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Extracranial internal carotid artery aneurysm in a child: a diagnostic and surgical challenge

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Abstract
This paper reports the presentation and management of an extra-cranial internal carotid artery aneurysm in a 15-year-old male. To our knowledge there is no previous report of a similar case in childhood.

Key words: Aneurysm; Carotid artery, internal; Surgery, operative

Case report

A 15-year-old male was referred with a four-month history of occipital pain, dysphagia and hoarseness with a right-sided neck swelling. This was associated with severe malaise and an 18 lb weight loss. There was no history of cervical injury. Examination revealed a hard mass in the jugulo-digastric area extending into the oropharynx and pushing the tonsil to the midline. A partial Horner's syndrome (myosis and ptosis) was present with ipsilateral adductor vocal fold and hypoglossal nerve palsies. An urgent computerized tomography (CT) scan revealed a non-enhancing 7 cm parapharyngeal mass eroding the skull base and extending inferiorly to the level of the epiglottis. The carotid sheath was displaced laterally (Figure 1).

In view of the nerve palsies and weight loss a neoplastic and possibly malignant lesion was suspected and repeated attempts were made to obtain histology. Ultrasound was performed to guide percutaneous biopsy. Large vessels were seen in close relation to the apparently solid mass and the procedure had to be abandoned because of rapid development of a large cervical haematoma. Subsequent tonsillectomy with ultrasound-guided biopsy through the tonsillar fossa produced normal skeletal muscle and fibrous tissue. A brisk secondary haemorrhage occurred one week later. Finally an open biopsy of the mass was performed through a lateral approach. Chronic inflammatory tissue and sympathetic ganglion cells were obtained from the wall of the mass with a separate reactive lymph node. The mass contained 3 ml pus which grew *Staphylococcus aureus*.

Initial symptomatic improvement on flucloxacillin was not sustained and magnetic resonance imaging (MRI) was performed. This showed a well-encapsulated heterogenous mass and thrombosis of the internal carotid artery in the carotid canal (Figure 2). MR angiogram showed the mass to be highly vascular, consistent with an aneurysm or possibly glomus tumour (Figure 3).

The patient had several small oropharyngeal bleeds and was taken to theatre for excision. A tracheostomy was performed under local anaesthesia to avoid potential intubation hazard and a transmandibular transcervical approach used to give adequate access to the parapharyngeal space and infratemporal fossa (Krepsi and Har-El, 1993) (Figure 4). An encapsulated mass was identified which on further dissection was found to be a large

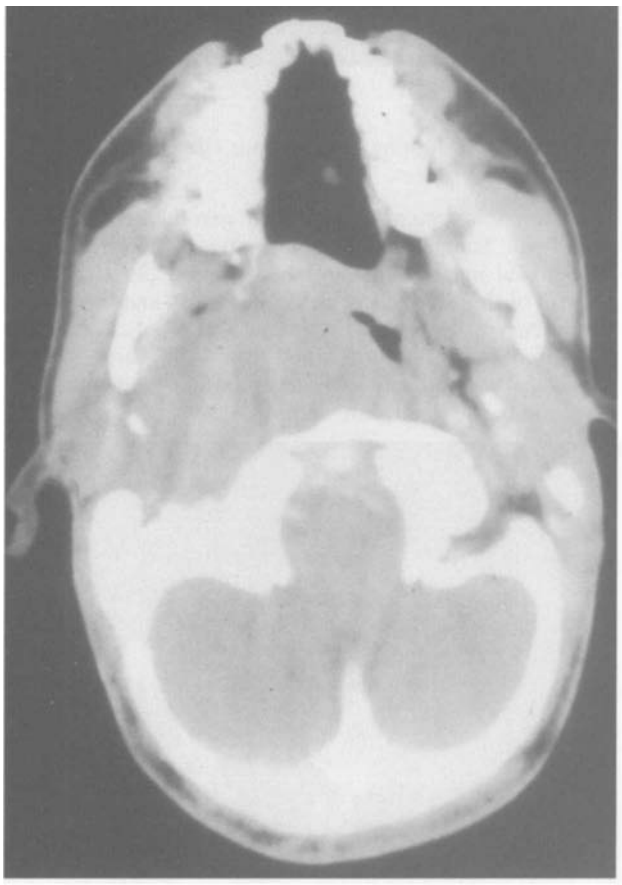

Fig. 1
Axial CT scan with contrast showing non-enhancing mass extending from infra-temporal fossa into pharynx.

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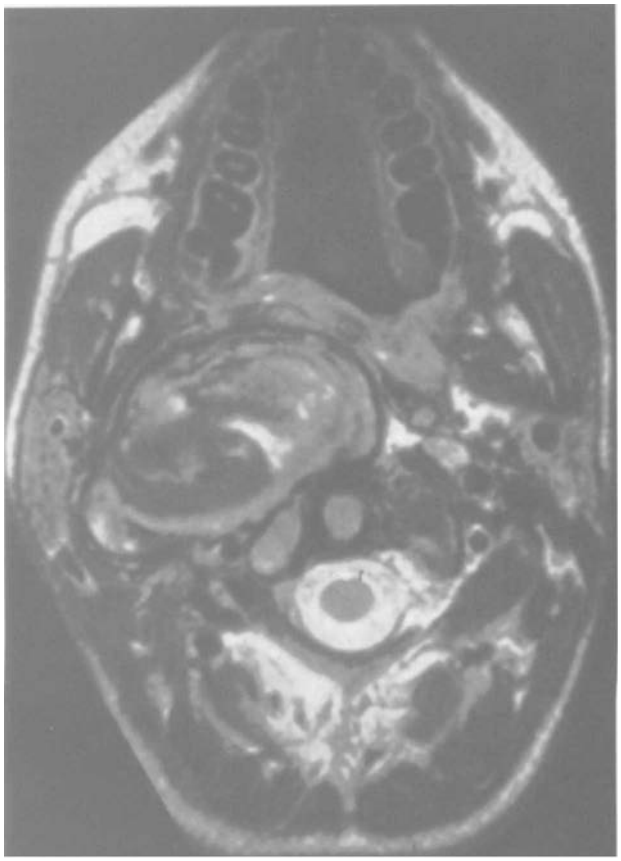

FIG. 2
Axial MRI scan showing heterogenous mass in infratemporal fossa.

saccular aneurysm of the internal carotid artery (ICA) filled with haematoma and thrombus. The artery and hypoglossal nerve were stretched around the aneurysm. Control of the ICA was achieved inferiorly and the wall of the aneurysm excised. Complete thrombotic occlusion prevented back flow of blood from the distal artery. Histology showed myxoid degeneration and muscular dysplasia in the aneurysm wall.

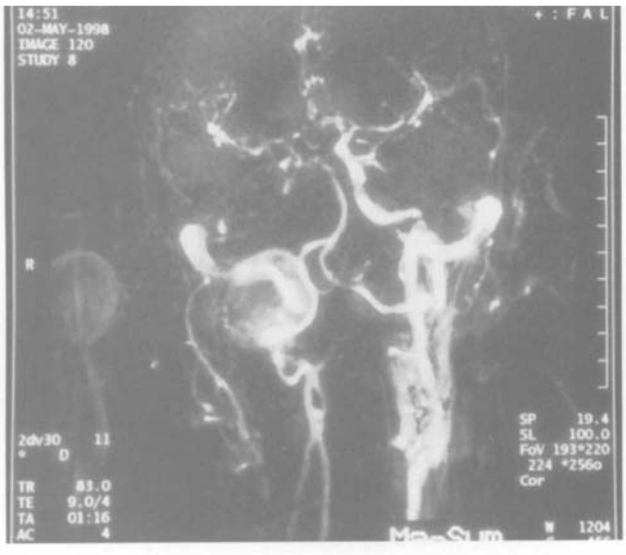

FIG. 3
MR angiogram illustrating vascular nature of lesion of internal carotid artery consistent with aneurysm containing thrombus.

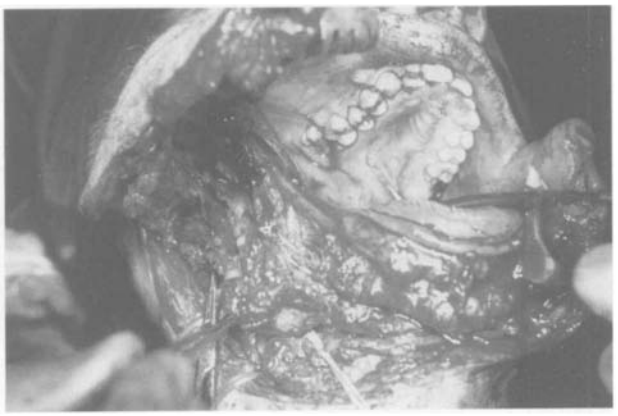

FIG. 4
Operative photograph of transmandibular transcervical approach to skull base. Right hemimandible swung laterally and superiorly.

The patient made a good recovery with resolution of all presenting features except the vocal fold palsy.

Discussion

Aneurysm of the extra-cranial internal carotid artery is a rare condition particularly in childhood. Lower cranial nerve palsies are unusual at presentation and have not previously been reported in childhood. In adults it typically presents with ipsilateral pain and Horner's syndrome and also with a contralateral weakness from cerebral infarction. Atherosclerosis, trauma and surgery are the common causes (Ameli *et al.*, 1983).

The presence of myxoid degeneration and muscular dysplasia in the wall of this young patient's aneurysm suggests that the aneurysm may have been congenital (Hammon *et al.*, 1972; Mettinger, 1982). Although he had no history of major trauma, many extracranial ICA aneurysms are thought to evolve by dissection following trauma and dissections have been reported from seemingly trivial actions, e.g. a blow to the back (Waespe *et al.*, 1988).

Sturzenegger postulates that a vessel wall anomaly allows dissection along the subadventitial layer. Expansion occurs easily in this plane and the resultant aneurysm stretches adjacent cranial nerves causing palsies. This process differs from most ICA dissections which occur in the subintimal layer so causing luminal compression and thromboembolic stroke (Sturzenegger and Huber, 1993).

Mycotic carotid aneurysms are now rare (Worley *et al.*, 1998) and the presence of *Staphylococcus aureus* in the abscess associated with this aneurysm is more likely to be secondary infection from earlier biopsy.

It has been reported that the absence of CNS involvement often delays diagnosis of ICA aneurysms (Tanaka *et al.*, 1983). Diagnosis in this case was delayed as thrombosis of the aneurysm gave it a solid appearance on contrast CT and ultrasound scans. Also, the systemic symptoms and nerve palsies were consistent with malignancy. As in other reports MRI was critical in forming a diagnosis (Coley *et al.*, 1998; Hsu *et al.*, 1998), but excision was necessary to confirm the diagnosis. While a standard approach to the carotid artery is suitable for many ICA aneurysms, close proximity to the skull base requires an infratemporal fossa approach (Rosset *et al.*, 1994). The transmandibular transcervical approach used in this case (Krespi and Har-El, 1993) allows good access and can be extended transpalatally if necessary (Krespi and Cusumano, 1991).

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Summary

Extra-cranial internal carotid artery aneurysm is exceptionally rare in childhood. This case may have arisen from a congenital weakness aggravated by minor trauma. Thrombosis of the aneurysm led to delay in diagnosis and should be considered when performing biopsies at this site.

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