LESS

Case Report

Laparosc Endosc Surg Sci 2023;30(1):32-35
DOI: 10.14744/less.2023.76892

Retroperitoneoscopic resection of a jejunal gastrointestinal stromal tumor masquerading as an adrenal incidentaloma; a unique reminder of the importance of clinical decision-making

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ABSTRACT

Although gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors, they are rare, especially those arising from the small bowel. Adrenal incidentalomas (AI) are much more common, the majority of which are non-functional. Radiological features largely guide the diagnosis and management of both types of tumors and due to investigative limitations, gastric and small bowel GISTs can be misdiagnosed on imaging as AI, especially if present in the left upper quadrant. A 58-year-old male was referred for the management of a left adrenal incidentaloma following investigations for weight loss. An adrenal protocol computed tomography demonstrated a 32-mm left adrenal mass without atypical features. Investigations including gastroscopy and adrenal biochemistry were normal. Positron emission tomography revealed a highly avid adrenal mass suggesting a non-functioning adrenal carcinoma. A prone retroperitoneoscopic left adrenalectomy was performed, but no abnormal adrenal lesion was found. An intraoperative re-review of imaging, further retroperitoneoscopic exploration of the retroperitoneum and the peritoneum identified a pedunculated tumor attached to the proximal jejunum. The tumor was successfully resected retroperitoneoscopically. Histopathology revealed a GIST. No previous reports of a retroperitoneoscopic GIST resection have been published nor has a misdiagnosis of a small bowel GIST as an adrenal tumor been published. This highlights the importance of intraoperative correlation of imaging and intraoperative findings and exploring alternative diagnoses when encountering discordance. Attention to detail is required when tumors are solely radiologically diagnosed without additional confirmatory investigations, especially so in potentially anatomically unclear regions.

Keywords: Adrenal incidentaloma, intraoperative decision-making, jejunal gastrointestinal stromal tumors, retroperitoneoscopic

Introduction

Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal tumors of the gastrointestinal system.[1] The small bowel is the second most common location for GISTs after the stomach.[2] However, overall, small bowel GISTs are exceedingly rare, with an annual incidence of approximately 4–14 cases/1 million.[3] Approximately 70% of cases will present with symptoms

Received: 21.02.2023 **Revision:** 21.02.2023 **Accepted:** 25.02.2023
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which are generally vague in nature and may include abdominal discomfort, nausea, and weight loss and can depend on the location of the tumor. 20% of cases are incidentally found and 10% found at autopsy.[3] Much more common than small bowel GISTs are adrenal incidentalomas, with an overall prevalence of 4.2%, rising in prevalence with age to approximately 10% in patients aged 70 and older.[4] By definition, they are classified an incidentalomas as they are only incidentally found on imaging, and unsurprisingly, the majority of them are non-functioning and asymptomatic.[5] For both types of tumors, GISTs and non-functioning adrenal tumors, preoperative confirmatory diagnosis with tissue biopsy is uncommon or largely unhelpful, and thus, the diagnosis is reliant largely on imaging features and where possible, investigations unique to location (endoscopy for gastric or upper small bowel GIST) and function (hormonal assays for functioning adrenal incidentalomas).[2,6] In the literature, there have been only six published cases worldwide of gastrointestinal GISTs (all gastric) being misdiagnosed on imaging as left adrenal tumors.[7-11] We present the first case where a small bowel GIST was misdiagnosed as a left adrenal incidentaloma. Additionally, this is the first case where a GIST was resected entirely retroperitoneoscopically. We discuss the importance of intraoperative decision making after a preoperative misdiagnosis and the importance of correlating the clinical picture with discordant operative findings. This case is a reminder to the reader regarding the imperfect nature of investigations for such conditions and describes a unique approach to resecting a small bowel GIST.

Case Report

A 58-year-old man was referred to the endocrine surgical service for treatment of a left adrenal incidentalinoma after investigation for weight loss and dysphagia. An initial computed tomography (CT) and a dedicated adrenal protocol CT demonstrated a 32 mm left adrenal mass with features not typical for an adrenal adenoma (Fig. 1a).

Additional investigations included a gastroscopy, did not reveal any further information. Adrenal biochemistry was normal and a FDG PET revealed a highly avid left adrenal mass suggestive of a non-functioning adrenal tumour (Fig. 1b). The patient was consented for a prone retroperitoneoscopic left adrenalectomy.

A prone left retroperitoneoscopic approach adrenalectomy was performed. During surgery, the left adrenal

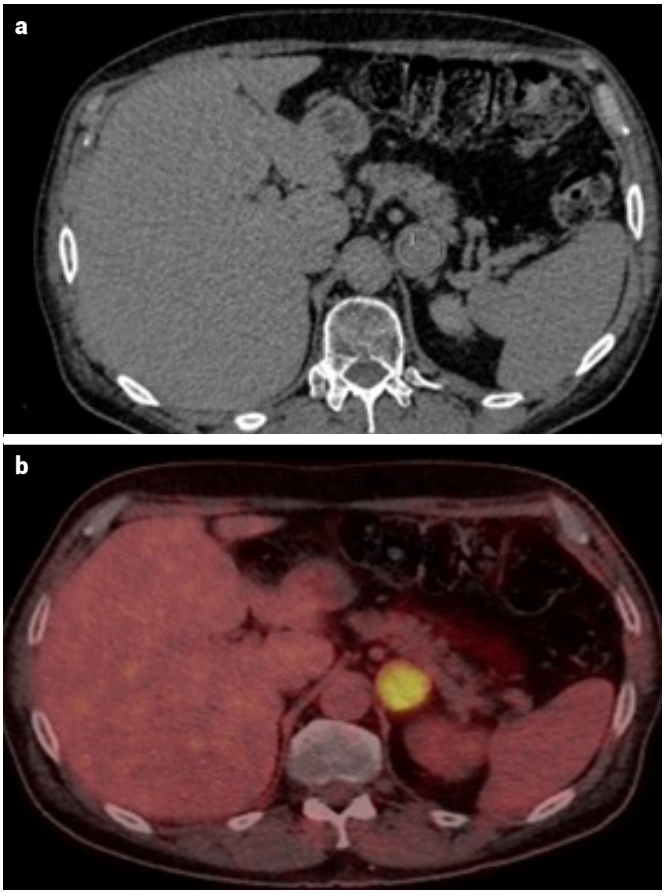

Figure 1. Computed tomography (CT) adrenal protocol **(a)** and PET scan **(b)** axial images revealing a 32 × 30 mm left adrenal lesion with features not typical for an adrenal adenoma including Hounsfield units of 58 and a washout of −62.5% at 15 min. FDG PET CT revealing an intensely FDG avid adrenal lesion suggesting malignancy.

gland was identified and although it did not entirely correlate with the preoperative imaging, the left adrenal gland was resected without any complication. Once the adrenal gland was exteriorised, the surgical team examined and measured the specimen to correlate the dimensions of the specimen to the imaging dimensions of the tumour and left adrenal gland. Examination revealed a normal appearing gland without any evidence of tumour and its dimensions did not correlate to that of the tumour. The decision was then made to re-explore the retroperitoneum with the current retroperitoneoscopic approach from the upper pole of the left kidney to the uppermost margins of the resection border. This allowed for identification of an abnormal mass adjacent to and abutting the resection border, covered by posterior peritoneum. The posterior peritoneum was subsequently breached to this position and a large solid vascular, pedunculated and exophytic lesion was identified, attached by a thin stalk to the proximal jejunum (Fig. 2).

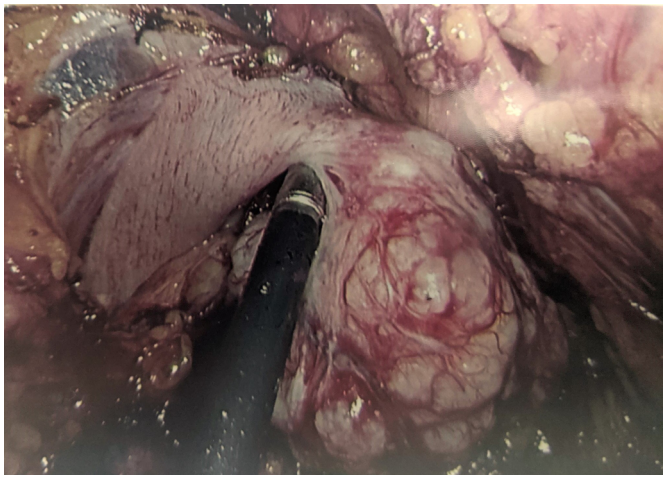

Figure 2. Intra-operative retroperitoneoscopic image of the malignant appearing intra-peritoneal solid, exophytic lesion tethered to the serosa of the proximal jejunum in the left upper quadrant.

The lesion was resected using a laparoscopic tri-stapler (Endo-GIA® 60 mm Tan, Medtronic, Minneapolis, USA) across the stalk in an orientation as to not comprise both the bowel lumen as well as the margins of the tumour. The patient recovered rapidly and was discharged day 1 postoperatively. The histopathological findings of the two specimens were a normal left adrenal gland and a 35 mm low grade GIST with clear margins. The multidisciplinary consensus for the GIST was for annual PET scan surveillance and no requirement for adjuvant treatment.

Written informed consent was obtained from the patient.

Discussion

Compounding their rare incidence, 0.32/100,000 in the United States, the vague and location-specific nature of the symptomatic portion of GISTs does not usually yield an immediate diagnosis.[12] They are often found incidentally on imaging. Gastroscopy can aid in the diagnosis of gastric GISTs; however, due to accessibility, small bowel GISTs are usually only discovered incidentally or following complications such as obstruction or bleeding.[2] Additionally, blood tests will be generally normal in GISTs unless a complication such as gastrointestinal bleeding or metastasis has occurred. Adrenal incidentalomas occur at a frequency between 3% and 10% worldwide, much more common than GISTs. The large majority of which are non-functioning adenomas. Our patient presented with vague gastrointestinal symptoms, had normal blood tests and a normal gastroscopy was subsequently diagnosed only by way of CT imaging with a left adrenal incidentaloma. Given the size, an FDG-PET scan was warranted and the highly avid lesion, within the context to all the clinical and investigative information, led to a suspicion of a non-functioning left adrenal carcinoma. Given the relatively smaller size of the tumour a retroperitoneoscopic approach was planned as evidence has shown there is minimal risk of spillage and tumour breach comparable to open resection of adrenal carcinomas.[13]

Several factors from this and previous cases highlight some learning points. The six previous cases of adrenal lesions were all left sided lesions that underwent transabdominal laparoscopic surgery. All cases were GISTs attached to the greater curve of the stomach.[7-11] In the majority of cases, the misdiagnosis was unrecognised until only after the transabdominal left adrenalectomy was performed, either at the time of initial surgery or after the surgery when pathology and imaging revealed the misdiagnosis.[7-11] 50% of cases underwent an enbloc resection of a LUQ mass presumed to be the adrenal tumour and only one case recognised the misdiagnosis intra-operatively prior to adrenalectomy and correct surgical resection of the isolated GIST was performed.

Our case highlights the importance of intraoperative correlation of imaging and the surgical findings when clinical discordance is evident and the importance of exploring other possibilities. This is an important learning case and should be of interest to all proceduralists diagnosis or operating on retroperitoneal tumours or GISTs, including gastrointestinal, endocrine and urological surgeons and gastroenterologist. This is especially so given that this same misdiagnosis has occurred at least six other times, as reported in the literature. The preoperative trap regarding the misdiagnosis relates to the rarity of GISTs, especially in the small bowel in the left upper quadrant, adjacent to the left adrenal gland with similar radiological characteristics.[8,9] In such cases, where clinically the tumour is likely to have vague or no symptoms or complications and non-functioning and in an anatomical position to readily accessible by endoscopy, the preoperative diagnosis relies primarily on imaging without any other clinical or investigative clues. Given the above, radiologists, gastroenterologists and surgeons alike can understandably be drawn into diagnosing the much more common pathology, an adrenal incidentaloma rather than consider an exponentially rarer diagnosis.[10] As such, in these cases laden with potential pre-and intraoperative pitfalls, clinician leadership and judgment are

crucial to snatch victory from the pre-determined jaws of clinical defeat. Evidence reveals that clinicians rely mainly on intuitive (experienced-based) or analytical modes of thinking to make successful key intraoperative decisions in times of uncertainty.[14] As intuitive decision making usually relies on years of clinical experience, an important element to highlight in this case was the analytical decision making. Given the discordance between preoperative imaging and intraoperative findings, the surgical team closely analyzed the resected specimen including measuring the specimen and compare this to the measurement dimensions of the tumour and the adrenal gland on pre-operative imaging. This is evidence-based practice in areas including head and neck, breast and upper gastrointestinal surgery largely to inspect the surgical margin to reduce the risk of leaving positive oncological margins.[15] However, in our case, this practice resulted in the immediate recognition of the surgical mistake as it was clear there was no infiltrating tumor, that the tumour was still in vivo and not associated with this gland. Following this recognised mismatch and the intra-operative findings, a controlled breach of the peritoneum was performed and subsequently allowed for the identification and safe surgical resection of the jejunal GIST. Given adequate access and visibility with the retroperitoneoscopic approach, there was no indication for repositioning the patient and attempting a transabdominal or open conversion.

Conclusion

This case highlights an uncommon but significant clinical scenario to remind the reader to always analytically question atypical clinical findings and formulate alternative diagnoses and solutions.

Disclosures

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

Authorship Contributions: Concept – M.G.; Design – M.G., M.O.J.; Supervision – M.G., M.K.Y.; Materials – M.G.; Data collection and/or processing – M.G., M.O.J.; Analysis and/ or interpretation – M.G., M.O.J.; Literature search – M.G., M.O.J.; Writing – M.G., M.O.J.; Critical review – M.G., M.O.J., M.K.Y.

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