Juvenile Angiofibroma: Case Report and the Role of Endoscopic Resection

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ABSTRACT

Juvenile angiofibroma is benign yet aggressive. Due to its inaccessible location in the nasopharynx, open surgery usually requires osteotomies which cause stunting of facial growth in adolescent males. Advances in imaging and treatment techniques have now facilitated more accurate staging of this disease. For small, extracranial tumours limited to the nasal cavity and paranasal sinus, endoscopic resection is a viable alternative. This case report will illustrate how endoscopic resection allows good control with minimal morbidity compared to open surgery or radiotherapy.

Keywords: Juvenile angiofibroma, endoscopic resection

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INTRODUCTION

Juvenile angiofibroma is a rare (<0.5% of head and neck tumours), locally invasive, histologically benign, vascular neoplasm. It occurs almost exclusively in adolescent males. Distinguishing features include the site of origin, patterns of spread and network of arterial supply. Though benign, its aggressive nature is apparent as it spreads along natural tissue planes and foramina.

It originates[1] in the superior margin of the sphenopalatine foramen, formed by the trifurcation of palatine bone, horizontal ala of vomer and root of pterygoid process. It grows[2] forward into the nasal cavity, upwards into the sphenoid, laterally into the pterygopalatine fossa, and via the pterygomaxillary fissure into the infratemporal fossa.

Intracranial extension can occur through the floor of middle cranial fossa, via foramen rotundum into cavernous sinus, or through the sphenoid sinus into cavernous sinus. Extracranial tumours are supplied by the external carotid system, with a strong contribution from the internal carotid system once there is intracranial extension.

Recent advances in imaging and in surgical instrumentation have allowed an evolution of techniques in treatment. Previously, treatment included expectant management (awaiting the rare spontaneous regressing at sexual maturity), irradiation, surgery, embolisation and hormonal treatment. With the advent of accurate staging of disease, the choice of primary treatment modality has been fine-tuned.

The role of minimally invasive therapy in the form of endoscopic resection is now possible for small, extracranial tumours. We shall now illustrate with one such case how endoscopic resection is an alternative to open surgery, offering good control yet minimising morbidity, for early stage juvenile angiofibroma.

CASE REPORT

NCK is a 20-year-old Chinese gentleman who was previously healthy. He presented with increasing left nasal obstruction and intermittent left-sided epistaxis of spontaneous onset. There were no associated otologic complaints. No generalised bleeding tendencies. No loss of appetite or weight.

Examination revealed a healthy gentleman with a vascular mass in the left post-nasal space which bleeds on contract. Cranial nerves were all intact. The rest of the physical examination was normal. NCK was diagnosed to have juvenile angiofibroma.

CT scan confirmed a well defines mass with strong contrast enhancement, measuring 3 x 2 cm, in the left post-nasal space, projecting into the anterior nasal cavity. The pterygopalatine fossa was uninvolved. Selective angiography demonstrated the feeding vessels to be from the internal maxillary and the ascending pharyngeal arteries. Embolisation was not performed for reasons stated below.

Due to the limited disease extent, it was possible to excise the mass by endoscopic means. This also allows haemostatic control via transantral ligation of the internal maxillary artery (IMA). This obviates the need for pre-operative arterial embolisation.

Appropriate measures were taken against the potential event of massive haemorrhage. Consent obtained included the possible need for external carotid ligation. The patient was grouped and matched.

Excision was carried out under general anaesthesia. Endoscopic removal of the medial maxillary wall, together with middle turbinectomy and ethmoidectomy, allowed good visualisation of the posterior antral wall. The lamina papyracea was left intact. A window was

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created in the posterior antral wall, through which branches of IMA were identified and liga-clipped. The tumour was then carefully dissected from its attachments. Haemostasis was secured with diathermy. A post-nasal pack was left in place for 72 hours. Estimated blood loss was 750 mls.

The post-operative recovery was uneventful.

Final histology revealed a polypoidal lesion comprising haphazardly arranged vessels in a collagenous stroma, diagnostic of angiofibroma.

Patient was free from recurrence at review one year later.

DISCUSSION

The aim of treatment of juvenile angiofibroma is to achieve symptomatic control, if not, complete eradication of the disease. This will depend on accurate staging of the disease extent, appropriate choice of treatment modality and surveillance for disease recurrence.

The discussion will highlight the current thinking in the management of juvenile angiofibroma and the role of endoscopic resection for the small, extracranial angiofibroma.

Staging

Various staging systems have been developed since Sessions et al[3] in 1981. Chandler et al[4] (1984), Antonino[2] (1987) and Radkowski[1] (1996) have proposed new staging criteria. Each staging system identifies the tumour extent, differentiating localised from extensive disease and extra- from intracranial disease. Careful staging allows selection of optimal treatment modality, minimising treatment morbidity yet maximising local control.

Imaging

Appropriate imaging facilitates accurate assessment of disease extent. Lloyd et al (1999)[5] in his review of 20 years of imaging, illustrated that CT is not only diagnostic (pathognomonic enlargement of sphenopalatine foramen), but also prognostic. He found that cases with CT findings of simple pressure erosion (40% of cases) of sphenoid bone accounted for only 7% of recurrent tumours, whereas, cases with deep invasion of sphenoid (60%) accounted for the other 93%. Based on CT findings of sphenoid invasion, more rigorous therapy should be planned.

Angiography

Selective angiography identifies the feeding vessels and allows the option of pre-operative embolisation for vascular control. Studies[2,6] have reported a reduction of estimated blood loss to <1L for cases operated within 48 hours of embolisation.

However, the role of embolisation in tumour recurrence has been controversial. Improved visualisation post-embolisation facilitates more complete tumour excision, hence decreasing the risk of recurrence. However, Lloyd et al[5] found that embolisation results in a greater risk of incomplete excision, due to less defined margins, especially if there is deep invasion of the sphenoid bone. Hence, embolisation is not recommended for all cases.

Treatment Modality

Amongst the treatment modalities available, the primary forms have been surgery and radiotherapy. Combination therapy has been used for extensive tumours which are either surgically unresectable or have intracranial extension.

Radiotherapy

Despite having similar control rates[5,7,8] as that for surgery, when used as the primary treatment modality, radiotherapy is now reserved for unresectable or life-threatening disease. Its side effects[9] include stunted craniofacial growth and potential carcinogenicity, hence, it is not recommended as the treatment of choice for adolescent males.

Surgery

Selection of the surgical approach[9] is dictated by several factors – tumour location and extent, tumour vasculature and effectiveness of embolisation, surgical expertise available and craniofacial age of the patient. Approximately 40% of vertical maxillary growth occurs after 12 years of age[9]. Hence, craniofacial surgery in pubertal males undesirably results in facial growth retardation.

The surgical approaches include endoscopic transnasal, transpalatal, medial maxillectomy (via midfacial degloving or lateral rhinotomy), LeFort 1 osteotomy, and infratemporal with/without craniotomy.

Tumour limited to the nasal cavity and paranasal sinuses can be managed by endoscopic or transpalatal approaches. More extensive pterygopalatine involvement needs the exposure afforded by the LeFort 1 or medial maxillectomy approach. Lesions extending to the infratemporal or to the cavernous sinus requires an infratemporal approach.

Endoscopic Transnasal Approach

This[9-11] is the only approach which causes minimal soft tissue and bony disruption, hence, it offers better cosmesis and no craniofacial growth retardation. It offers excellent endoscopic visualisation of the paranasal sinuses compared to other approaches. It obviates the need for pre-operative embolisation as it allows transantral control of internal maxillary artery (the main feeder of juvenile angiofibroma). Hence, it is the approach of choice for this case.

However, due to limited access, it is recommended only for small, extracranial tumours confined to the nasopharynx, nasal cavity and sphenoid sinus.
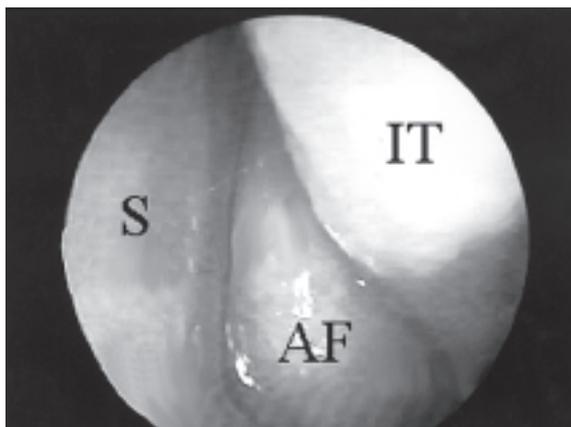
Fig. 1 Endoscopic appearance of the juvenile angiofibroma in the left nasal cavity (AF: angiofibroma, IT: inferior turbinate, S: nasal septum).
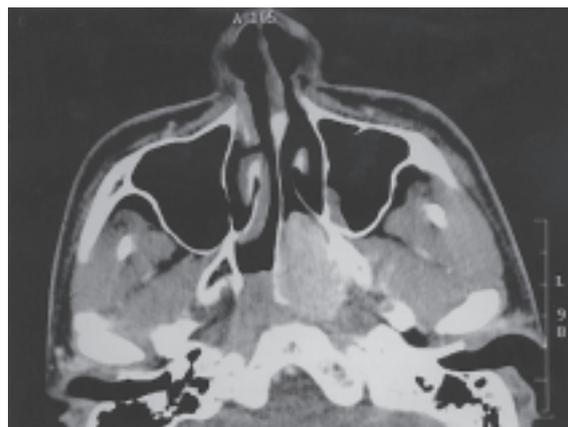

Fig. 2 Axial CT scan demonstrating an enhancing mass in the left posterior nasal space, in close proximity to the sphenopalatine territory.
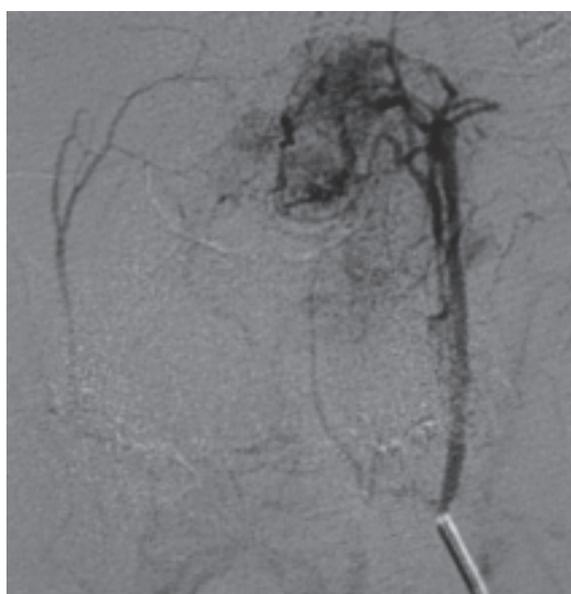

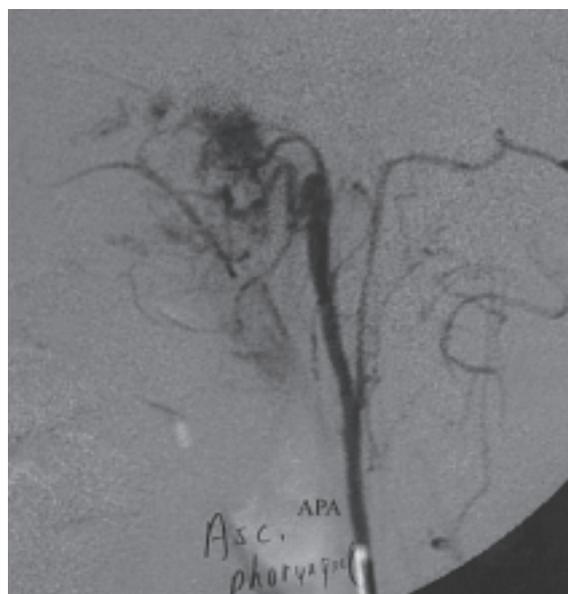**Fig. 3** Antero-posterior view of the angiogram demonstrating tumour vascularity.

Fig. 4 Lateral view of the angiogram demonstrating the feeding vessels from branches of the internal maxillary and ascending pharyngeal arteries.

CONCLUSION

Endoscopic transnasal resection of juvenile angiofibroma is a viable surgical approach for small, extracranial tumours. It offers excellent visualisation of intranasal disease, and vascular control via transantral ligation of internal maxillary artery obviates the need for embolisation and hence its attendant risks. It also spares craniofacial skeletal osteotomies hence avoiding stunting of facial growth in adolescent males.

Accurate staging of disease after appropriate imaging will identify patients with limited disease in which this approach is possible.

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