RADIOLOGICAL CASE

CLINICS IN DIAGNOSTIC IMAGING (15)

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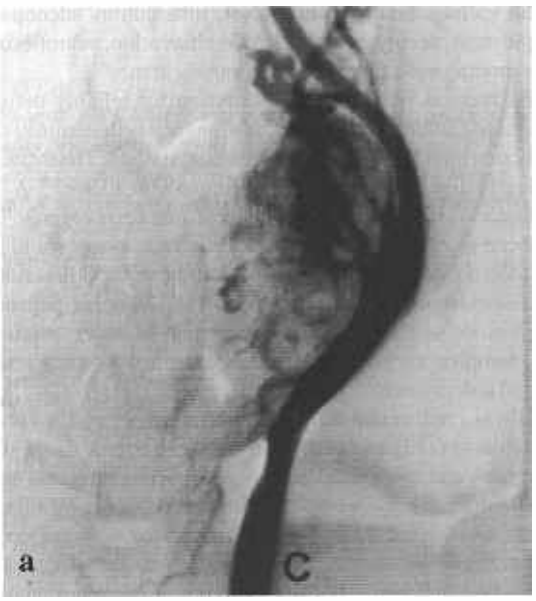
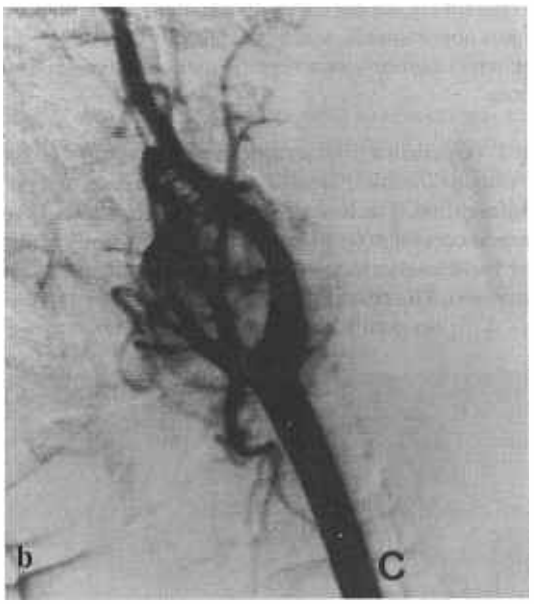
CASE REPORT

A 30-year-old Chinese woman first presented 2 years ago with bilateral neck swellings, more prominent on the right side. Since then the swellings had enlarged progressively and were associated with facial pain and discomfort on chewing. A mass was excised from the right neck at another hospital. Because of residual unilateral vocal cord paralysis post-operatively, she had been reluctant to undergo further surgery to her left neck until her current admission to our institution - by which time she had already recovered her right vocal cord function with speech therapy. On examination, a smooth firm swelling was palpable on the left side of her neck, anterior to the sternomastoid muscle and just below the angle of the mandible. The mass was pulsatile but no bruit was heard on auscultation. Digital subtraction angiography (DSA) of the left common carotid artery was performed (Fig 1a-c). What do these show? What is your diagnosis?

Fig 1 - DSA of the left common carotid artery (C). (a) Early phase (10 seconds), frontal view; (b) early phase (9 seconds), 25° left anterior oblique view; (c) late phase (18 seconds), 25° left anterior oblique view.





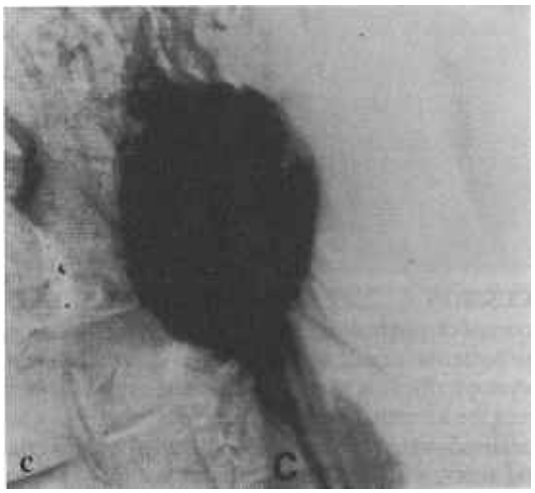


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IMAGE INTERPRETATION

DSA images demonstrated an oval-shaped, highly vascular tumour at the bifurcation of the left common carotid artery. Its blood supply was derived from the lingual and ascending pharyngeal branches of the external carotid artery. The mass displayed early tumour blush and produced typical splaying of the origins of the internal and external carotid arteries (Fig 1a and b). The intense vascularity of the tumour was well shown on delayed images (Fig 1c). The right common carotid DSA was essentially normal with no evidence of tumour recurrence. The previous resected lesion was a carotid body tumour.

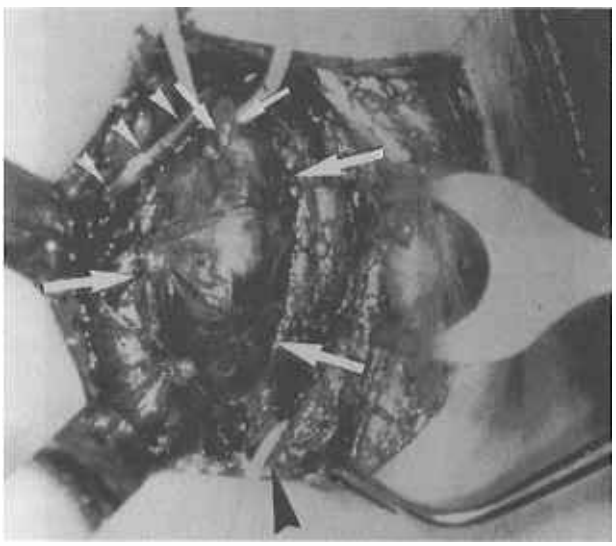
DIAGNOSIS

Carotid body tumour

CLINICAL COURSE

The patient underwent primary surgical excision of the left carotid body tumour. At operation, a well-encapsulated vascular tumour, measuring 45mm x 30mm x 25mm, was found encasing the proximal external carotid artery. The vagus and hypoglossal nerves were also involved (Fig 2). Complete resection was achieved, with preservation of the 2 cranial nerves, internal carotid artery and internal jugular vein. Histopathological examination confirmed the diagnosis of carotid body tumour. Post-operatively, the patient developed left vocal cord paralysis and bronchopneumonia, which subsequently resolved. Screening of her family members were negative for carotid body and related tumours.

Fig 2 – Operative photograph showing the carotid body tumour (arrows) located at the left carotid artery bifurcation. The lower silicon band loops round the common carotid artery (large black arrowhead) while the upper band loops around the external carotid artery (small arrows). The course of the hypoglossal nerve is also marked (small white arrowheads).



DISCUSSION

The normal carotid body measures 3mm by 6mm and is situated at the posterior aspect of the common carotid bifurcation. Embryologically, it is of neuroectodermal origin, developing between the adventitia and media of arterial wall. It is a highly vascularised organ, predominantly supplied by the external carotid artery, which functions in reflex control of heart rate, blood pressure and respiration by the detection of temperature and chemical composition (pH, PO_2, PCO_2) of blood. Physiological hypertrophy may occur in response to chronic hypoxia. A link between carotid body tumour and chronic hypoxic stimulation has been reported, though the vast majority of these tumours have no known predisposing factors[1].

Carotid body tumours can be classified surgically, according to ease of removal[2], or pathologically, according to anatomical site and function[3]. In the pathological classification proposed by Glenner and Grimley, carotid body tumours should more precisely be named 'carotid body paraganglioma non-functional'. These tumours are very rare, with an incidence of 0.012% in one series[3]. About 6% of carotid body tumour patients develop second primary tumours, mainly other paragangliomas, suggesting a large neurocristopathy syndrome of multiple tumours of cells of neural crest origin[4].

The average age of presentation of carotid body tumours is in the late forties with no predisposition to either sex. Bilateral tumours occur most frequently when they are familial. They appear most commonly as a painless, slow growing mass in the anterior triangle of the neck. Due to its attachment to the underlying artery, the tumour is more mobile in the horizontal than in the vertical plane. It is firm to rubbery in consistency and is often pulsatile due to arterial transmission. A bruit may sometimes be audible. Cranial nerve dysfunction is uncommon although vagal, hypoglossal or sympathetic involvement have been reported[1]. Differential diagnoses of a solitary mass in this region include branchial cleft cyst, tuberculous adenopathy, thyroid mass, aneurysm of the carotid bifurcation, neurofibroma, schwannoma and other cervical paragangliomas[1].

As there is no histological criterion for reliably defining malignant carotid body tumour, malignancy is determined only by documentation of nodal or distant metastases[1]. The incidence of local malignant invasion ranges from 3% to 30 %[2,3,5]. A 20% incidence of lymph node involvement has been reported[5]. It has been suggested that all carotid body tumours should be considered malignant because of progressive local invasion of the neurovascular structures, especially in younger patients[6]. This has led to the recommendation that, in order to effect a cure, complete surgical excision be performed for early lesions in good risk patients[1,3,6,7].

Ultrasound examination is non-invasive and a readily available investigation in most centres. It can distinguish aneurysms and cysts from solid homogeneous tumours, and it can also delineate the relationship between tumour and adjacent carotid arteries[6,8]. With duplex Doppler or colour Doppler modes, hypervascularity of the lesion can be detected and the diagnosis established[3]. Because of its non-invasiveness and low cost, ultrasound is ideal for follow-up and screening of familial cases. Radionuclide angiography is also a safe technique but the relatively high false positive and false negative rates, as well as its relatively high cost, render this procedure less reliable for screening[6]. CT gives additional information on the size and extent of the lesion, and is recommended for assessment of intracranial extension, especially if its upper limit is not palpable[3].

Conventional or digital subtraction angiography will accurately delineate the tumour blood supply and provide information about patency of the internal carotid artery prior to surgery. Preoperative assessment of the cerebral collateral circulation is of value if interruption of the carotid arteries during operation is anticipated. Bilateral angiography should be performed in order to detect the contralateral tumour which is present in 5% to 10% of cases. Percutaneous biopsy has no place in the diagnosis of this condition as it carries a high risk of complications like haemorrhage, pseudoaneurysm and carotid artery thrombosis[1,3,6,8].

Primary surgical excision is the preferred treatment. Early operation is desirable as the tumour will be less extensive and

there is less risk of cranial nerve involvement. Regional lymph node dissection in conjunction with complete tumour excision may be performed if malignancy is suspected. An arterial shunt may be necessary where there is extensive or full thickness invasion of the carotid artery or if the artery is damaged during dissection. Bilateral tumours should be removed in a two-stage operation in order to avoid having bilateral vocal cord palsy[3,5,8-10].

The other treatment options of radiotherapy and embolisation are reserved for selected situations. Radiotherapy is only recommended for patients with extensive tumour or intracranial extension where complete resection is impossible or in those with poor surgical risk. Although embolisation has been reported as a useful preoperative measure, bleeding would not be significantly reduced in those cases where the internal carotid artery also supplies the tumour. This procedure also carries a risk of hemiplegia and should only be considered as palliation for irresectable tumours[3].

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ABSTRACT

A 30-year-old Chinese woman presented with an enlarging pulsatile swelling in the left neck. A similar right neck mass was excised 2 years previously. Digital subtraction angiography showed a well-defined, hypervascular tumour situated at the left common carotid arterial bifurcation, typical of a carotid body tumour. Complete resection of the second carotid body tumour was achieved. The clinical and imaging features of this tumour are described.

Keywords: *angiography, carotid body tumour, head and neck tumours, paraganglioma*