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Angiofibroma of the maxillary sinus mimicking mucocele: a diagnostic challenge

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Abstract

Background Angiofibroma is a rare, histologically benign but locally aggressive vascular tumour that typically arises within the nasopharynx. Angiofibroma in sites other than the nasopharynx is extremely uncommon. Despite the histological similarities, the clinical features of this tumour are very much different from the typical nasopharyngeal angiofibroma and pose a diagnostic challenge, hence emphasizing the importance of a thorough evaluation and high index of suspicion in establishing the correct diagnosis.

Case presentation A 25-year-old gentleman presented with progressive right nasal blockage and intermittent nasal discharge for 3 years duration. He was clinically and radiologically diagnosed as a mucocele. Surgical excision was done via endoscopic medial maxillectomy with complete relief of all the symptoms.

Conclusion Although uncommon, angiofibroma can occur outside the nasopharynx. Therefore, thorough evaluation and a high index of suspicion are crucial in establishing the correct diagnosis.

Keywords Angiofibromas, Mucocele, Extranasopharyngeal

Background

Nasopharyngeal angiofibroma (NA) accounts for approximately 0.5% of all head and neck tumours [1]. It occurs almost exclusively amongst adolescent and young adult males between the ages of 14 and 25 years [2]. The most common site of origin of NA is the superior margin of the sphenopalatine foramen. From here, it tends to extend along the natural foramina and fissures to involve the adjacent structures. Sporadic occurrence of angiofibroma outside the nasopharynx is rare and is generally referred to as extranasopharyngeal angiofibroma (ENA) [3]. The clinical features of ENA are rather non-specific and to a great extent related to the sites involved. ENA affects more females, and the tumour is less vascularized in comparison with NA [4]. Similar to typical NA, the mainstay of treatment is surgery [5]. In contrast, a mucocele is a cyst-like structure containing secretion from continuous mucus production. The condition is treated by marsupialization. In this report, the patient presented with a vascular mass which was diagnosed as mucocele preoperatively after a clinical, radiological and histopathological examination. However, a different entity of diagnosis was revealed from the postoperative histopathological examination. Although both diseases are treated with surgical management, the underlying pathophysiology is completely different.

Case presentation

A 25-year-old gentleman presented with progressive right nasal blockage and intermittent nasal discharge for 3-year duration. Otherwise, he had no epistaxis, facial pain or facial swelling. There was no noteworthy past medical or surgical history. There was no facial swelling or tenderness upon examination. Nasal endoscopy

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revealed a vascular mass arising from the lateral wall of the right nasal cavity (Fig. 1).

Contrast-enhanced computed tomography (CT) scan of the paranasal sinuses revealed a mildly enhancing soft tissue lesion fully occupying the right maxillary sinus obliterating the right osteomeatal complex with extension into the right nasal cavity and right ethmoid sinus. The lesion displaced the maxillary sinus roof superiorly and the nasal septum to the contralateral side. There was erosion of the right maxillary sinus's medial and lateral walls and the ethmoid sinus's inferior wall. The sphenopalatine foramen was normal (Fig. 2). These CT findings did not conform to that of typical NA; they were more suggestive of mucocele of the right maxillary sinus. Because of the ambiguous diagnosis, a biopsy was taken preoperatively. As clinically it is a slow-growing and relatively not aggressive soft tissue mass and more of a localized pressure effect, a contrast-enhanced CT scan should have provided the sufficient required information. Magnetic resonance imaging (MRI) is not a routine procedure at least in our centre due to its long waiting time.

On histopathological examination (HPE) of the biopsy specimen, fragments of mucus and inflamed fibrous tissue lined with respiratory epithelium with infiltrating acute and chronic inflammatory cells were seen. These features were suggestive of mucocele.

As the radiological assessment and preoperative biopsy do not show any feature of malignancy, surgical excision was carried out via an endoscopic approach. Some authors suggest endoscopic marsupialization by a middle and/or inferior meatal antrostomy. However, in our case, considering the size and other potential differential diagnosis, an endoscopic medial maxillectomy was performed, and this is thought to be the most conservative window at this stage as any small possibility of inverted papilloma cannot be missed at the first surgery and a revision surgery is almost impossible given the logistic reason in our patient. Intraoperatively, the mass which was occupying the entire right maxillary sinus was removed completely in pieces (Fig. 3). Blood loss was minimal. HPE of the surgical specimen showed fibrous stroma consisting of plump spindles and stellate-shaped cells with varying amounts of fine and coarse collagen fibres. There was marked myxoid degeneration with areas of fibrinoid necrosis and calcification. Scattered vessels ranging from thin-walled, slit-like to occasionally large ones were seen (Fig. 4). These microscopic findings confirmed the diagnosis of angiofibroma.

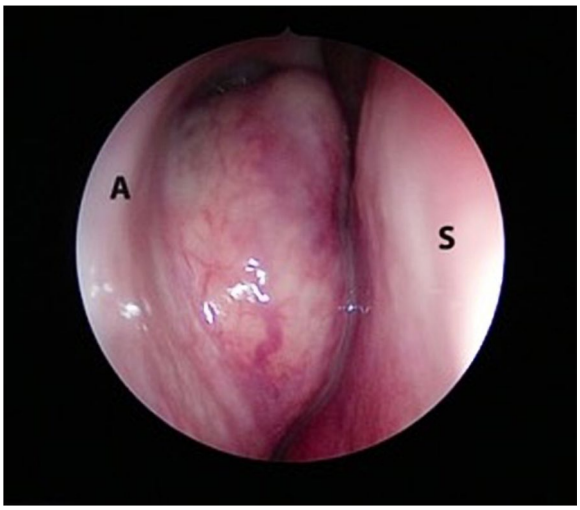

Fig. 1 Endoscopic view of the right nasal cavity showed a vascular mass arising from the osteomeatal complex, seen between the axilla of the right middle turbinate (A) and the nasal septum (S)

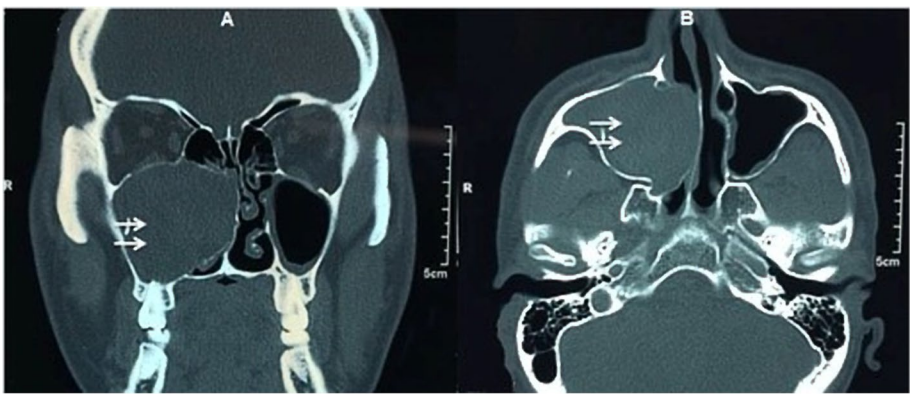

Fig. 2 Contrasted CT showing mildly enhancing lesion in the right maxillary sinus (white arrows) extending into the right nasal cavity and ethmoid sinus seen in coronal view (**A**). The roof of the maxillary sinus is displaced superiorly, and the septum is pushed towards the opposite side as demonstrated in the axial view (**B**)

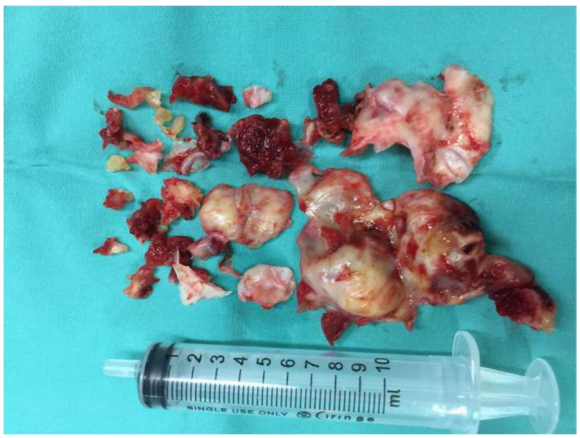

Fig. 3 Angiofibroma removed via endoscopic medial maxillectomy

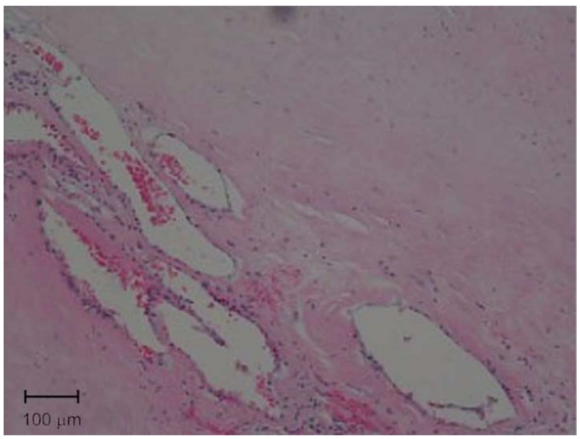

Fig. 4 Resected specimen showing predominant fibrous tissue (upper right) with some vascular component (lower right)

The postoperative recovery was uneventful. The patient was discharged with alkaline nasal douching. At 3-month follow-up, the patient was symptoms-free, and the nasoendoscopy showed no signs of residual disease. Hence, a postoperative CT evaluation was not performed.

Discussion

ENA had been reported to arise from various sites in the head and neck region. The most common location for ENA is the maxilla with the ethmoid, nasal cavity, septum and other sites being involved less frequently. Other rare sites reported are the external ear, external nose, hard palate, lacrimal sac, carotid bifurcation, trachea, oesophagus, facial nerve, middle cranial fossa and infratemporal fossa [6]. There is a possible role of ectopic tissue in the occurrence of angiofibroma in these atypical locations, but the exact cause remains uncertain to date [7].

Some authors have argued that ENA should be considered a separate clinical entity because they have virtually nothing in common with NA regarding the clinical presentation. The use of the term angiofibroma for these lesions may therefore lead to confusion. One of the most striking differences between ENA and NA appears to be gender predilection. Where NA is almost exclusively described in males, ENA is seen more in females. Also, ENA occurs in older patients in comparison to NA, with a mean age of 22 years and 13–15 years, respectively [4, 6]. In our case, the patient's older age of presentation was more consistent with ENA rather than NA.

In terms of the clinical symptoms, unlike NA which typically presents with progressive nasal blockage and recurrent epistaxis, the symptoms of ENA are very much relevant to the affected site and hence highly variable. ENA originating in the nasal cavity may be diagnosed relatively early due to limited space for tumour growth. On the other hand, ENA arising in paranasal sinuses may present relatively late, because of the larger closed space available for the tumour to expand without causing many symptoms for a long period [8]. In our case, the patient presented with 3-year duration of nasal blockage with intermittent nasal discharge. These symptoms were indeed non-specific, mimicking various lesions of the sinonasal cavity including mucocele and chronic sinusitis [9].

Diagnosis of ENA remains a challenge owing to the rarity of this condition and the non-specificity of the symptoms. Albeit the classic radiological findings signifying NA are not shared by ENA, imaging studies are still helpful in clinching the diagnosis. The preferred investigation usually is a CT scan as it better depicts bony changes like expansion or erosion [4]. Magnetic resonance imaging (MRI) is complementary to CT and shows better soft tissue delineation, which is helpful particularly when intracranial involvement is suspected. Due to the frequent poor vascularity of the tumour, contrast enhancement may not be overt in ENA [5]. Having said that, when there is evidence of hypervascularity in imaging studies, angiography is warranted before surgery so that the necessary precautions such as embolization can be exercised to reduce the risk of bleeding. In our case, the poor vascularity of the lesion did not require preoperative embolization.

Histologically, angiofibroma consists of proliferating, irregular vascular channels within a fibrous stroma, with varying ratios between both of them. Oftentimes, the vessels are just lined by a single layer of endothelium without a muscular coat, which provides the vessels with the ability to contract. The abundant vascular components and the lack of contractile ability of the vessels are the contributing factors for sustained bleeding in NA. In

ENA, the vascular components are not as abundant as in typical NA. This explained the low vascularity depicted in the imaging and the lack of torrential bleeding during biopsy and surgery in ENA. It is worth mentioning that in our case, the initial biopsy that was performed in the clinic preoperatively was reported as a mucocele. The possible explanation for this discordance between the biopsy and the surgical specimen is that the biopsy was superficial and not representative of the typical histological appearance of the angiofibroma, which was only seen internally.

Similar to NA, complete surgical excision remains the mainstay of treatment for ENA [5]. The choice of surgical approach is tailored to the site of involvement. Recent advances in transnasal endoscopy, however, have made many of the surgeries amenable to the endoscopic approach [10]. Radiotherapy is an alternative treatment modality that is reserved for those in whom the tumour is unresectable. Where recurrence of NA is not uncommon, recurrence was not observed in ENA [6].

Conclusion

Angiofibroma is a rare condition that can occur beyond the nasopharynx, warranting consideration when diagnosing vascular lesions in the nasal cavity and surrounding structures. A high index of suspicion and a thorough examination are important as the symptoms of ENA are often non-specific.

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SC, EH and MH involved in the literature review and manuscript preparation. RH and IR involved in reviewed.

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Declarations

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Not applicable.

Consent for publication
Written informed consent for publication of the patient's clinical details and clinical images was obtained from the patient.

Competing interests
The authors declare that they have no competing interests.

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