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Extra-adrenal retroperitoneal paraganglioma: report of a rare case

Abstract

Extra- adrenal retroperitoneal paragangliomas are extremely rare neuroendocrine neoplasms with an incidence of 2-8per million. They arise from embryonic neural crest cells and are composed mainly of chromaffin cells located in the para-aortic sympathetic chain. They synthesize, store and secrete catecholamines because of which they may present with headache, sweating, palpitation and symptoms of hypertension.1 On the other hand, they may remain silent and non- functional and present with vague symptoms like pain abdomen due to episodic release of catecholamines.

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Introduction

A pheochromocytoma (adrenaline-producing tumor) that grows outside of the adrenal glands is known as extra-adrenal paragangliomas, or extra-adrenal pheochromocytomas. They are closely related to pheochromocytomas. While pheochromocytomas originate in the core of the adrenal glands, extra-adrenal paraganglioma tumors originate in the ganglia specifically the paraganglia, of the sympathetic nervous system. Adrenal glands are located just above each kidney and are part of the endocrine system. One function of the adrenal glands is to produce hormones called catecholamines, such as epinephrine and norepinephrine. The sympathetic nervous system's main function is to control the organs of the body. Ganglia is a tissue mass of nerve cells. The paraganglia is a small group of chromaphil cells in the abdomen. In 95percent of cases, extra-adrenal paragangliomas are located in the abdomen regions, but they can also occur in the head, neck, and thoracic regions. They tend to be malignant in more cases than pheochromocytomas. This case report aimed at analyzing the clinical presentation, diagnosis and treatment outcomes.

Case report

A 55-year-old female patient presented to surgical department with complaints of pain abdomen mass in the left hypochondrium since 5 years along with history of headache off and on. The patient was a known hypertensive and was on medication. General physical examination did not reveal any significant abnormality. Per abdominal examination revealed tenderness and a mass in the left hypochondriac region which did not move with respiration. Bowel sounds and rectal examination were normal. Her routine hematological and biochemical parameters including serum calcium were within normal limits.

Ultrasonography showed a hypoechoic, round retroperitoneal mass suspected to be of mesenchymal origin measuring approx. 6.9x5.9cm in left paraaortic region anterior to left kidney with multiple anechoic cystic areas seen within the mass. CECT abdomen and pelvis was performed subsequently. There is evidence of large well marginated centrally necrotic enhancing mass situated left anterior to abdominal aorta starting from the level of SMA extending to the level of lower pole kidney showing distinct planes with aorta, left ureter and SMA.

Laparotomy was undertaken for exploration by an upper midline incision. Per-operatively, a retroperitoneal mass was found inferior to the renal vein on the left side of aorta occupying 12th thoracic, 1st and 2nd lumbar vertebral levels. The tumor was situated left to abdominal aorta starting from the level of SMA extending to the level of lower pole kidney showing distinct planes with aorta, left ureter and SMA. Many direct arterial connections were seen supplying the mass from the aorta. The mass was found located anterior to the sympathetic and lymphatic chain. Intra-op BP varied from 220/120 to 100/50 due to catecholamine surge, which was managed successfully by anesthesia team. Meticulous hemostsis was achieved. The mass was completely respected. There were no significant perioperative and postoperative complications.

Discussion

Paragangliomas can develop anywhere along the midline of the retroperitoneum. The exact incidence of retroperitoneal paragangliomas is unknown, although males are typically affected more frequently than Females. In addition, most patients are diagnosed between 30 and 45years of age.[1] Clinically, patients with a retroperitoneal paraganglioma often present with back pain or a palpable mass.[2] Conventional treatment for paragangliomas typically involves complete surgical excision, while surgical debulking is considered a mainstay of palliative therapy for malignant paragangliomas. In some cases, complete excision is difficult due to the highly vascular nature of paragangliomas and their proximity to major blood vessels (Figure 1-4).

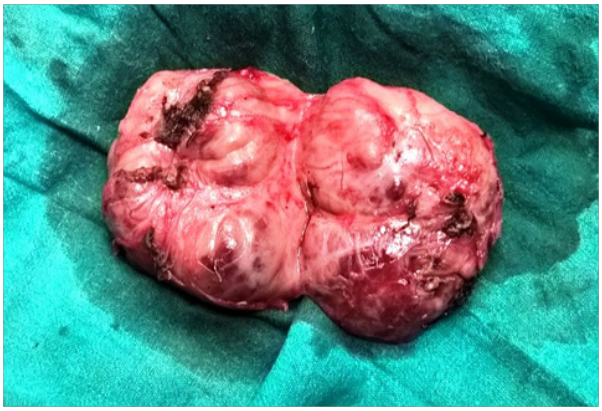

Figure 1 Gross specimen of excised tumor.

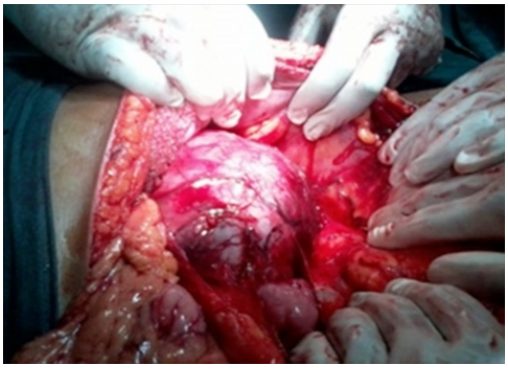

Figure 2 Intraoperative photograph of tumor *in-situ.*

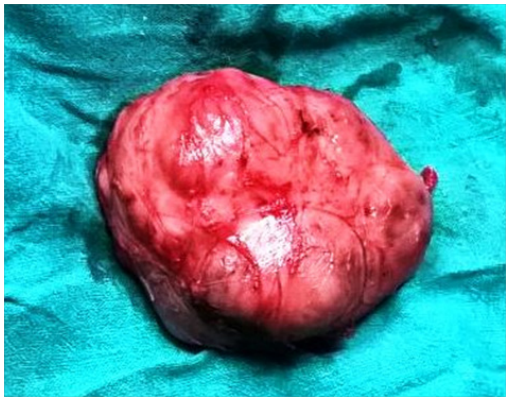

Figure 3 Excised tumor.

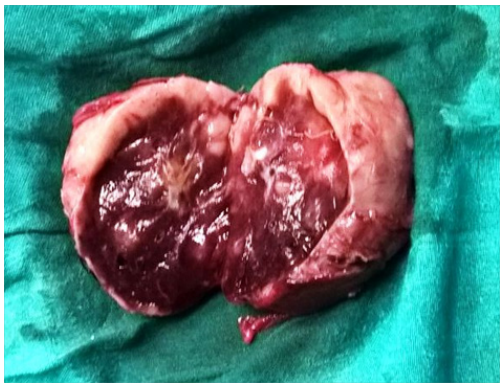

Figure 4 Cut section of the tumor.

Abdominal paragangliomas are mostly retroperitoneal in location, accounting for 85% of all extra adrenal paragangliomas. The most common site for retroperitoneal paragangliomas is between the origin of inferior mesenteric artery and the aortic bifurcation known as organ of Zuckerkandl. Paragangliomas arising from jugulotympanic body are called chemodectomas, whereas paragangliomas originating from the carotid body are known as carotid body tumors. Paragangliomas located in the second part of duodenum are called gangliocytic paraganglioma.[3] Functional paragangliomas can be diagnosed based on presentation and subsequent laboratory investigation revealing elevated catecholamines and their metabolites in the blood and urine. Nonfunctional paragangliomas are mostly found incidentally or present as a mass with symptoms of surrounding organ compression. On CT scan these tumors appear as soft-tissue masses with either homogenous enhancement or central areas of low attenuation. It appears as highly vascular structure with areas of intralesional hemorrhage and necrosis.[4] Patient with metastatic disease will require adjuvant radiotherapy while chemotherapy is restricted to patients not accessible for surgery and resistant to radionuclide therapy.[1] Because of malignant potential and higher recurrence rate in paragangliomas, lifelong follow up is usually recommended.[5–10]

Acknowledgements

None.

Conflict of interest

Author declares that there is no conflict of interest.

References

1. Andersen KF, Altaf R, Krarup-Hansen A. Malignant pheochromocytomas and paragangliomas the importance of a multidisciplinary approach. *Cancer Treat Rev.* 2011;37(2):111–119.
2. Moslemi MK, Abolhasani M, Vafaeimanesh J. Malignant abdominal paraganglioma presenting as a giant intra-peritoneal mass. *Int J Surg Case Reports.* 2012;3(11):537–540.
3. Disick GIS, Palese MA. Extraadrenal pheochromocytoma:50 diagnosis and management. *Curr Urol Reports*. 2007;8(1):83– 88.
4. Sangster G, Do D, Previgliano C, et al. Primary retroperitoneal paraganglioma simulating a pancreatic mass: a case report and review of the literature. *HPB Surg.* 2010;4p.
5. Shah U, Giubellino A, Pacak K. Pheochromocytoma: complications in tumorigenesis and the actual management. *Minerva Endocrinol.* 2012;37(2):141–156.
6. Sclafani LM, Woodruff JM, Brennan MF. Extraadrenal retroperitoneal paragangliomas: natural history and response to treatment. *Surgery*. 1990;108:1124–1130.
7. Lack EE, Cubilla AL, Woodruff JM, et al. Extra-adrenal paragangliomas of the retroperitoneum: a clinicopathologic study of 12 tumours. *Am J Surg Pathol*. 1980;4:109–120.
8. Arrabal-Polo MA, Arrabal-Martin M, Lopez-Leon VM. Spontaneous retroperitoneal abscess as the first clinical manifestation of a nonfunctioning retroperitoneal paraganglioma. *Ann R Coll Surg Eng.* 2010;92(3):W17–W19.
9. Lee KY, Oh YW, Noh HJ. Extraadrenal aragangliomas of the body: imaging features. *Am J Roentgenol*. 2006;187(2):492–504.
10. Van Hulsteijn LT, Dekkers OM, Hes FJ, et al. Risk of malignant paraganglioma in SDHB-mutation and DHDmutation carriers: a systematic review and meta-analysis. *J Med Genet.* 2012;49(12):768–776.