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Juvenile angiofibroma – Imaging techniques in diagnosis

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INTRODUCTION

In 1847 Chelius described a fibrous mass arising in the postnasal region at around puberty and 30 years later Gosselin remarked on its propensity for young males and its potential for regression. Whilst Chaveau (1906) introduced the term "juvenile nasopharyngeal fibroma" the vascular nature of the lesion was recognised by Friedberg (1940) in the name "angiofibroma".

Because of the natural reluctance of ENT surgeons to biopsy such a potentially vascular mass presenting in the nasopharynx of a young male patient, it may devolve on the radiologist to make the initial diagnosis and thereafter to define the extent of the mass which may influence the surgical approach. These objectives may be achieved by conventional radiology, CT scanning and more recently MR.

METHOD AND MATERIALS

There were 37 patients with histologically proven angiofibromata investigated over a 16 year period. All were males aged between 8 and 29 years (average 15.7 years) and all were initially investigated with plain X-ray though these were not available for review in seven cases as the patients were secondary referrals. Twenty-six underwent hypocycloidal tomography and 27 were investigated by computerised tomography. Angiography was performed in nine and five have been examined by MR.

RESULTS

In all cases, a soft tissue mass was demonstrated in the nose or nasopharynx. Erosion of the base of the medial pterygoid plate, associated with enlargement of

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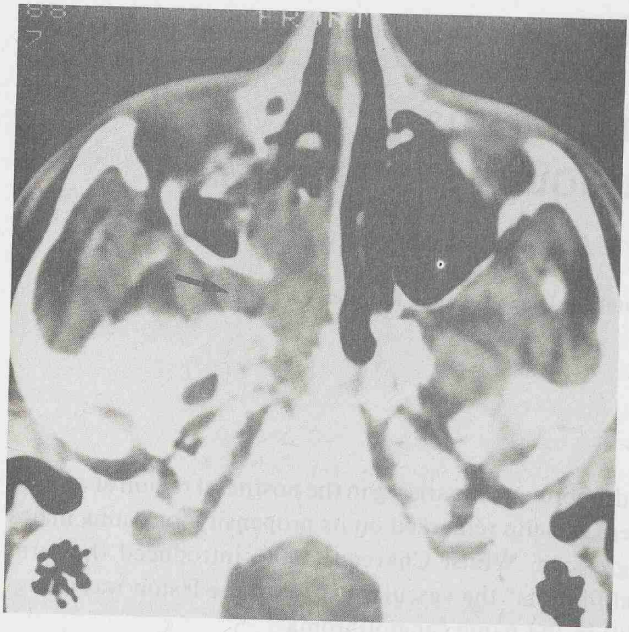

Figure 1. Axial CT scan showing extension of an angiofibroma into the infratemporal fossa (arrow) with widening of the pterygopalatine fossa and forward bowing of the posterior wall of the antrum.

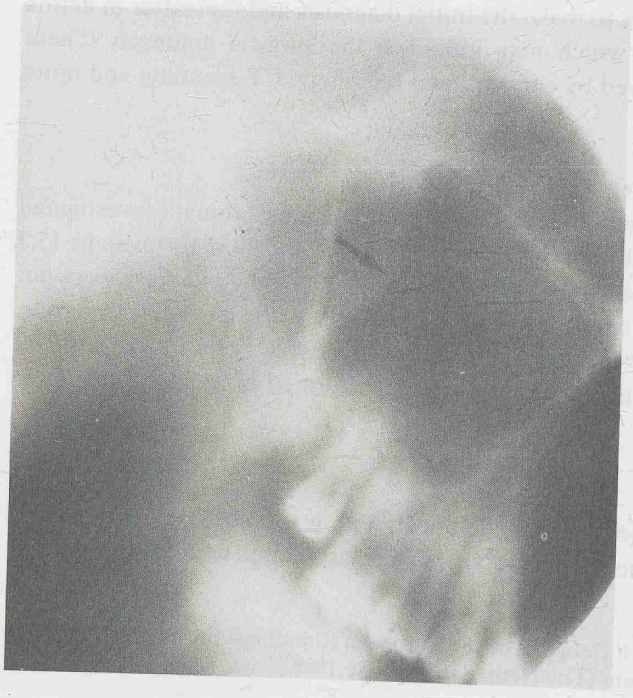

Figure 2. Lateral hypocycloidal tomogram of the sinuses showing an early antral sign (arrow) with forward bowing of the posterior antral wall.

the sphenopalatine foramen was demonstrated in 100% of the 36 patients examined by conventional or computerised tomography (Figure 1).
Indentation of the posterior wall of the maxillary antrum (antral sign) was shown on lateral plain X-rays in 81% of cases (Figure 2).
The sphenoid sinus was involved by angiofibromá in 73% of patients, the infratemporal fossa in 54%, the orbit in 24% and the middle cranial fossa in 16%. All patients who underwent angiography demonstrated a hypervascular tumour, with a strong tumour "blush" in all but one patient.

DISCUSSION

The clinical history of a young or adolescent male with epistaxis and/or nasal obstruction with a nasopharyngeal mass is highly suggestive of an angiofibroma. As growth continues, facial deformity may result in swelling of the cheek and proptosis but radiological investigations should be diagnostic.
The lesion can be shown to originate in the sphenopalatine fossa and whilst secondary attachment may occur (Friedmann and Osborn, 1983) the mass expands, enlarging the foramen and grows medially in the direction of least resistance, into the nasopharynx and nasal cavity (Figure 1). Thus erosion of the roof of the medial pterygoid plate is one of the earliest demonstrable signs on coronal CT (Figure 3). This is followed by indentation of the postero-superior border of the maxillary sinus producing the classic "antral" sign described by Holman and Miller (1965) and is easily seen on plain lateral views and tomography (Figures 1 and 2).
Invasion of the sphenoid and extension into the infratemporal fossa follows via the pterygomaxillary fissure (Figure 3) and this may lead to the mass coming to lie beneath the skin of the cheek by passing between the upper molar teeth and ascending ramus of the mandible (Figure 4). As a consequence, a bilobed mass results, its medial component in the nose and nasopharynx, its lateral part in the infratemporal fossa, the two lobes joined by an isthmus between maxillary antrum and pterygoid plates (Figure 3). It is also possible for the orbit to be invaded via the infraorbital fissure and thence the middle cranial fossa or this can occur by direct lateral extension from the sphenoid.
The relative diagnostic importance of these radiological features must be assessed. Plain X-ray will demonstrate the "antral" sign but it is not pathognomonic nor completely reliable in angiofibroma. Holman and Miller (1965) found it in 87% of their series and it occured in 81% of our patients. Any slow-growing lesion in the infratemporal fossa such as a neurilemmoma, haemangiopericytoma and rhabdomyosarcoma can produce anterior bowing of the posterior maxillary wall. However, medial pterygoid plate erosion is a constant feature in all patients in this series and does not occur in antrochoanal polyps which may, when squamous metaplasia is present, simulate angiofibroma.

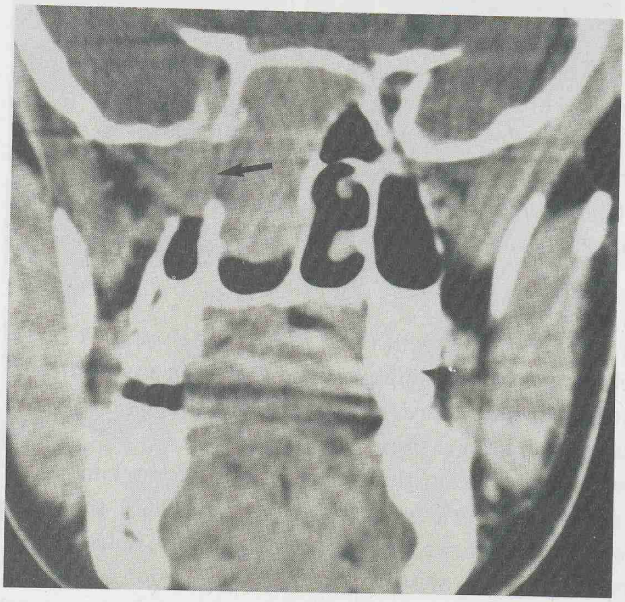

Figure 3. Coronal CT scan showing erosion of the base of the medial pterygoid plate (arrow) with extension into the sphenoid sinus.

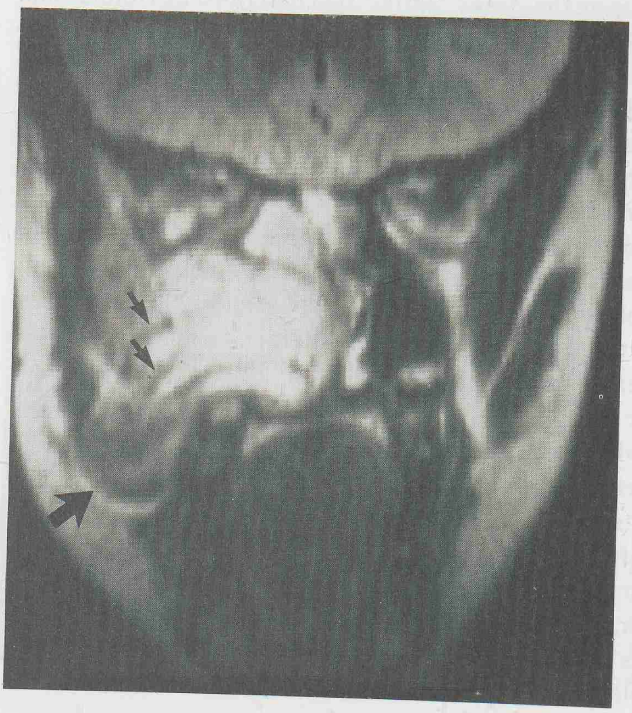

Figure 4. Coronal magnetic resonance scan showing angiofibroma with large vessels within the tumour (small arrows) and bulging beneath the cheek skin (large arrow). (From Lloyd, 1988).

The majority of cases can be managed via a lateral rhinotomy approach but the demonstration of extensive infra-temporal extension may necessitate a Weber-Fergusson approach and this can be shown by contrast-enhanced CT scanning in the immediate post-injection phase or more recently and effectively by MR. The exact size of the lesion relative to adjacent structures can be defined with MR using 3-plane imaging and T_2 weighted spin echo sequences (Figure 5). The distinction between the mass and the fluid in an obstructed sinus can easily be made and the vascularity of the lesion clearly indicated by signal voids from the vessels within the tumour which is usually diagnostic (Figure 6). In contrast, an antrochoanal polyp can be demonstrated arising from the antrum, passing via the ostium into the posterior choana, with a characteristically high homogeneous signal on T_2 weigthed spin echo sequences.

There is little if any evidence that angiofibromas involute spontaneously and the suggestion that they gradually become more fibrous based on random biopsy is meaningless as the distribution of vascular and fibrous components is extremely variable. However, it has been possible to demonstrate some regression of residual remnants after surgical intervention has removed the majority of the lesion (Stansbie and Phelps, 1986).

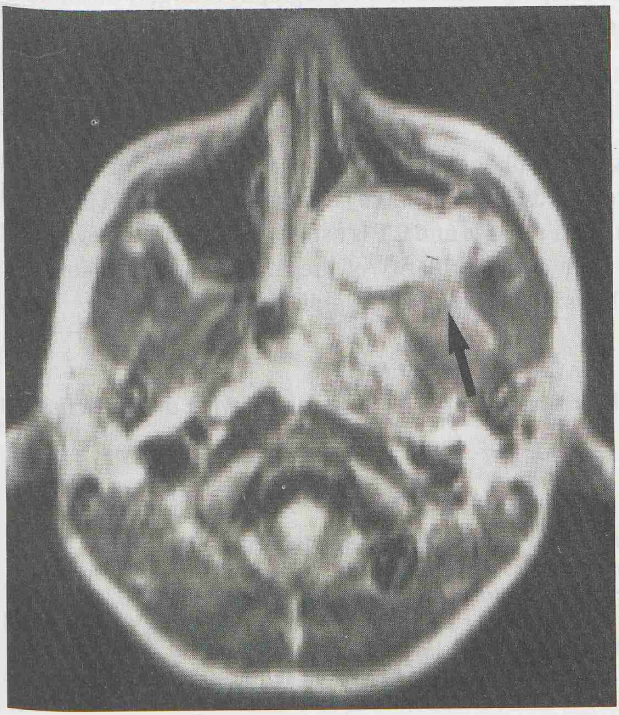

Figure 5. Axial magnetic resonance showing angiofibroma extending into infratemporal fossa.

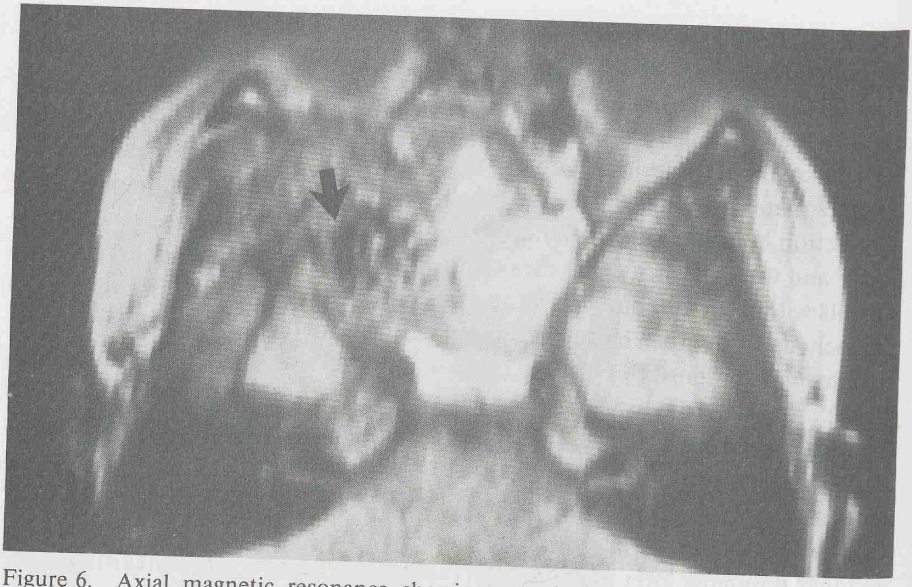

Figure 6. Axial magnetic resonance showing vascularity of the angiofibroma, with vessels as areas of negative signal (arrow). (From Lloyd, 1988).

As a consequence of these imaging techniques, angiography is largely obsolete. It is superfluous to diagnosis and embolisation is rarely employed in the management of these patients.

CONCLUSION

Angiofibroma originates in the sphenopalatine foramen, eroding bone locally at the base of the medial pterygoid plate which is a pathognomonic sign. The antral sign on plain X-ray is not 100% reliable and CT is usually required. However, MR best demonstrates the extent and vascularity of the tumour with non-ionising radiation and angiography is now redundant as a diagnostic technique.

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