ABDOMINAL RETROPERITONEAL PARAGANGLIOMA WITH PREAORTIC VASCULAR INVOLVEMENT; A CASE REPORT

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ABSTRACT:
Retroperitoneal paragangliomas are a rare clinical entity. Literature regarding the encasement of celiac axis and superior mesenteric artery by these tumours is scarce. In this case report we discuss about a 13 year old boy who underwent successful removal of a functional retroperitoneal paraganglioma with preaortic vascular encasement.

Keywords: Paraganglioma; Retroperitoneal; Pheochromocytoma; Celiac Axis; Superior Mesenteric Artery.

INTRODUCTION
Paragangliomas are neuroendocrine tumours arising from chomaffin cells of sympathetic and parasympathetic ganglia outside adrenal glands. They can be found from below the base of the skull to the pelvic floor. Retroperitoneal paragangliomas are rare and they originate from visceral autonomic pre aortic ganglia and ganglia of sympathetic chain. Only few case reports describe their involvement of celiac axis and superior mesenteric artery (¹,²).

CASE PRESENTATION
A 13 year old boy presented with epigastric pain and was found to have concentric left ventricular hypertrophy. About one year subsequently he was found to be hypertensive with no other examination abnormalities. His thyroid functions and 24 hour urinary matanephrines (136µg/day) were normal. A large para-aortic soft tissue mass was revealed in the ultra sound scan. The germ cell tumour markers (β-hCG, α fetoprotein, LDH) were within normal limits. Further investigation with contrast enhanced CT scan of the abdomen demonstrated a lobulated retroperitoneal soft tissue mass in the upper para-aortic region with no space occupying lesions in adrenal glands (Fig1).
CT angiogram demonstrated that the lesion to be a highly vascular and was encircling celiac axis and superior mesenteric vessels with an associated superior mesenteric artery aneurysm (fig2). Preoperative blood pressure control was achieved with prazocin and three weeks later atenolol was added. Patient underwent surgery under general anaesthesia and epidural block. Intraoperative blood pressure fluctuations were controlled with IV labetolol and phentolamine.

Mercedes Benz incision was made and left lobe of the liver was mobilized. Gastrohepatic omentum was divided to access the celiac axis and lower extent of the tumour was dissected by dividing gastrocolic omentum. An en
Block resection of the tumour was done. Patient had uneventful post operative period with blood pressure returning to normal without drugs. The resected specimen (60×65×38mm) was compatible with a paraganglioma (neuron specific enolase and chromogranin highly positive). The margins were focally involved (Figure 3). KI 67 index was 18%. A MIBG scan was planned for follow up. Repeat contrast CT scan of the abdomen performed in 5 months after the surgery revealed no recurrent lesions.

**DISCUSSION**

Paragangliomas are rare with estimated incidence of 2-8 per × 10⁶ people per year. Retroperitoneal paragangliomas account for up to 85% of such cases. In literature although pelvic, pancreatic and mesenteric abdominal paragangliomas reported (3, 4), vascular involvement confined to about four case reports.

Figure 3 - Specimen

About 40-70% of retroperitoneal paragangliomas are functional and secretes catecholamines. Compared with pheochromocytoma a high proportion of them are malignant (10% vs. 30-50%). Most are sporadic but about 10–50% are associated with inherited syndromes. Hereditary paragangliomas, MEN syndrome type II, Von Hippel-Lindau disease and neurofibromatosis type I represent associated hereditary syndromes. Genetic mutations involving succinate dehydrogenase B and D subunits, RET, VHL and NF I genes are associated with these tumours. Although our patient did not have neurocutaneous manifestations, palpable neck mass or hypercalcaemia to suggest genetic syndromes his age render genetic testing valuable.

The classic triad of episodic headache, sweating, and tachycardia is a presentation of catecholamine secreting functional tumours. Non secretory tumours without suspicious features often present at an advanced stage. Some tumours have presented with chest pain but epigastric pain is a very rare presentation. Urinary metanephrines are highly specific, but intermittent secretion and low levels of secretion may cause normal values even in some large lesions as seen in our patient. Plasma catecholamines have poor sensitivity in diagnosis. Chromogranin A levels are not specific but it is an important prognostic indicator.

CT scan and MRI has sensitivity of 93-100% in localizing adrenal tumours and for extra-adrenal lesions it is about 90%. MRI is more sensitive than CT for paragangliomas.¹²³ I MIBG is the choice of investigation which has a sensitivity of about 80-95% with superior specificity than CT or MRI. In¹¹¹ Octreotide, FDG PET/CT may be valuable imaging options but not routinely recommended. All these isotope studies are invaluable in detecting metastatic disease.

Surgery is often not straightforward as in our case and technically demanding but remains the mainstay of treatment. Preoperative α blockade (to reduce blood pressure) followed by β blockade (to control tachycardia) and hydration (to circumvent vasoexpansion upon tumour resection) are important steps in perioperative blood pressure control. Metyrosine (alpha-methyl-para-tyrosine), which inhibits catecholamine synthesis may provide smoother perioperative course than those given by phenoxybenzamine alone.
External beam radiotherapy and sequential I131 MIBG has achieved about 70% tumour regression in some studies. Other local therapies like radiofrequency ablation, ethanol ablation can be safely performed for metastatic disease but all can induce a hypertensive crisis. Malignancy is seen in about 20-40% of cases (2-10% in adrenals) but its definition is difficult. Clinical, histopathological, and biochemical features are taken into account to predict the biological behaviour. KI 67 index has a prominent value in this assessment. For patients with metastatic disease whose tumours secretes catecholamines and take up MIBG, $^{131}$I-MIBG has shown therapeutic benefit in terms of palliation. Octreotide, Cytotoxic chemotherapy and target therapy with sunitinib are the other available options.

REFERENCES