International Journal of Surgery Case Reports 89 (2021) 106613

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Retroperitoneal GIST: An exceptional location of a rare tumour. A case report from Ouagadougou and review of the literature

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ARTICLE INFO

Keywords:
Gastro-Intestinal-Stromal-Tumour
Retroperitoneum
Surgery
Imatinib
Case report
Ouagadougou

ABSTRACT

Introduction: GIST can occur in all segments of the gastrointestinal tract with a predilection for the stomach. Retroperitoneal localization remains exceptional. We report a case to describe our diagnostic and therapeutic approach.

Case presentation: A 55-year-old patient was admitted with borborygms and a sensation of lumbar swelling for 6 months. He was diabetic and hypertensive. The clinical examination noted a right lumbar mass with perception of bowel sound anterior to the mass, with minimal discomfort and mobility. Ultrasound revealed a hypervascularised, encapsulated, well-limited retroperitoneal tissue mass in contact with the right psoas muscle. Abdomino-pelvic CT scan showed a large, hypervascularized, encapsulated, calcified tissue mass measuring 147 × 106 mm in close contact with the outer edge of the right psoas muscle, suspected of being malignant. MRI noted a suspicious process developed at the expense of the right psoas muscle in its lumbar and iliac portion suggestive of a psoas rhabdomyosarcoma. The patient underwent laparotomy with a retroperitoneal approach by lumbar incision. Histology and immunohistochemistry revealed a GIST expressing CD117. The patient was put on imatinib for 6 months. He is complaint-free after 4 months.

Discussion: Despite the rarity of retroperitoneal GIST, it should be considered in the presence of any retroperitoneal mass. This will allow for early management.

Conclusion: Retroperitoneum is an exceptional location for GIST. Surgery remains the mainstay of curative treatment. Adjuvant imatinib reduces the risk of recurrence. The prognosis is usually good.

1. Introduction

Gastrointestinal stromal tumours are rare, accounting for less than 1% of cancers [1]. They arise from spindle cells, often epithelioid, rarely mixed, called Cajal cells [2,3]. Their main location is in the digestive tract with a predilection for the stomach [1,4]. Retroperitoneal location is exceptional and is only reported in case reports [2,3]. Treatment is mainly surgical and consists of excision without rupture of the pseudocapsule [5]. Adjuvant imatinib reduces the risk of recurrence [2,5]. The treatment of GIST is not codified because of its rarity, and is based on the treatment of both retroperitoneal sarcoma and digestive GIST [2,5]. We report a case to describe our diagnostic and therapeutic approach. This case has been reported in line with SCARE criteria [6].

2. Case presentation

A 55-year-old patient, Mossi ethnicity, was admitted to the oncology clinic with borborygma and a sensation of lumbar swelling for 6 months. His history included diabetes and arterial hypertension which were regularly monitored. We did not note any family history of GIST. The patient has no previous surgical history. Any allergies and/or adverse reactions weren't identified. He is an electrical engineer, is married and comes from Ouagadougou. He is not a smoker and does not drink alcohol. His body mass index was 28.9 kg/m^2. The clinical examination

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https://doi.org/10.1016/j.ijscr.2021.106613
Received 5 October 2021; Received in revised form 21 October 2021; Accepted 16 November 2021
Available online 19 November 2021

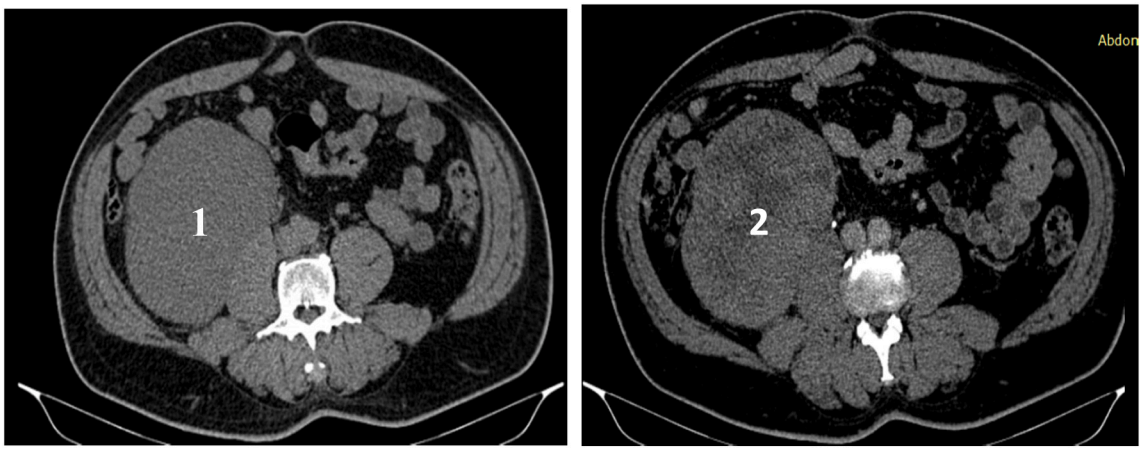

Fig. 1. Axial slices CT scan.
1, 2: retroperitoneal tumor
1: Without contrast injection
2: With contrast injection at portal time.

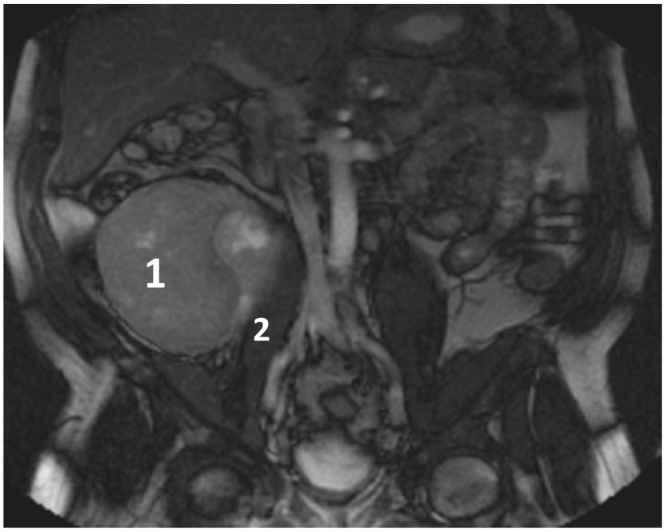

Fig. 2. Abdominopelvic MRI showing the mass and its relationship to the psoas muscle: coronal sequence.
A retroperitoneal mass of heterogeneous signal, compressing the psoas muscle, whose signal is not modified and remains comparable to the contralateral psoas
1: Mass
2: right psoas muscle.

noted a right lumbar mass with perception of bowel sound anterior to the mass, with minimal discomfort and mobility. Any concurrent treatments were notified. The rest of the examination was normal.

Hemoglobin level was 11.8 g/dl. Ultrasound showed a hypervascularised, encapsulated, well-limited retroperitoneal tissue mass in contact with the right psoas muscle. Abdomino-pelvic CT scans showed a large, hypervascularised, encapsulated, calcified tissue mass measuring 147 × 106 mm in intimate contact with the outer edge of the psoas muscle and suspicious of malignancy (Fig. 1). MRI noted a suspicious process developed at the expense of the right psoas muscle in its lumbar and iliac portion suggestive of a psoas rhabdomyosarcoma (Fig. 2). Preoperative biopsy, histology and immunohistochemistry revealed a gastrointestinal stromal tumour. It overexpressed CD117 receptors with a Ki 67 of 20%. The patient was informed of the need for surgery by his surgeon and he gave his consent. He received psychological care before surgery and a preoperative resuscitation. We performed under general anaesthesia, a right lumbar incision on top of the swelling. Exploration revealed a retroperitoneal, encapsulated mass with significant peripheral vascularisation. These arteries originated from the lumbar and iliac arteries. The mass laid on the right psoas muscle without invading it. It was a mass that developed independently of the retroperitoneal organs. We proceeded with a meticulous dissection freeing the mass without breaking it. The psoas fibres in contact with the wall of the mass were removed. The mass measured 16 cm × 14 × 13 cm (Fig. 3). The procedure remained retroperitoneal without rupture of the peritoneum. After checking the haemostasis, we left a tube drain in place for three days. The patient received postoperative resuscitation including administration of analgesics and antibiotics to prevent complications. He was discharged on the third postoperative day. He was put on imatinib 400 mg per day for 12 months. Presently, 6 months later, he has no complaint and his CT scan does not show any stigma of recurrence.

3. Discussion

Until 33 years ago, the majority of mesenchymal tumours were considered to be smooth muscle tumours (leiomyomas, leiomyosarcomas, etc.) [7]. In 1983, Mazur and Clark introduced the term gastrointestinal stromal tumour (GIST) to describe a distinctive type of non-smooth muscle mesenchymal tumours overexpressing surface receptors c-KIT or CD117 and CD34 [1,4]. GISTs are rare tumours identified by advances in immunohistochemistry [4]. Prior to this era, they were considered leiomyosarcomas [7]. They occur preferentially in the gastrointestinal tract with a preference for the stomach (60%) [1,4]. They may present by pain and/or lumbar curvature [1,4]. CT scan is a key examination that shows a retroperitoneal mass pushing the intra-abdominal viscera forward, the large vessels laterally [1]. It is an organ-independent mass, arising from isolated Cajal's cells in the retroperitoneum [2,3,8]. Abdominopelvic MRI provides valuable information by showing the absence of a fatty component, eliminating liposarcoma [2,5,8]. It also provides a better description of the relationship of the mass to adjacent structures [1,8]. The diagnosis is made either on biopsy specimens as in our case, or on the surgical specimen [1,7]. The vast majority (94.6%) overexpress CD34 or CD117 surface receptors [4,7]. Their grade of malignancy has been established by Fletcher who distinguishes between low, intermediate and high grades of malignancy [4,7]. All extragastric GISTs larger than 5 cm or of any size with more than 10 mitoses per 50 fields are considered Fletcher high grade [7]. In our case, the mass was larger than 10 cm. It was therefore at high risk of recurrence. Furthermore, the presence of tumour necrosis classifies the tumour as high grade malignant with a significantly higher

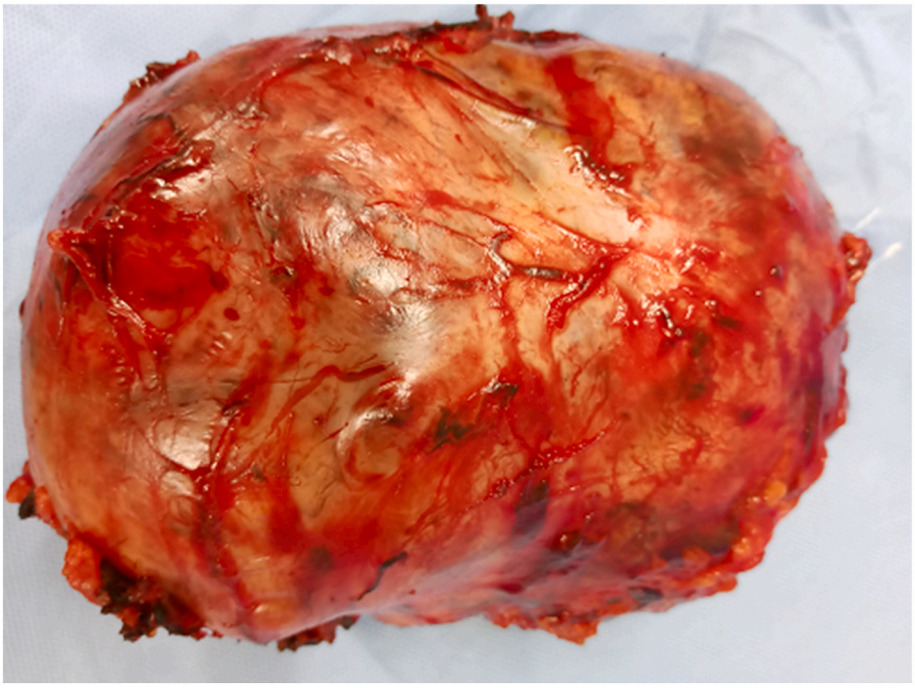

Fig. 3. Postoperative specimen: encapsulated mass.

risk of recurrence compared to non-necrotic tumours [8,9]. The treatment of retroperitoneal GISTs is not clearly standardised because of their rarity [2,3]. It is based on the treatment of digestive GIST [5]. It consists of a monobloc resection of the tumour without rupturing the capsule [2,5]. Surgery can be performed via a conventional or laparoscopic approach [10]. Surgery is the main treatment and is combined with adjuvant imatinib for retopritonal GISTs overexpressing surface C-kit receptors and classified as high-grade by Fletcher [Şentürk, Krunal]. For metastatic or non-resectable GIST, imatinib can be used as neoadjuvant or adjuvant therapy [11]. The daily dose varies between 400 and 800 mg [4,5]. It lasts between 6 and 18 months depending on the author [5]. In our case, surgery was the first treatment followed by imatinib for 12 months. The prognosis of GIST has generally been good since the advent of imatinib [2,5]. In our case, despite the size of the tumour, the patient had no complaint or signs of recurrence. Surgeons should keep in mind that despite the rarity of retroperitoneal GIST, it should be considered in the presence of any retroperitoneal mass. This will allow the diagnosis to be made preoperatively and neoadjuvant treatment with imatinib to reduce the size, facilitate surgery, reduce the risk of recurrence and improve the prognosis.

4. Conclusion

Retroperitoneum is a rare location for GIST. They can be distinguished from other mesenchymal masses by immunohistochemistry. Surgery is the main treatment. It consists of a monobloc excision without capsular rupture. Imatinib reduces the risk of recurrence in high-grade C-kit overexpressing tumours. The prognosis is generally good.

CRediT authorship contribution statement

ZONGO Nayi, OUEDRAOGO Nabonwindé Lamoussa Marie: Conceptualization, Methodology, Writing - Original Draft, Investigation ZONGO Nayi, OUEDRAOGO Nabonwindé Lamoussa Marie, KOAMA Adjirata: Formal analysis, acquisition of data, Statistical analysis and interpretation of data. OUATTARA Souleymane, WINDSOURI Mamadou, YAMEOGO, Paratyande Bonaventure: Visualization, Investigation ZONGO Nayi: Supervision.

Declaration of competing interest

The authors declare that they have no competing interests regarding the publication of this manuscript.

Acknowledgements

We thank the whole team of Yalgado Ouédraogo Teaching hospital of Ouagadougou for administrative and technical support.

Sources of funding

No sponsors to declare.

Ethical approval

Ethical approval is not needed for this case report as patient consent and we are not trialing a new device.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

N/a.

Guarantor

Nayi ZONGO.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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