Case Report

Unusual Cause of Acute Abdomen—Ruptured Retroperitoneal Paraganglioma

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Ruptured retroperitoneal paraganglioma is a rare cause of acute abdomen. Its clinical presentation and laparoscopic features have seldom been reported in the literature. Herein, we report a case of ruptured retroperitoneal paraganglioma that presented as acute abdomen, and its subsequent management.


Key Words: acute abdomen, paraganglioma, rupture

Introduction

Nonfunctional retroperitoneal paraganglioma is a rare tumour. They are usually asymptomatic until attaining a remarkable size or when complications arise. Occasionally, they may be revealed incidentally during investigations for other reasons. Spontaneous tumour rupture can cause severe abdominal pain that mimics other acute surgical conditions. Herein, we report a case of ruptured retroperitoneal paraganglioma that presented as acute abdomen, and its subsequent management.

Case report

A 76-year-old man with a medical history of diabetes and hypertension was admitted to casualty for sudden onset of right lower quadrant pain. Blood tests revealed leukocytosis with white cell count of $17.4 \times 10^9/L$. Chest X-ray, amylase level, liver and renal function tests were all normal. Physical examination found local peritonism in the right lower quadrant of the abdomen, suggestive of acute appendicitis. Laparoscopic appendectomy was arranged. During laparoscopic examination, the appendix was found to be normal. The abdominal cavity was clean and unremarkable. Further examination found a non-pulsatile retroperitoneal haematoma in the right para-aortic region (Figure 1A). Laparoscopic ultrasound revealed a 5.5-cm retroperitoneal mass that was surrounded by blood (Figure 1B). The aorta, iliac vessels, inferior vena cava and kidneys were normal. The patient was treated conservatively and further investigations were performed to define the nature of the mass.

Contrast-enhanced computed tomography (CT) (Figure 2A), magnetic resonance imaging (MRI) (Figure 2B) and angiographic studies found a hypervascular tumour (Figure 3) extending from the right renal hilum down to the level of the aortic bifurcation. Neoplastic lesions such as retroperitoneal sarcoma or extra-adrenal phaeochromocytoma were suspected. However, the patient had normal serum and 24-hour urinary concentrations of catecholamines.

Early elective open operation was performed for tumour resection. Intraoperatively, a highly vascular tumour was found lying between the aorta and inferior vena cava. En bloc excision was successful and the patient had an uneventful recovery. Histopathological examination confirmed retroperitoneal paraganglioma (Figure 4). The patient was asymptomatic after operation, and follow-up CT scans at 6, 18 and 36 months showed no recurrence.
Figure 1. (A) Laparoscopic view shows a retroperitoneal haematoma (arrow). (B) Laparoscopic ultrasound reveals a retroperitoneal mass (arrow) surrounded by a rim of blood (arrowhead).

Figure 2. (A) Computed tomography reveals a lobulated isodense/hyperdense heterogeneously enhanced mass (arrow) in the retroperitoneal cavity. (B) The corresponding magnetic resonance image shows a T2-weighted hyperintense lesion (arrow).

Figure 3. Arteriography reveals an oval-shaped hypervascular mass with blood supply from the right L2 lumbar artery (arrow) and an ascending branch of the right superficial gluteal artery (arrowhead).

Figure 4. The tumour cells are arranged in small cell balls separated by vascular septae that contain nerve bundles and ganglion (haematoxylin & eosin, 200×).
Discussion

The nomenclature for paraganglioma is controversial.3–6 The 2004 WHO classification of endocrine tumours defines phaeochromocytoma as a tumour arising from chromaffin cells in the adrenal medulla. Closely related tumours in extra-adrenal sympathetic and parasympathetic paraganglia are classified as extra-adrenal paraganglioma. They can be found from the upper cervical region to the pelvis, along the autonomic nervous system. These tumours are most commonly present in the organ of Zuckerkandl at the aortic bifurcation, in the upper abdomen in association with coeliac, superior and inferior mesenteric ganglia, and in the mediastinum. A paraganglioma can be functional when they secrete catecholamine or nonfunctional.

Ruptured phaeochromocytomas are more frequently reported in the literature than nonfunctional paraganglioma. It is because ruptured phaeochromocytoma has very high mortality. In addition, it has some unique features due to hypersecretion of catecholamines. Therefore, most reports in the literature only describe ruptured phaeochromocytoma and not nonfunctional paraganglioma. However, the differentiation between these two tumours can only be confirmed through careful clinical, radiological and biochemical studies, which would be impossible under an emergency condition.

Spontaneous rupture of nonfunctioning retroperitoneal paraganglioma has not been reported in the English literature. It can cause severe abdominal pain that mimics other acute surgical conditions. Diagnostic laparoscopy, with the aid of laparoscopic ultrasound, allows detailed assessment and immediate exploration can thus be avoided. Paragangliomas are highly vascular tumours and often adhere to vital structures. Furthermore, they can be a functionally active tumour and secret catecholamines during tumour manipulation. Therefore, adequate preoperative work-up, including biochemical and radiological investigations, is mandatory for their management. The measurement of serum and 24-hour urinary concentrations of catecholamines is required as paragangliomas can be functionally active. CT and MRI can define the local extent of the tumour, although there are no characteristic features that are peculiar to paraganglioma.7,8

In a review of 236 cases of paraganglioma,9 [131I]- or [123I]-metaiodobenzylguanidine (MIBG) scintigraphic study was the least sensitive imaging study compared to CT and MRI. Although lacking in sensitivity, MIBG scan is highly specific and may be the only positive imaging test in some patients. We therefore suggest that MIBG scan should be included in the panel of investigations.

Preoperative angiographic study for the arterial supply of the tumour could be optional. However, preoperative embolization of the feeding vessels has been suggested by some authors to decrease the amount of operative blood lost during the removal of this vascular tumour.10 Since these tumours are radiosensitive and there are no reliable morphological criteria to separate benign from malignant paraganglioma, surgery is the treatment of choice for curative intent.11 In a review of 22 cases of retroperitoneal paraganglioma, the 5-year survival for tumours not resected was 19% compared with 75% after complete resection.12 The prognosis of paragangliomas can be very difficult to predict. However, their anatomical location may influence their biological behaviour: carotid body paragangliomas have a relatively low rate of malignant behaviour while retroperitoneal paragangliomas tend to have a higher incidence of malignant transformation.12 Therefore, regular surveillance with CT scan is necessary.

It is well known that paraganglioma, whether functional or nonfunctional, is a highly vascularized tumour. Emergency exploration of such a mass without adequate preoperative assessment could be dangerous. It is even worse in the presence of haematoma which obscures the tumour anatomy. For functional paraganglioma, immediate exploration can have a mortality rate of up to 81%!13 In this era of minimally invasive surgery, diagnostic laparoscopy is frequently used for patients with acute abdominal pain. The laparoscopic features of a ruptured retroperitoneal paraganglioma reported in the present case can remind our surgical colleagues of this rare condition so that potential lethal immediate exploration is avoided.

Ruptured retroperitoneal paraganglioma is an unusual cause of acute abdomen. This rare condition should be included in the differential diagnosis when a retroperitoneal mass is found during diagnostic laparoscopy for acute abdomen. Immediate exploration of such a vascular and potential catecholamine-secreting tumour under emergency conditions could be lethal.

References