

Laparoscopic Excision of Functional Ganglioneuroma Near Renal Hilum

PRK Bhargav, V Amar, SK Uday

ABSTRACT

Ganglioneuromas are one of the frequent retroperitoneal tumors arising from extraadrenal sympathetic ganglia or chromaffin tissue. The commonest extraadrenal site is periadrenal region near renal hilum. We present a case of ganglioneuroma in a narrow quadrangular space near right renal hilum, treated laparoscopically.

Keywords: Ganglioneuroma, Renal hilum, Laparoscopic excision.

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INTRODUCTION

Paragangliomas and ganglioneuromas can arise from extraadrenal sympathetic ganglia or chromaffin tissue of retroperitoneum.¹ Surgery either with open or laparoscopic approach is the treatment of choice for these tumors. We describe a case of functional ganglioneuroma arising from retroperitoneum in a precarious areas near right renal hilum managed by successful laparoscopic excision.

CASE REPORT

A 28-year-old lady presented with nonradiating pain in right loin since 3 months. There was no history of gastrointestinal disturbance or urinary complaints. She was detected to have episodic hypertension with maximum blood pressure of 170/110 mm Hg. There was no history of classic triad of headache, sweating and palpitations, which is suggestive of pheochromocytoma. There was no family history or stigmata of multiple endocrine neoplasia or familial paraganglioma syndromes. Computerized axial tomogram (CT scan) showed a 4 × 4 cm heteroechoic mass in the right adrenal region close to the renal hilar vasculature (Fig. 1). The patient and first-degree relatives were clinically screened for syndromic associations, such as MEN syndrome, neurofibromatosis, paraganglioma syndrome, Von-Hippel Lindau disease, Struge-Weber syndrome and tuberous sclerosis, but no suggestive stigmata was found. Genetic testing of MEN 2 syndrome was normal. There was no thyromegaly or thyroid nodularity on sonography. According to our departmental protocol, other causes of

secondary hypertension, such as renal, vascular, neurological, drug abuse were excluded by clinical evaluation and relevant investigations. Twenty-four hours urinary total metanephrine level was 1.2 mg (0.7–0.95), plasma metanephrine level was 29 pg/ml (<80) and 24 hours urinary vanillylmandelic acid level (VMA) was 11.4 mg (<8 mg). Serum calcium, phosphorus and alkaline phosphatase were within normal limits. The latter was particularly suggestive of a functional chromaffin tumor. In view of secondary hypertension in a young lady with evidence of tumor in right adrenal region a diagnosis of pheochromocytoma or neurochromaffin tumor was considered, though metanephrine levels were marginally elevated. Preoperative preparation was done with titrated escalating doses of prazosin (maximum = 8 mg/d), fluid and high salt intake.

Operative details: After 9 days of preparation, she was taken up for surgery. Approach was transperitoneal laparoscopic with four ports in right hypochondrium and lumbar regions. Contrary to our first diagnosis of adrenal originated pheochromocytoma, we found a highly vascular tumor being unrelated to the adrenal gland. It was located in the retroperitoneum in a narrow quadrangular space flanked by renal hilum, ureter on right, renal vessels superiorly, inferior vena cava on left side and right colic flexure inferiorly (Fig. 2). Schematic diagram of the same laparoscopic view is depicted in Figure 3. With gentle dissection using bipolar diathermy and hydrodissection with irrigation/suction

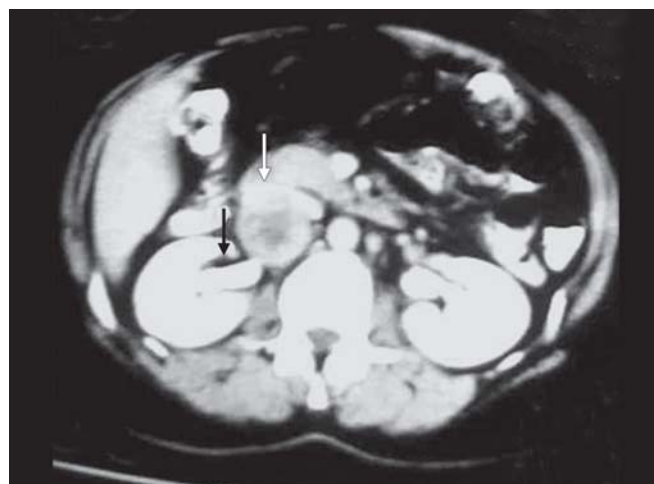


Fig. 1: Computerized tomogram of abdomen—heterogeneous mass (white arrow) in adrenal area abutting renal vasculature (black arrow)

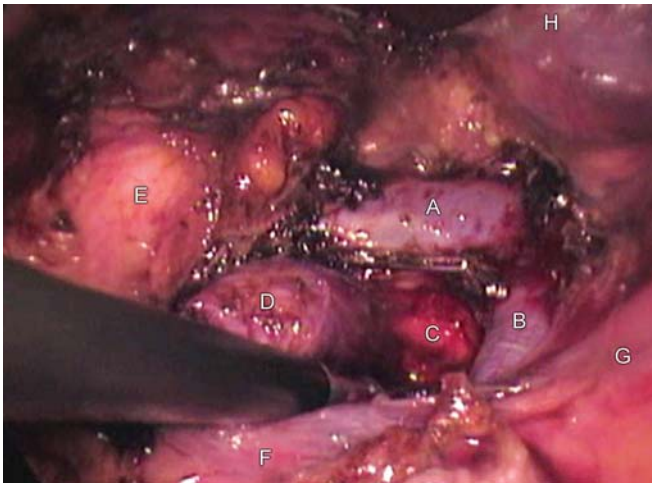


Fig. 2: Intraoperative view of tumor: (A) right renal vein; (B) inferior vena cava; (C) renal artery; (D) tumor; (E) right kidney; (F) hepatic colic flexure; (G) duodenum; (H) liver

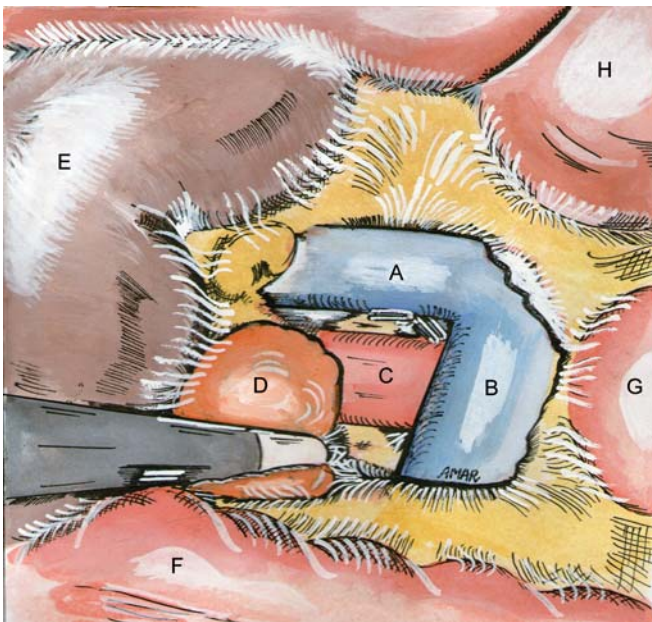


Fig. 3: Schematic diagram of structures shown in Figure 2

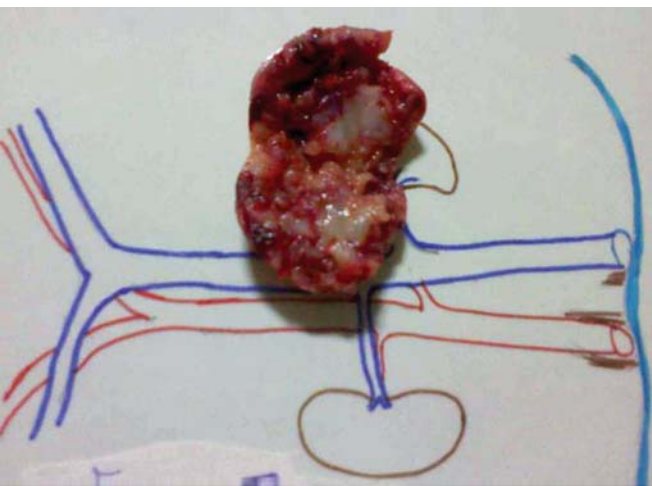


Fig. 4: Cut section of *ex vivo* specimen of tumor on grossing chart

cannula, intact tumor without any capsular breach was extirpated without injury to adjacent vital structures. The extirpated *ex vivo* specimen is shown in Figure 4. Postoperative period was uneventful with mild hypertension of 136/90 mm Hg and patient was put on angiotensin inhibitor. Histopathology of the tumor was suggestive of a benign ganglioneuroma with schwannian stroma consisting of Antoni A areas intermixed with mature, few immature ganglion cells and chromaffin cells (Fig. 5). Immunohistochemistry demonstrated reactivity for neuron-specific enolase and chromogranin A. On regular follow-up, she became normotensive at 5 months follow-up.

DISCUSSION

Neurochromaffin tumors, such as paragangliomas and ganglioneuromas usually arise from lumbar or thoracic sympathetic ganglia and rarely in the neck and skull base.² Occasionally, mixed ganglioneuromapheochromocytoma may occur suggesting their common origin from neural crest of embryonic neurectoderm.² Ganglioneuromas can be associated with disturbance in catecholamine metabolism and secretion of dopamine, VMA in contrast to metanephrine or normetanephrine as in pheochromocytoma.³ Serum chromogranin A is a nonspecific marker for neuroendocrine tumors, which is especially useful in a unknown tumor pathology to predict its nature in the setting of normal levels of specific markers (in this case metanephrines/VMA). We evaluate chromogranin A level in a case of neuroendocrine tumor of unknown origin in the absence of elevated specific markers or in the follow-up monitoring after treatment of neuroendocrine tumor. As seen in this case, the tumor had a predominantly neuronal differentiation with few chromaffin elements as confirmed by histopathology and immunohistochemistry.

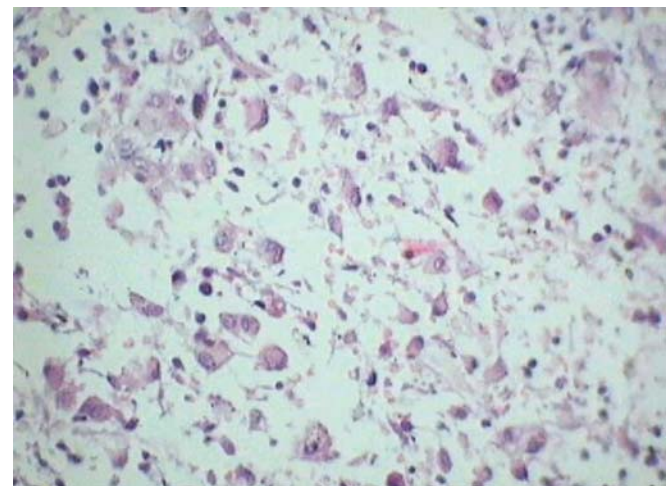


Fig. 5: Microphotograph of the tumor showing combination of ganglion cells (predominantly) and chromaffin elements interspersed in a scanty neuronal stroma

The commonest location of EAP and extraadrenal ganglioneuroma (EAGN) is perirenal area and renal hilum is one of the most frequent site of these tumors.¹ Sometimes, the differentiation between adrenal and extraadrenal masses in the vicinity is a difficult task. Though, the present thin slice computed tomography (CT) scan with reconstructive protocols offer a virtual intra-abdominal topography of the retroperitoneal tumors, especially in three-dimensional and coronal sections.⁴ But, we have not performed any CT scan reconstruction of the tumor, which has resulted in our broader differential diagnosis. The role of preoperative MIBG scintigraphy is a debatable issue with both supportive and opposing data. In our department, we believe in postoperative MIBG scan and when planning MIBG therapy, because of its poor sensitivity^{5,6} for smaller tumor sites, metastatic deposits and familial paragangliomas in the preoperative setting, which are masked by the primary tumor mass. Moreover, latest thin slice CT protocols serve the purpose better than preoperative MIBG scan. Their clinical presentation ranges from asymptomatic incidentaloma to abdominal lump or functional tumors. Functional neurogenic or chromaffin tumors are associated with hypertension secondary to hormonal hypersecretion or renal vascular compression. Surgery is usually done via open approach, though technical advances and surgical expertise allows laparoscopic approach. The laparoscopic excision of retroperitoneal ganglioneuromas can be arduous task due to precarious proximity to great vessels, often with adhesions, direct vascular supply or encasement of blood vessels.⁷⁻⁹ There have been few case reports and short series of laparoscopic excision of retroperitoneal tumors and EAGN, establishing their laparoscopic feasibility.^{8,10} There is a case report laparoscopic excision of ganglioneuroma using vessel seal device to separate it from posterior inferior vena cava.⁷ This case was done with successful use of electrocautery, hydrodissection without the use of harmonic shears, in spite of proximity of vital structures and direct vascular supply. With conventional CT scan, caution needs to be exercised in differentiating between renal and extrarenal masses in the perirenal region, though reconstructive CT scan with coronal sections can serve this purpose better.⁴ This case further highlights the feasibility of laparoscopic excision of EAP, even in the precarious area of renal hilum in experienced hands.

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