Case Report

Non-functional retroperitoneal paraganglioma: A case report

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Abstract

Paragangliomas are extra-adrenal tumors of the autonomic nervous system and may be found within the skull base, neck, chest, and abdomen. When presenting within the abdominal cavity, they may arise as a primary retroperitoneal neoplasm and can mimic vascular malformations or other conditions related to specific retroperitoneal organs such as the pancreas, kidneys.[1] They synthesize, store, and secrete catecholamines because of which they may present with headache, sweating, palpitation, and symptoms of hypertension (functional).[2] In the absence of histological diagnosis and symptoms of catecholamine excess (non-functional), these may be mistaken for GISTs.[3] We are reporting a case of a 36-year-old female who was clinically diagnosed as GIST, underwent excision, and postoperative histopathological examination was found to be paraganglioma.

Keywords: Catecholamine excess, non-functional paraganglioma, retroperitoneal GIST

Introduction

Paragangliomas (also known as extra-adrenal pheochromocytomas) are rare neuroendocrine neoplasms which are derived from paraganglia, a diffuse neuroendocrine system dispersed from the skull base to the pelvic floor, and these tumors are observed in patients of all ages. Some of the tumors (named as functional paragangliomas) have been discovered to originate, synthesize, store and secrete catecholamines, which leads to elevated levels of urine/serum catecholamines and the typical clinical symptoms such as episodic headache (72%), sweating (69%), and palpitations (51%).[4] Approximately 10-15% of such tumors is non-functional. They are often locally invasive and associated with a high incidence of local recurrence. Non-functional paragangliomas pose a significant diagnostic challenge.[5] On abdominal CT, there are no unique imaging characteristics specific for paragangliomas. Consequently, these tumors may be mistaken for other primary epithelial or mesenchymal abdominal tumors.[3]

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Case Presentation

A 36-year-old female, presented with intermittent pain over epigastrium for 3 months. Her symptoms started after undergoing cholecystectomy. Occasional dyspepsia and nausea present. No history of vomiting, altered bowel habits. The pain is dull aching in nature, occurs anytime during the day, and not related to food intake or position. It is relieved with regular analgesics. She has no any comorbid illness. The physical examination was unremarkable with no palpable abdominal mass present. She has undergone open cholecystectomy in April 2018. Her post-operative recovery was uneventful and the gall bladder biopsy was Chronic cholecystitis. CECT abdomen revealed a non-enhancing mass of 7cm × 4cm just inferior to the pancreas. The mass was in contact with the pancreas but not infiltrating it. There was no lymphadenopathy [Figure 1]. A clinical diagnosis of GIST (Gastro-Intestinal Stromal Tumor) was made and decision taken to excise the mass. Surgery was done on 14/8/18. The abdomen was opened by a midline incision and the entire mass was excised completely. The mass was located inferior to the pancreas, medial to the duodenojejunal flexure. The mass not invading into any surrounding structure [Figure 2]. The patient

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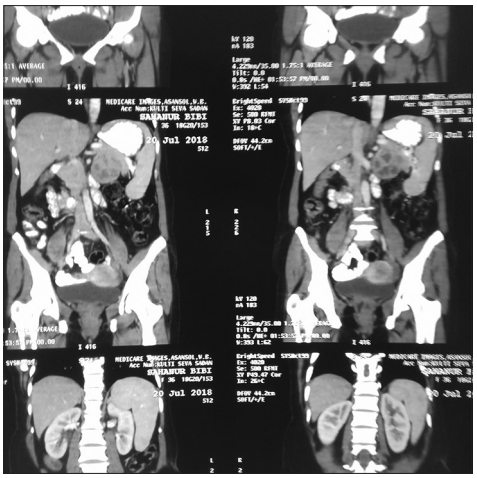

Figure 1: CECT abdomen report—There is a well-defined SOL in left side with regular outline located between inferior border of Pancreas and the DJ flexure. It is abutting inferior border of body of stomach without any obvious sign of its involvement. The lesion shows heterogenous enhancement on contrast with a few cystic/necrotic areas. No calcification or fatty component is seen. Superior mesenteric vessels appear normal and away from the SOL. Size of the lesion is 7 × 4 cm

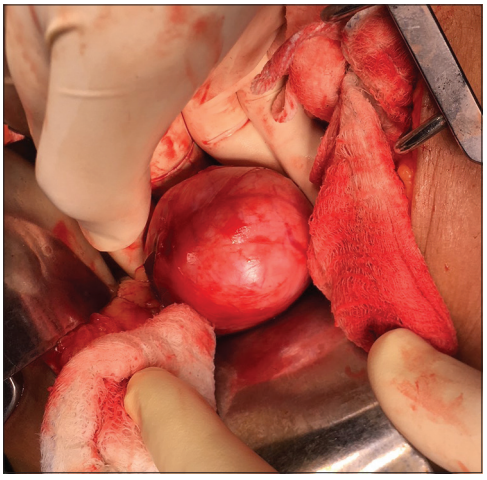

Figure 2: Intraoperative picture

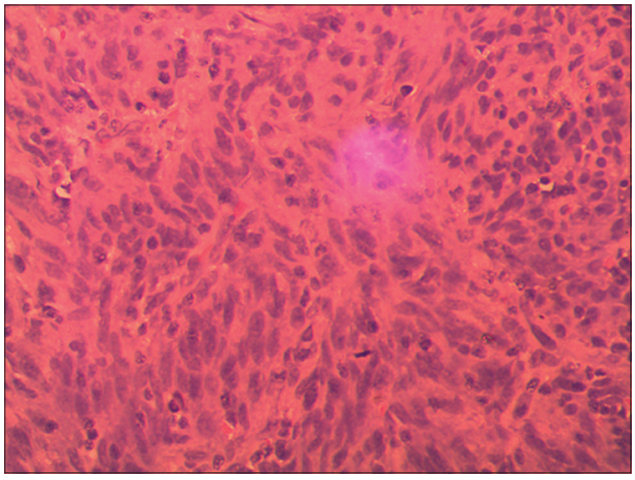

Figure 3: Histopathological slide

had complete and an uneventful recovery and was discharged from the hospital on 18/8/18. The histopathology of the mass was a Paraganglioma. [Figure 3]. The patient was advised urinary VMA and metanephrines post-operatively which were within normal values. Postoperatively patient is undergoing follow up with CECT abdomen every 3 months for the first year.

Discussion

Paragangliomas are rare tumors arising from sympathetic and parasympathetic paraganglia, which are derived from the neural crest cells. Extra-adrenal paragangliomas are found arising from chromaffin tissue along the autonomic nervous system and can thus be found in the head, neck, thorax, abdomen, and pelvis.[5] Retroperitoneal paragangliomas are rare tumors. Non-functioning retroperitoneal forms are even more rare and are most often isolated. They are characterized by their asymptomatic profile and normal levels catecholamines in the urine and blood.[6] The median age of diagnosis for retroperitoneal paragangliomas is 37-43 years and the incidence is similar between men and women.[5] Nevertheless, patients will sometimes present with non-specific symptoms such as lower back pain, abdominal heaviness, urinary symptoms, or changes in their general condition. Computed tomography scans typically show a solid round or oval mass that is homogenous but that may have a cystic or necrotic appearance at its centre, or it may appear calcified. A definitive diagnosis can be reached only by histology. Histologically paragangliomas are diagnosed by their highly vascular appearance, with chief cells and sustenacular cells arranged in clusters called zellballen. Chief cells are often positive for neuroendocrine markers (synaptophysin, NSE, chromogranin) on immunohistochemistry, while sustenacular cells are positive for S-100 protein.[5] However, total excision is the basis of curative treatment because these tumors are potentially malignant. Only the appearance of distant metastases can confirm that the tumor is cancerous.[6] Sites of metastasis include lymph nodes, bone, liver, and lung.[5] Follow-up using scintigraphy with metaiodobenzyl-guanidine enables the detection of metastases or recurrence.[6,7] In our case, since we achieved complete excision of the mass (R0), we are following up the patient with CECT abdomen every 3 months in the first year.[8]

Conclusion

Non-functional retroperitoneal paragangliomas are rare group of tumors which are difficult to diagnose, owing to their silent and asymptomatic behavior, often misdiagnosed as GISTs. Our case emphasizes the importance of including paragangliomas in differential diagnosis of retroperitoneal tumors and the role of histology as only definitive way in diagnosing non-functional retroperitoneal paragangliomas. Surgical excision is the only treatment of choice. As recurrence and metastasis are common, lifelong follow-up is required.

There are numerous causes of upper abdominal pain. While gastritis, pancreatitis, and cholecystitis with gall stones are very common, such uncommon conditions need to be kept in

mind, so as not to miss these diseases. The patient underwent cholecystectomy but her pain persisted, which has got relieved following successful treatment of this paraganglioma.

Consent

Written consent was obtained from the patient to publish this case report.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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