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Primary intra-abdominal paraganglioma: A case report and literature review

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Abstract

BACKGROUND

Paragangliomas are rare neuroendocrine tumors. We hereby report a case of a localized paraganglioma found in the abdominal cavity, and review the relevant literature to improve the understanding of this disease.

CASE SUMMARY

A 29-year-old Chinese female patient was referred to our hospital due to an abdominal mass found on physical examination. Imaging revealed a mass in the left upper abdomen, suggestive of either a benign stromal tumor or an ectopic accessory spleen. Laparoscopic radical resection was subsequently performed, and histopathological analysis confirmed the diagnosis of a paraganglioma. The patient was followed up 3 months post-operation, and reported good recovery with no metastasis.

CONCLUSION

Radical resection can effectively treat intra-abdominal paragangliomas, with few side effects and low recurrence risk. In addition, early and accurate diagnosis and timely intervention are essential for the prognosis of this disease.

INTRODUCTION

Paragangliomas are rare neuroendocrine tumors that can occur at any age. It represents an important cause of secretory hypertension as it is often characterized by the excessive production of catecholamines. This study aimed to report a case of an intra-abdominal paraganglioma, and improve the understanding of this disease by elucidating the clinical features of the patient, while reviewing the relevant literature.

CASE PRESENTATION

Chief complaints

A 29-year-old woman was referred to the Department of General Surgery of the Second Hospital of Jiaxing on July 20, 2022 following findings of an abdominal mass 5 d ago.

History of present illness

Initial abdominal computed tomography (CT) revealed soft tissue nodules with central calcification in the left upper abdomen, which warranted further imaging with contrast-enhanced CT.

History of past illness

The patient had no surgical or tumor history. Besides a 2-year history of Hashimoto's thyroiditis and hyperthyroidism, no other underlying diseases such as hypertension and diabetes were reported.

Personal and family history

The patient denied any family history of cancer.

Physical examination

Physical examination was grossly unremarkable. No gastrointestinal symptoms such as abdominal pain, distension, nausea and vomiting, jaundice, abnormal bowel movements, or abnormal stool forms were noted. Urinary symptoms such as gross

hematuria, frequency, urgency, and dysuria were not noted as well. The patient was not pyrexic, and denied any chest tightness or shortness of breath.

Laboratory examinations

Laboratory results were as follows: Thyroid stimulating hormone, 0.003 μIU/mL; anti-thyroglobulin, 282.92 IU/mL, anti-thyroid peroxidase antibody, 149.91 IU/mL (reference range, 0–34 IU/mL); and CA19-9, 2.0 U/mL (reference range 0–37 U/m).The remaining laboratory test results were otherwise unremarkable.

Pathological examination of the lesion suggested a paraganglioma (Figure 1). Immunohistochemical staining was subsequently performed, which demonstrated the following: Syn (-), CgA (-), CD56 (-), Ki-67 (+ 5%), CD10 (-), S-100 (+), NES (+), AE1/AE3 (-), SOX10 (-), and P53 (wild type). A diagnosis of an intra-abdominal paraganglioma was thus confirmed.

Imaging examinations

Contrast-enhanced CT confirmed a mass in the left upper abdomen. At this stage, a benign stromal tumor of mesenchymal origin or an ectopic accessory spleen was considered (Figure 2).

FINAL DIAGNOSIS

The patient was diagnosed with a primary intra-abdominal paraganglioma.

TREATMENT

Laparoscopic radical resection of the abdominal mass was indicated. During the operation, the mass was found adhered to the surrounding omentum in the left upper abdominal region, in close proximity to the spleen, stomach, small intestine, and colon. The lesion was approximately 22 mm × 26 mm in size. Postoperative chemotherapy was not indicated.

OUTCOME AND FOLLOW-UP

The patient recovered well without any discomfort. At 3 mo follow-up, no complications such as recurrence or metastasis were observed.

DISCUSSION

Paragangliomas are rare neuroendocrine tumors originating from either the adrenal medulla, or the extra-adrenal sympathetic and parasympathetic ganglia. Symptoms involve the classic triad of headache, palpitation, and sweating. Adrenal and extra-adrenal sympathetic paragangliomas are often characterized by the excessive production of catecholamines, which can not only result in hypertension, but also associate with the risk of acute cardiovascular complications. Diagnosis is often based on plasma or urinary metanephrine measurements and nuclear imaging. Moreover, normal catecholamine levels have been reported to virtually exclude the presence of a sympathetic paraganglioma[1]. Intra-abdominal paragangliomas are particularly rare, with incidence of approximately 1 in 500000.

Parasympathetic paragangliomas are usually located in the head and neck, while sympathetic paragangliomas are more commonly located in the abdomen, followed by the chest and pelvis[2]. And the tumer that may present with cranial neuropathies when located along the skull base[3]. Head and neck paragangliomas are usually painless and slow growing, and are thus often an incidental clinical finding. They mainly manifest as carotid body tumors and vagal paragangliomas. As parasympathetic paragangliomas are non-secretory, symptoms are usually secondary to mass effects. These may include neck pain and dysphagia, conductive hearing loss and pulsatile tinnitus in cervical tympanic paragangliomas, as well as lower cranial nerve defects in advanced tumors[4]. Paragangliomas located outside of the head and neck may also be non-secretory, and are often accompanied by mild symptoms[5].

Due to the catecholamine-secreting nature of abdominal paragangliomas, the common clinical symptoms may include malignant hypertension, palpitation, headache, dizziness, anxiety, metabolic disorder syndrome, orthopnea, oliguria, anuria,

and hepatic encephalopathy, among others. In rare cases, patients may experience paraganglioma crises, an endocrine emergency resulting in life-threatening hemodynamic instability and end-organ damage[6]. This can often be misdiagnosed as septic shock, heart failure, thyroid storm, and malignant hyperthermia[7,8]. Given its mortality rate of approximately 15%, early recognition of the signs and symptoms of paragangliomas is thereby critical[7,9]. It was found that in patients with paraganglioma, the metaiodobenzylguanidine (MIBG) uptake intensity ratio was significantly higher in malignant lesions than in benign lesions. Therefore, iodine-131 MIBG uptake was able to distinguish between benign and malignant diseases, which was not helped by magnetic resonance imaging[10]. Furthermore, one study used radio-labeled MIBG and somatostatin analogues for scintillation imaging for correct localization. The results showed that MIBG was more accurate in imaging pheochromocytoma than somatostatin analogues. But somatostatin analogues are more accurate than MIBGs in detecting neuroendocrine tumors[11]. While symptoms are often paroxysmal, the clinical manifestations of paragangliomas can vary based on catecholamine subtypes, and may range from asymptomatic to life-threatening[12]. As exemplified in our case, the lack of obvious clinical abnormalities such as abdominal pain, abdominal distension, hypertension, and dizziness could have led to misdiagnosis or missed diagnosis.

Paragangliomas are mostly benign in nature, with surgical resection being the main treatment of choice. In contrast, malignant paragangliomas often warrant a multidisciplinary approach, involving endocrinology, oncology, surgery, nuclear medicine, radiotherapy, interventional radiology, and histology. So far, there are no standardized treatment regimens for metastatic diseases. Current treatment measures mainly involve beta-blockers and catecholamine synthesis inhibitors (A-methyl-p-tyrosine) to prevent tumor progression and minimize catecholamine-induced symptoms[13].

The diagnosis of paraganglioma in our case was mainly based on pathology, and was confirmed upon findings of Syn (-) and S-100 (+) on immunohistochemical staining. While morphologically similar to malignant perivascular epithelioid cell and stromal

tumors, S-100 played a role as a differentiating factor. As a neurogenic index, the positive expression of S-100 observed in our patient was in keeping with the origin of paragangliomas from chromaffin cells of the neural crest. In contrast, perivascular epithelioid cell tumors are mostly malignant in nature, and are characterized by positive HMB45, SMA, and Desmin, the latter 2 of which are myogenic indices. Stromal tumors are also commonly malignant, and are distinguished by the expression of CD117. In our case, the low expression of Ki-67 indicated a benign tumor.

CONCLUSION

In conclusion, intra-abdominal paragangliomas are clinically rare with no characteristic imaging findings, and are, as such, easily missed. However, surgical resection can associate with good clinical prognosis.

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