Case Report

Haemangiopericytoma of the Internal Jugular Vein: an Unusual Neck Mass

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INTRODUCTION

Haemangiopericytomas (HPC) are rare vascular tumours arising from pericytes of Zimmermann, contractile spindle cells that surround the capillaries and post-capillary venules. They are ubiquitous, in the body due to their vascular origin though lower extremity and retroperitoneum are common sites. We report here a case of HPC of internal jugular vein (IJV), which is an extremely rare site of occurrence, and the findings using different imaging techniques such as including colour Doppler sonography (CDS) and computed tomography (CT).

CASE REPORT

A 26-year-old lady presented with progressively increasing painless swelling on the left side of the neck over 2 months. She underwent fine-needle aspiration cytology (FNAC) of the lesion at a peripheral hospital, and was referred because of a sudden increase in the size of the swelling. In view of the clinical presentation, CDS of the neck was performed to exclude a vascular lesion and revealed a mixed echogenic mass measuring 4 x 5 x 6 cm, highly vascular with low resistance flow pattern in the left carotid space from the IJV with partial obliteration of its lumen. Non-enhanced CT of the neck revealed a blood–fluid level within the lesion suggestive of haemorrhage (Fig. 1). On contrast medium administration, the left IJV was not seen separate from the lesion and there was intense enhancement of the lesion (Fig. 2). There were no foci of calcification or infiltration into adjacent structures. Peroperatively, a highly vascular lesion from the IJV was noted. Excision of the lesion along with a segment of IJV was performed. The histopathological examination showed a cellular lesion separated by dilated vascular channels surrounded by round to oval cells with vesicular nuclei; mitoses 3–4/10 high-power field (HPF) were present; Sections from the vein also showed similar lesion and collagenous connective tissue. Gomori’s reticulin staining technique revealed reticulin condensation around individual cells and blood vessels confirming the vascular nature of the tumour. The final diagnosis was HPC of the IJV (Fig. 3). The chest radiograph and routine laboratory investigations were normal.

DISCUSSION

HPC is an uncommon vascular tumour. Stout and Murray [1] introduced this entity in 1942 to distinguish this puzzling tumour of blood vessel origin from the closely related glomus tumour (benign) and angiosarcoma (malignant). It occurs in all age groups with equal incidence in both males and females. Enzinger and Smith [2] evaluated 106 haemangiopericytomas and concluded that the commonest site is lower extremity (35%) followed by pelvis or retroperitoneum (25%), head and neck (16%), trunk (14%) and upper extremity (10%). It has also been described occurring in the brain and spine, oesophagus, breast and lung [3–7]. In the head and neck region it has been described in the orbit, nasal cavity, oral cavity, jaw, parotid gland, parapharyngeal space, masticator space, jugular foramen, etc. [8–15]. However, there has been no report of it occurring in the carotid space or from IJV in the neck.

Patients present with a slowly increasing painless swelling, symptoms being mostly due to pressure on adjacent structures. Paraneoplastic syndromes like hypoglycaemia, hypophosphataemic osteomalacia, hypertrophic pulmonary osteoarthropathy. Other features like hypertension, congestive cardiac failure are known to occur with these neoplasms.

On ultrasonography, HPC is a well-defined hypoechoic lesion occasionally hyperechoic or anechoic. On CT, it appears as a soft tissue density mass lesion with heterogeneity due to foci of necrosis, haemorrhage and cystic degeneration; with intense heterogeneous enhancement on contrast medium administration; calcification and bone erosion can occur occasionally. On magnetic resonance imaging (MRI), it appears isointense on T1-weighted images and isointense or hyperintense with flow voids within on T2-weighted images. Contrast-enhanced MRI shows heterogeneous enhancement.
However, angiographic findings are characteristic of the lesion: it is hypervascular with dilatation of regional arteries in the arterial phase and later a diffuse capillary blush and dilatation of the draining veins [2]. CT and angiography demonstrate the vascular nature of the tumour, reveal the exact source of the blood supply and demonstrate the size and relationship to adjacent structures [16].

The differential diagnoses considered for this highly vascular mass in the left carotid space were paraganglioma and angiosarcoma. Paraganglioma is a tumour of neural crest origin whose common sites include common carotid artery bifurcation splaying the origins of external and internal carotid arteries, jugular bulb, middle ear (glomus tympanicum) and nodose ganglion of the vagus nerve. It is also a strongly enhancing lesion with early and prolonged vascular staining on angiography. It has a characteristic “salt and pepper” appearance on MR imaging due to the numerous flow voids. Angiosarcoma is a malignant vascular neoplasm that on ultrasonography is hypoechoic or hyperechoic and complex with cystic areas resulting from haemorrhage; on CDS there is arteriovenous shunting. On CT, it appears as a heterogeneous soft tissue mass with marked contrast enhancement.

Pathologically, HPC is a large well-encapsulated tumour consisting of pericytes surrounding branching, thin-walled vascular channels with hyperchromatic nuclei and mild to moderate pleomorphism. It is difficult to classify a lesion as benign or malignant. Increased mitoses (>4 per HPF), increased cellularity, foci of necrosis are suggestive of malignancy [16]. Immunohistochemically HPCs are positive for vimentin and negative for factor VIII and desmin [5].
Local recurrence can occur even many years after removal of the tumour. Metastases are known to occur to the lung, bone, liver, regional lymph nodes and pancreas [3]. Complete excision of the tumour is the definitive treatment. The role of radiotherapy or chemotherapy is not yet well established [17]. Preoperative embolization to reduce the tumour vascularity and preoperative radiotherapy to reduce the tumour size are useful therapies. The prognosis is fair with 5-year survival rate being 50% [16].

HPC should always be considered in the differential diagnosis of any highly vascular tumour anywhere in the body.

REFERENCES


Fig. 3 – Contrast-enhanced CT with reconstruction showing the location of the lesion in relation to internal jugular vein (IJV).