Case Report
Gastrointestinal stromal tumor masquerading as an adrenal tumor: a case report with literature review

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Abstract: Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms of the gastrointestinal tract. They are rarely encountered in adrenal region. Here we report the case of a 55-year-old man with GIST masquerading as an adrenal tumor. The patient was admitted to our hospital with epigastric discomfort lasting 20 days. Computed tomography scan revealed a 5×5.5 cm round-like mass located in the left adrenal region with mild homogeneous enhancement. Laboratory testing was without positive findings. The patient was initially believed to have a left adrenal neoplasm in the preoperative diagnosis. Complete resection of the tumor was performed using a laparoscopic approach. The pathologic diagnosis was suggestive of a gastrointestinal stromal tumor. The patient had no evidence of recurrence during the 14-month follow-up. GISTs that arise in adrenal area are difficult to distinguish from nonfunctional adrenal masses. Prompt recognition of these ectopic tumors is conducive to minimize the chance of tumor rupture during operation.

Keywords: Gastrointestinal stromal tumor, adrenal neoplasm, laparoscopy

Introduction
Gastrointestinal stromal tumors (GISTs) are potentially malignant mesenchymal tumors of the gastrointestinal (GI) tract and typically arise from stomach or small intestine in adults [1]. These tumors frequently occur in the 50 to 60-year-old patients, without preference for gender [2]. Patients are often asymptomatic or experience nonspecific symptoms including epigastric discomfort and pain, but some severe symptoms such as GI bleeding and ulcer-like symptoms may also occur [3]. Localization of GISTs in adrenal area is extremely rare that, as our knowledge, only a few cases have been reported to date [4-6]. The imaging characteristics of this entity are similar to those of hormonally inactive adrenal masses, leading to diagnostic difficulties. A definitive diagnosis could only be achieved through histopathological examination and immunohistochemical staining of the tumor. Here, we report a rare case of GIST disguising an adrenal tumor that was successfully resected laparoscopically.

Case presentation
A 55-year-old male patient presented with discomfort in the left upper quadrant for 20 days and was admitted to our hospital. He had medical history of hypertension for 5 years, and the blood pressure was controlled satisfactorily with medication treatment (<140/90 mmHg). Apart from cardiac valve operation that was performed 10 years before, no other surgical history was present. On physical examination, no positive findings were found. Furthermore, he denied any history of smoking, alcohol, and drug abuse.

A plain helical computed tomography (CT) scan found a 5×5.5 cm round-like mass with density similar to that of the soft tissue in adrenal area (Figure 1A). During enhancement scanning, the tumor was slightly enhanced uniformly (Figure 1B). Results of all laboratory tests including complete blood counting, biochemistry, liver function, blood coagulation test and metabolic parameters such as serum levels of aldosterone and cortisol were within normal limits. No
positive findings were revealed from electrocardiogram examination and chest x-ray. Based on these examinations, a clinical diagnosis of adrenal tumor was made. The patient was prepared to be treated by laparoscopic adrenalectomy.

Operatively, it was easy to see a solid mass above the upper pole of the left kidney. As the tumor could hardly be distinguished from the adrenal gland, we excised the tumor as well as surrounding glandular tissue. However, local tumor was found closely adhere to greater curvature of the stomach. Hence, we took a surgical consultation to a general surgeon.

Figure 1. Plain CT scan revealed a 5×5.5 cm soft tissue mass in the left adrenal region (A). The tumor was slightly enhanced during arterial phase (B).

Figure 2. The histopathologic examination showed abundant spindle cells arranged infascicular (hematoxylin and eosin stain, A: ×100; B: ×200). Immunohistochemical appearance demonstrated that tumor cells were positive for CD117, CD34, and DOG-1, focally positive for Ki-67, and negative for SMA and S-100 (C-H: ×100).
GIST masquerading as an adrenal tumor

Despite the many difficulties we faced, the operation was finally a success with interdisciplinary cooperation.

On gross examination, the tumor specimen was found to be intact. Microscopically, the tumor was composed of spindle cells arranged in fascicular (Figure 2A, 2B). Immunohistochemical analysis showed that the tumor was positive for CD117, CD34, and DOG-1, focally positive for Ki-67, but negativity for SMA and S-100 (Figure 2C-H). According to these results, the pathological analysis showed a gastrointestinal stromal tumor.

During the 14-month follow-up, the patient was without any uncomfortable and free of recurrence detected by radiology.

Discussion

GISTs are the commonest gastrointestinal mesenchymal neoplasms that derive from interstitial cells of Cajal [7]. According to statistics, GISTs occur in around 0.32 per 100,000 in the United States [8]. They can arise anywhere within the GI tract, but rarely do they occur in sites outside digestive system. In 2011, Sereg et al. [6] first reported a case of gastrointestinal stromal tumor presenting as a hormonally inactive adrenal mass. Until date, only four such cases (two gastric GISTs mimicking adrenal mass and two primary adrenal GISTs) have been documented in the English medical literature (Table 1) [4-6]. Unlike the pheochromocytoma or aldosteronoma, preoperative endocrine tests of GISTs are within normal range. CT images of these tumors are usually without characteristic signs and can often appear as soft tissue mass with slight enhancement. In the present case, the patient was diagnosed as adrenal tumor in the initial diagnosis. In retrospect, however, we found the tumor closely related to the greater curvature of the stomach when reading the CT images again. Therefore, as they can easily be misdiagnosed as non-functioning adrenal tumors, the preoperative diagnosis of adrenal GIST requires a careful reading of the imaging data and a high index of clinical suspicion.

Surgical resection of the tumor is the current priority management for localized GISTs [9]. Owing to GISTs have a risk of tumor rupture during operation which may lead to the spread of tumor cells, en bloc resection of the tumor is of importance. In recent years, with the development of minimally invasive techniques, the utility of laparoscopic wedge resection (LWR) for gastric GISTs has been well established and increasingly emerged as the preferred treatment option. And even gastric GISTs larger than 5 cm were also be resected by LWR [10]. Since GISTs seldom spread via the lymphatic system, it is generally accepted that lymph node dissection is not necessary [11]. For patients with unresectable tumors or advanced GIST, tyrosine kinase inhibitors are the suggested treatment. Abou Al-Shaar et al. [4] reported an adrenal GIST patient with almost complete resolution of the tumor metabolism after receiving a 3-month anti-tyrosine kinase therapy (imatinib). In addition, as GISTs are generally resistant to radiotherapy and chemotherapy, these treatments are seldom used in clinical practice [12].

The definitive diagnosis of the disease mainly depends on the postoperative pathological examination and immunohistochemistry. The histologic features of GISTs are presenting as spindle, epithelioid, or mixed cells, usually arranged in fascicular clusters or sheets. Immunohistochemically, these tumors are characterized by positive staining of CD117. Additionally, about 60%-80% of the lesions are

Table 1. Summaries of previously published articles on GISTs outside GI tract

<table>
<thead>
<tr>
<th>Author</th>
<th>Title</th>
<th>Year</th>
<th>No. of cases</th>
<th>Main content</th>
<th>Clinical features</th>
<th>Therapeutic method</th>
<th>Pathological findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sereg et al.</td>
<td>Gastrointestinal stromal tumor presenting as a hormonally inactive adrenal mass.</td>
<td>2011</td>
<td>1</td>
<td>Primary adrenal GIST</td>
<td>Non-special</td>
<td>By laparotomy</td>
<td>Positive for CD117 and a-SMA</td>
</tr>
<tr>
<td>Chung et al.</td>
<td>Laparoscopic resection of gastric gastrointestinal stromal tumors presenting as left adrenal tumors.</td>
<td>2012</td>
<td>2</td>
<td>Laparoscopic excision of GIST</td>
<td>Non-special</td>
<td>Laparoscopic resection</td>
<td>Positive for CD117</td>
</tr>
<tr>
<td>Abou Al-Shaar et al.</td>
<td>Gastrointestinal stromal tumor of the adrenal gland: a case report and review of the literature.</td>
<td>2015</td>
<td>1</td>
<td>Primary adrenal EGIST</td>
<td>Non-special</td>
<td>Imatinib</td>
<td>Positive for CD117 and CD34</td>
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positive for CD34, and 85%-95% of CD117-positive GISTs as well as 30%-36% of CD117-negative GISTS are discovered on DOG-1 expression [13]. Other antigens such as S-100 and smooth muscle actin can also be detected in some cases [14, 15]. In this study, GIST was identified immunohistochemically by positive staining of CD117, CD34 and DOG-1.

It can be difficult to determine the biological behavior of GISTs, ranging from benign to malignant. However, their prognosis is considered to be associated with tumor size, mitotic rate, and location of the primary lesion [16]. On the basis of the modified National Institutes of Health (NIH) classification that evaluated the risk of aggressive behavior [17], our case has showed a potentially malignant biological behavior due to large size (maximum diameter of >5 cm) and much higher mitotic index (more than 5 mitoses per 50 HPFs). Meanwhile, it was reported that approximately 40% to 50% of GIST patients will have local recurrence or metastasis after radical resection [16]. Therefore, a close surveillance after operation is needed.

Conclusion

The occurrence of GISTs in the adrenal region is very rare and can be difficult to make differential diagnosis from nonfunctional adrenal tumor. The radiological presentation of the close anatomical relationship between the tumor and gastrointestinal tract was suggestive and should arouse surgeons’ suspicion. As there is a potential risk of tumor spread, a delicate and complete tumor excision is suggested. Histopathology examination is golden standard for definitive diagnosis of the tumor. Additionally, long-term follow up is indispensable in monitoring the progression of GIST.

Disclosure of conflict of interest

None.

Abbreviations

GIST, Gastrointestinal stromal tumor; CT, computer tomography; LWR, laparoscopic wedge resection.

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GIST masquerading as an adrenal tumor


