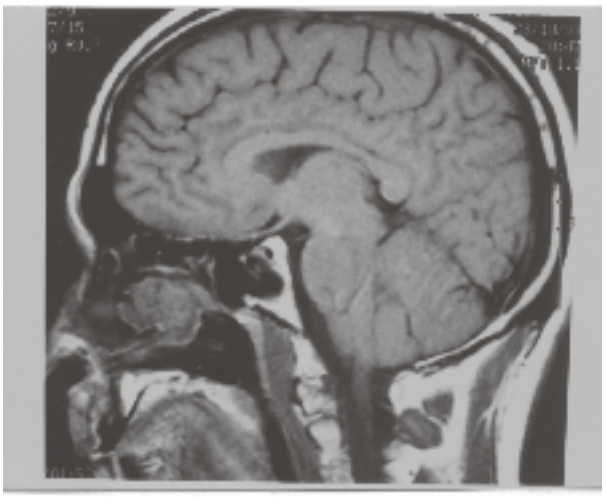


FIGURE 2
Coronal MRI image demonstrates a 3 cm mass (line 1), arising in the nasal septum.

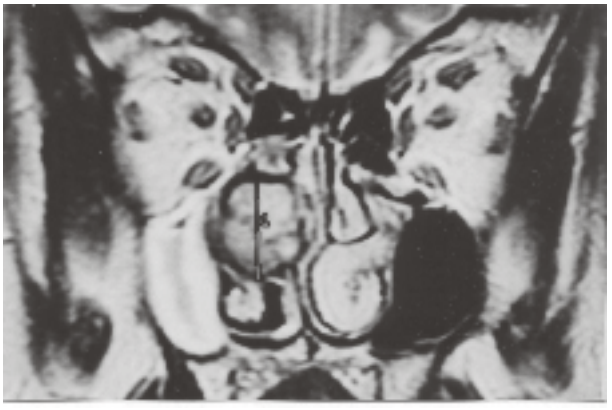
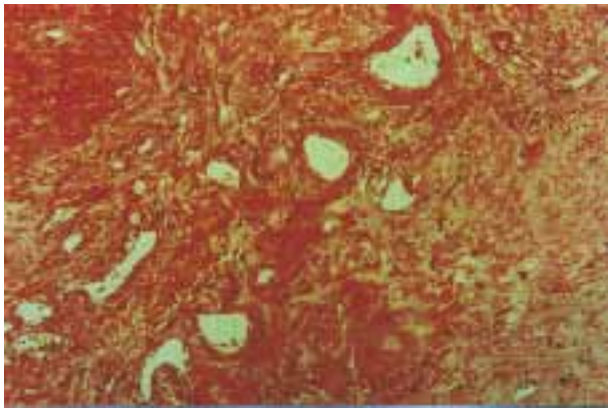


FIGURE 3
Histologic section at 250x magnification, demostrating the characteristic thin-walled vascular network surrounded by fibrous stroma



removal without disruption of the tumour with minimal bleeding. The endoscopic view demonstrated the septal pedicle and his real origin.

The tumour measured 3 x 2,5 cm and it was round in shape. Histological analysis showed an angiofibroma with characteristic erectile tissue appearance containing more fibrous tissue than capillary hemangiomas (figure 3). The tumour was composed of stellate or spindle fibrocytes in a varying amount of connective tissue stroma with many wide, thin-walled vessels.
Three years following surgery the patient remains tumour free.

DISCUSSION

Reports of extranasopharyngeal angiofibroma have appeared sporadically in the literature. From these it is evident that extranasopharyngeal angiofibromas occur in an older age group than nasopharyngeal angiofibromas. Also, the male sex preponderance of the nasopharyngeal angiofibromas is not shared by the extranasopharyngeal angiofibromas. Extranasopharyngeal angiofíbromas have heen reported from many sites in the head and neck region, a comprehensive list of which has been compiled by Sarpa and Novelly[3]. The commonest site of origin is the maxillary sinus.
Imaging is extremely important in the diagnosis and management of these tumours. The characteristic pattern of spread, angiographic appearance, and clinical situation generally obviates the need for a biopsy. CT, MRI, and angiography are the primary imaging modalities for the identification of these tumours. Although this lesion is microscopically benign, it may have an aggressive clinical course extending out of the bony vault of the nasopharynx to compromise vital structures if untreated. In the clinical setting, it is important to distinguish angiofibromas from capillary hemangiomas because of the different natural history of both lesions.
Primary radiation therapy for nasal angiofibroma was a common approach earlier in the 1990s[7]. Because of the excellent results with modern surgical techniques, the addition of CT-based tumour mapping, and concerns regarding the late effects of radiation therapy, the standard of care has become surgical excision of resectable tumours. The use of primary radiation therapy is usually described only for tumours that are considered to be unresectable on the basis of CT criteria.
To date few reports of angiofibromas arising in the nasal cavity have appeared in the literature, and the nasal septum represents an extremely rare localization.

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