Extra-adrenal retroperitoneal paraganglioma associated with duplication of inferior vena cava

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ABSTRACT

Retroperitoneal paragangliomas arise from specialized neural crest cells symmetrically distributed along the aorta in association with the sympathetic chain. If this tissue aggregates in the adrenal medulla pheochromocytoma may arise. When it remains in the paraaortic sites it could develop into extra-adrenal, retroperitoneal paraganglioma. We report a case of extra-adrenal paraganglioma in the renal hilum intimately related to the left side of a double inferior vena cava. To the best of our knowledge such an association has never been described before. The clinical significance of this venous anomaly is reviewed.

CASE REPORT

Introduction

A 17-year-old boy presented with a two year history of uncontrolled hypertension. He complained of severe headache, sweating and palpitations inspite of being on beta blockers. On examination his blood pressure was 180/120 mmHg. Opthalmoscopy revealed Grade 4 hypertensive retinopathy. There were no masses palpable per abdomen and auscultation did not disclose any bruits. Laboratory studies showed persistently elevated urinary vanillylmandelic acid (VMA) levels (28 mg / 24 hrs Normal: 2 - 10 mg / 24 hrs). An ultrasound ordered to rule out adrenal masses showed a 4 x 4 cm hypoechoic lesion in the left renal hilum. Based on these findings a diagnosis of extra-adrenal paraganglioma was arrived at. A CT, following administration of oral and intravenous contrast agents, done as a part of the pre operative evaluation showed a heterogeneously enhancing mass lesion in the left renal hilum (Figure 1). It also showed a round structure in addition to the inferior vena cava (IVC) and aorta located anterior to the vertebral bodies (Figure 2). The structure was identified both in the slice immediately cephalad and in a number of images more caudad. As this appearance could represent either paraaortic adenopathy or left sided anomalous venous drainage a colour Doppler study was performed. The demonstration of flow confirmed suspicions that the structure was a duplicated IVC. The left common iliac vein drained into the anomalous vein. There were no anastamotic vessels between the right sided IVC and the aberrant vessel. At the level of the renal vessels the tumour abutted the left sided IVC with no demonstrable cleavage between these two structures. The surgeons were cautioned about this anomaly and a vascular surgeon was included in the surgical team. At surgery a highly vascular tumour located at the junction between the renal vessels and the anomalous vein was excised. His convalescence was uneventful.

DISCUSSION

Paragangliomas can occur anywhere from the base of the brain to the urinary bladder. The common locations for extra-adrenal paragangliomas include the organ of Zuckerkandl, bladder wall, retroperitoneum, heart, mediastinum, carotid bodies and glomus jugulare bodies (1,2). The majority of paragangliomas present in the head and neck region are

nonfunctioning tumours of the parasympathetic system, while those below the neck are frequently functional and associated with the sympathetic system (1).

Extraadrenal paragangliomas affect patients in the 2nd or 3rd decade of life.It has been reported that 10%–40% of extra-adrenal paragangliomas are malignant (3). Distant metastasis is the only reliable criteria for confirming malignancy. Local tissue invasion or pathological evidence of nuclear pleomorphism or mitotic activity does not necessarily imply malignancy (1).

Functioning extra-adrenal paragangliomas represent more than 10% of all pheochromocytomas (4).

Duplication of the inferior vena cava is uncommon and has an incidence of 0.2 to 3% (5). The formation of the IVC involves development, regression and anastomosis of three pairs of posterior cardinal, subcardinal, and supracardinal veins (6). The normal IVC is composed of four segments: hepatic segment forming from the vitelline vein, suprarenal segment developing from the right subcardinal vein, renal segment from the right suprasubcardinal anastomosis, and infrarenal segment from the right supracardinal vein.

There are six major anomalies of the inferior vena cava which include duplication of the IVC, transposition of the IVC, azygos continuation of the IVC, circumaortic renal collar, retroaortic renal veins and retrocaval ureter (6). The persistence of right and left supracardinal veins results in double IVC. Transposition of IVC results from regression of the right supracardinal vein with persistence of the left supracardinal vein. Azygos continuation of the IVC is due to failure to form the right subcardinal–hepatic anastomosis, with resulting atrophy of the right subcardinal vein. Consequently, blood is shunted from the suprasubcardinal anastomosis through the retrocrural azygos vein. A circumaortic left renal vein results from persistence of the dorsal limb of the embryonic left renal vein and of the dorsal arch of the renal collar. A retroaortic renal vein develops from the persistence of the dorsal venous anastomosis of the supracardinal and subcardinal veins with regression of the ventral aspect of the two venous systems. A retrocaval ureter occurs when the right posterior cardinal vein persists and the right supracardinal system fails to develop.

The presence of duplicated vena cava can complicate retroperitoneal surgery. Cases of renal cell carcinoma associated with this anomaly have been described (7). Duplicated IVC can be mistaken for paraaortic lymphadenopathy during CT evaluation. Erroneous interpretation of the CT appearance of a duplicated IVC in patients with cancer has resulted in overstaging (8). Colour Doppler demonstrates flow in venous anomalies and maybe utilised to confirm the diagnosis. During retroperitoneal surgery a duplicated inferior vena cava can be inadvertently ligated due to the mistaken assumption that the vessel is a lumbar vein or a variant of the internal spermatic vein (7). Renal vein sampling to investigate secondary hypertension can be misleading if renal vein anomalies are not determined prior to procedure (7).

TEACHING POINT

In conclusion congenital anomalies of the inferior vena cava are uncommon, but can pose important clinical implications for surgeons contemplating retroperitoneal surgery. Clinicians as well as radiologists must be aware of the presence of such anomalies and should be able to distinguish these variants from pathological processes like paraaortic lymphadenopathy. Complications may arise if these anomalies are unexpectedly encountered during surgery. Cross-sectional imaging can provide an accurate anatomical definition thereby allowing an appropriate pre operative evaluation.

ABBREVIATIONS

VMA = vanillylmandelic acid
IVC = inferior vena cava
CT = computed tomography

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FIGURES

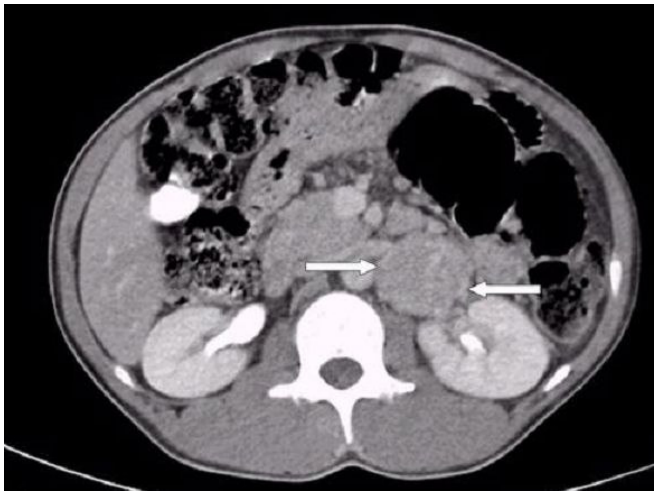

Figure 1: Contrast enhanced CT image in the portal venous phase showing an heterogeneously enhancing mass in the left renal hilum measuring ~2 x 2 cm (white arrows).

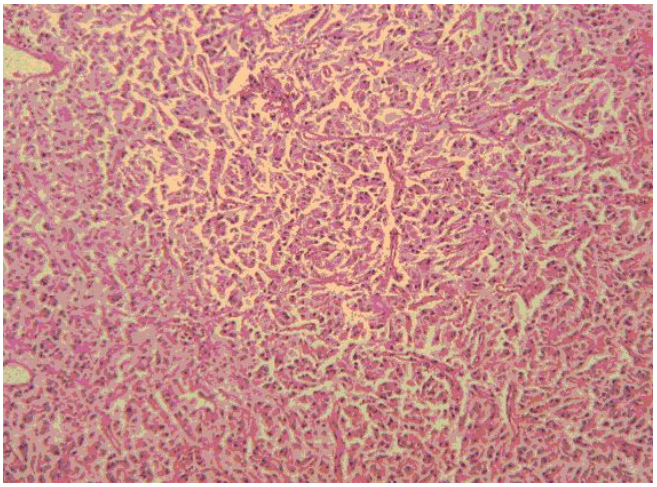

Figure 3: Microscopic histopathological slide showing tumour cells with abundant granular eosinophilic cytoplasm with round nuclei and finely granular chromatin with focal anisokaryosis [H&E,original magnificationx 100). Characteristic nesting pattern of cells-Zellballen pattern

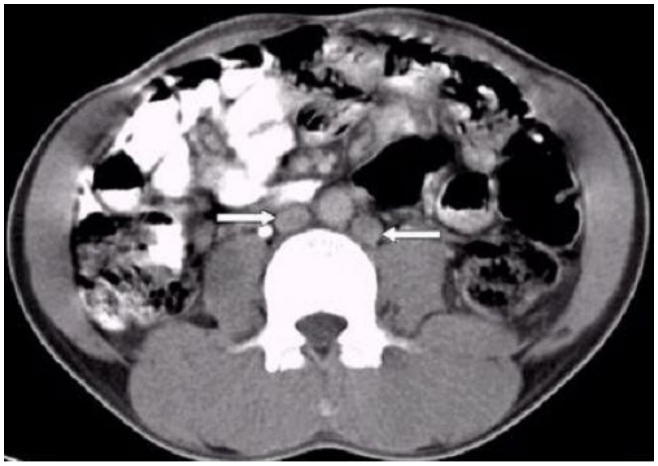

Figure 2: Contrast enhanced CT image in the portal venous phase showing two venous channels adjacent the aorta-duplication of inferior vena cava (white arrows).

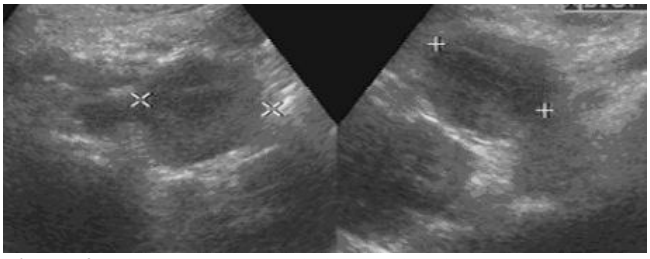

Figure 4: Ultrasound grayscale image showing a well defined hypoechoeic mass in the left renal hilum measuring ~2 x 2 cm.

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