Non-functioning Retroperitoneal Paraganglioma: A Case Report of Rare Intra-abdominal Tumour

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ABSTRACT

Paragangliomas are rare neoplasms arising from the primitive neural crest cell. These are often difficult to diagnose and treat. The aim of this work is to present a case of non-functional paraganglioma which is a rare manifestation; draw attention to diagnostic problems and treatment. We report a case of a 52 year old patient presented with vague abdominal pain and distension of abdomen for two years duration who had a retroperitoneal tumour situated at the aortic bifurcation. Complete resection of the tumour was performed. It has been recorded that 10% of all paragangliomas arise outside the adrenal glands. The histological examination and immune histochemistry of specimen gives the diagnosis of an organ of Zuckerkandl paraganglioma. Non-functioning paraganglioma remain silent without any clinical presentation and suddenly presents with metastasis, which is difficult to treat. The adrenal medullae represent the largest collection of paraganglionic tissue; tumours arises from these gland are much more frequently than in extra-adrenal locations. Our case highlights the importance of including the adrenal external paraganglioma in the differential diagnosis of retroperitoneal tumours.

Keywords: Paraganglioma; Tumours; Organ of Zuckerkandl.

INTRODUCTION

Paragangliomas are rare neuroendocrine tumours arising from a widely dispersed collection of specialized cells of the primitive neural crest.1 The adrenal medullae represent the largest collection of paraganglionic tissue; tumours arises from these gland are much more frequently than in extra-adrenal locations. It has been recorded that 10% of all paragangliomas arise outside the adrenal glands.2 The most common site of extra-adrenal occurrence is intra-abdominal, usually within the para aortic and perinephric spaces.3 Other rarer sites include the thorax, skull base and neck. These tumours may be functional and result in symptoms of excess catecholamine production. 10-15% of such tumours are non-functional, and in about another 10% hormone activity is not manifest clinically. Paragangliomas are associated with a high incidence of recurrence and locally invasive.4 Non-functional paragangliomas pose a significant difficulty in diagnosing. In the absence of typical symptoms of catecholamine excess, the diagnosis of such tumours is often difficult and delayed.5 In this report, the authors describe a new case of non-functional retroperitoneal paraganglioma discovered during investigation of abdominal pain.

Case report and results

A 52 year old gentleman presented with vague abdominal pain and distension of abdomen for two years duration. He didn’t have any other co-morbidity. His blood pressure was 138/80 mmHg and pulse was 84 bpm at the time of admission, however he denied symptoms of catecholamine excess. He was taking no regular medications and no family history of illness. The physical examination was un-remarkable with a palpable abdominal mass of 8x6 cm in the epigastric and umbilical region. It was mobile and retroperitoneal in origin. Abdominal ultra sonography showed a huge expansile mildly lobulated right paramedian solid heteroechocic mass of 16x14x12 cm with internal vascularity evident in subhepatic GB fossa region extending from inferiorly up to right iliac fossa (?GIST), initially thought to represent GIST (Fig-1).

Figure 1: Intraoperative before resection of tumour (16x14x12 cm)

Abdominal computed tomography (CT) showed a solitary well-defined heterogeneous lobulated mass in peritoneal cavity extending from right iliac fossa, right lumbar and sub hepatic region with calcific foci at places and central

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necrotic areas with possibly maintained fat planes with surrounding structure displacing proximal transverse colon anteriorly (?GIST) multiple peritoneal and retroperitoneal lymphadenopathy seen (Fig-2).

There was prominence of vascularity of the neoplastic cells, which were uniform, round to ovoid with vesicular nuclei with few cells showing nuclear enlargement, hyperchromasia and occasional intra nuclear pseudo inclusions. Areas of necrosis, haemorrhage, calcification, hyalinization and dilated congested blood vessels were also present. It was highly cellular, multi-nodular encapsulated tumour with a vascular framework containing many dilated and congested blood vessels. Immunohistochemical staining of the tumour showed that it strongly expressed chromogranin (Fig-5), vimentin and synaptophysin and while they are immune negative for cytokeratin and CD-10, S-100P highlights the sustentacular cells. These findings led to a histological diagnosis of retroperitoneal paraganglioma.

CT chest showed no intra-thoracic malignancy. USG guided FNAC revealed features suggestive of paraganglioma. The lesion was completely resected through a midline laparotomy incision (Fig- 3).

DISCUSSION

The paraganglioma is a rare tumour of the sympathetic and parasympathetic ganglion, which is produced from neural crest cells. Those produced by the adrenal medulla are called pheochromocytomas and contain about 80% of these tumours. The adrenal gland paraganglioma is found in the chrome tissue produced along the autonomic nervous system and can be found in the head, neck, chest, abdomen and pelvis thusbe. The most common site of the sympathetic paraganglioma is in the abdomen, usually located in the Zuckerkandl organ at the bifurcation of the aorta, with the paraganglion first in 1901 characterized by Zuckerkandl characterized by a consistent distribution in human fetuses.6,7 The median age of diagnosis for retroperitoneal paragangliomas is 37-43 years and the incidence is similar between men and women. A large proportion of paragangliomas are related to disease-causing mutations or hereditary syndromes such as neurofibromatosis type 1 (NF1), multiple endocrine neoplasia type 2 (MEN2), the Carney triad and gene mutations of the subunits of succinate dehydrogenase (SDH), Von Hippel-Lindau (VHL) gene mutations.8,9 Functional paragangliomas patient presents with flushing, hypertension, palpitations, tachycardia, anxiety, headache and/or profuse diaphoresis due to raised cate-cholamines level in blood.4,5,10 Those with non-functioning retroperitoneal ganglion tumours may be
incidentally diagnosed or have compression symptoms such as abdominal pain associated with nausea, vomiting, abdominal distension, and weight loss. Non-functional retroperitoneal ganglion tumors have also been reported to have atypical manifestations of chest pain or paralytic ileus. Display of different ranges often makes clinical diagnosis of these tumours difficult. Serum levels of chromogranin A, NSE, or vimentin are elevated with most neuroendocrine tumours and help to differentiate them from non-neuroendocrine tumours. By using hormonal assay in blood or urine; endocrine symptoms, neuroendocrine tumours can be differentiated. Paragangliomas may show central necrosis or haemorrhage, enhancement and calcification. CT and MRI have higher sensitivity than ultrasound however ultrasound may be used as a first-line investigation. Functional imaging using MIBG imaging can be used to better localize extra-adrenal disease or metastatic sites. Angiography may help to reveal vascular invasion or show small metastases. Ultimately, although imaging techniques may be helpful, the diagnosis of paragangliomas can only be confirmed by careful histologic and immunohistochemical evaluation. Because of the malignant potential of the paraganglioma, surgical resection is the preferred management. Resection is usually challenging because these highly vascular tumours are often located near multiple vital vessels. If the tumour is considered unresectable, an attempt to reduce its size using chemotherapy, radiotherapy or embolization may be indicated, as excision provides the only chance of cure. Radiation therapy can also be used for inoperable tumours or for palliative care. The radioactive nucletidates may be specialized therapies for diagnostic scans showing uptake of the tumour. Inoperable paragangliomas can be treated with octreotide. Histologically paragangliomas are diagnosed by their highly vascular appearance, with sustenacular cells and chief cells arranged in clusters called zellballen. Sustenacular cells are positive for S-100 protein, while chief cells are often positive for neuroendocrine markers (synaptophysin, NSE, chromogranin). Malignant tumours cannot be reliably diagnosed histologically and are often diagnosed based on the presence of metastasis. Approximately 20-42% of adenai sympathtic ganlgon tumours metastasize compared to only 2-10% of pheochromocytomas. Metastases include lymph nodes, bone, liver and lung. Metastatic disease prognosis is poor, it was reported that 5-year survival rate was 36%.4

CONCLUSION

Sub-peritoneal paraganglioma is a rare tumour with malignant potential, which causes considerable difficulty in diagnosis and treatment. Our case highlights the importance of including the adrenal external paraganglioma in the differential diagnosis of retroperitoneal tumours. Advances in genetic testing and discovery of new molecular markers are contributing to increased understanding of paragangliomas, however at present there is no way to definitively predict metastatic risk. As recurrence and metastasis are common, lifelong follow-up is required.

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REFERENCES


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