CASE REPORT

Rare case of primary leiomyosarcoma of sigmoid mesocolon

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SUMMARY

We experienced a rare case of primary leiomyosarcoma of sigmoid mesentery. A 45-year-old woman was presented to us with left iliac fossa mass and discomfort for 4-month duration. CT scan of abdomen and pelvis revealed a huge mass 14 cm×14 cm×16 cm occupying left iliac fossa mimicked having a large left ovarian carcinoma. She was subsequently planned for elective total abdominal hysterectomy and bilateral salpingo-oophorectomy by gynaecology team. During laparotomy, a huge mass was revealed arising from sigmoid mesentery invaded to the left lower ureter. Curative resection was done and pathological findings show the tumour being leiomyosarcoma with immunohistochemistry tests on caldesmon, desmin, smooth muscle actin and CD34 reagent all positive. Clinicopathological and literature review of this rare primary leiomyosarcoma of mesocolon was discussed in our case presentation

BACKGROUND

Before 1998, the common mesenchymal tumour, gastrointestinal stromal tumour (GIST), may have been misclassified as mesenteric leiomyosarcoma due to the lack of differentiation by immunohistochemistry. Primary leiomyosarcoma, which especially originated from mesentery of a colon, is a rare entity with only few cases reported over the world.¹ Thus, the characteristics of clinicopathology of mesenteric leiomyosarcomas that originated from the smooth muscle lineage remain unclear. The imaging by ultrasonography or CT scan of the abdomen can differentiate the presence of cystic, solid or necrotic malformation within the tumour compared with other tumours.² They usually attained a large size and possible distant metastasis at the time of diagnosis. A treatment strategy for mesenteric leiomyosarcoma is surgical excision with a wide margin.^{3,4}

CASE PRESENTATION

We reported a case of a 45-year-old woman presented to gynaecology outpatient clinic with complaints of left iliac fossa mass with discomfort for 4 months. Her gynaecological, urinary and bowel habits were normal. She was a pre-morbidly known case of hypertension on regular treatment and no previous admission to hospital before. On examination, the general physical examination was normal and vitals were stable. Abdomen examination noted there was an irregular and globular mass

arising from the pelvis measuring 32–34 weeks' size of pregnancy, non-tender, firm to hard in consistency with restricted mobility. The mass was not ballotable and no ascites were demonstrated. On per speculum examination, cervix and vagina were in normal conditions. Digital rectal examination showed no intraluminal mass.

INVESTIGATIONS

Baseline and routine blood workouts were within normal limits. Plain radiography showed no abnormality. CT scan of abdomen and pelvis was done and revealed a huge mass 14 cm×14 cm×16 cm occupying the pelvic cavity possibly arising from the left ovary. No distant metastasis was noted (figure 1).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis was possible left ovarian carcinoma, retroperitoneal tumour, intraperitoneal lymphoma or less likely perforated sigmoid colon tumour.

TREATMENT

The patient was admitted to gynaecology ward and subsequently planned for elective total abdominal hysterectomy and bilateral salpingo-oophorectomy by gynaecology team. Intraoperatively, the uterus, bilateral Fallopian tube and ovaries were normal. There was a mass of size 20 cm×15 cm, irregular and lobulated, and hard mass with some retroperitoneal adhesion connections with sigmoid mesentery and left ureter (figure 2). Colorectal surgeon was called intraoperatively; found that it was a huge sigmoid mesocolon tumour with lower part of ureter embedded to the tumour. There was also minimal dense adhesion surrounding the area. The tumour was excised from the lateral approach to release from the adjacent surrounding structure. After the lateral excision, it was posteriorly noted there was an invasion to the lower part of left ureter. The tumour was excised with wide margin of 5 cm proximal and 5 cm distal together with the involved part of lower ureter (figure 3). The urologist was called in to perform uretero-ureteral anastomosis. The recovery period was uneventful and she was discharged on 10th postoperative day with normal functional activities. The histopathological examination came back as leiomyosarcoma with the complete clear margin that has been excised, and immunohistochemistry tests on caldesmon (figure 4), desmin (figure 5), smooth muscle

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Figure 1 The CT image shows presence of mixed enhancing solid and cystic lesions which arise from the pelvic cavity measuring 14 cm×14 cm×16 cm.

actin (figure 6) and CD34 reagent (figure 7) were all positive, while on CD117 and S100 reagents were negative.

OUTCOME AND FOLLOW-UP

Nonetheless, she showed good response during postoperative recovery. The surveillances of CT scans every 3 months were within normal findings. She was on regular clinic follow-up for clinical and CT scan surveillances for every 3 months to monitor signs and symptoms of local recurrence and metastasis. She is now free of recurrence and having good quality of life after 9 months of operation.

DISCUSSION

The mesentery is a common site of metastasis place for the gastrointestinal malignancy via lymphatics and lymph nodes. However, for the primary pathology arising from the leaves of the mesentery is very rare; it can be from the leiomyosarcomas,

Figure 2 Intraoperative picture of huge mesenteric tumour involving sigmoid mesocolon.

Figure 3 The huge tumour embedded together with left lower part of ureter (marked with blue vessel loop).

liposarcomas, fibrosarcomas, malignant teratomas or haemangiopericytomas.⁵ Previously, mesenteric leiomyosarcoma was classified in the same group of gastrointestinal stromal tumour, which is the most common mesenchymal tumour of the gastrointestinal tract. However, leiomyosarcoma is a mesenchymal malignant tumour of the mesentery that exhibits common immunohistochemistry patterns such as C-kit (CD117), CD34 or actin.⁶ Hence, the true smooth muscle malignancies are currently very uncommon and its clinicopathological characteristics remain unclear.

Mesenteric tumour is arising from mesodermal origins of the mesentery of blood vessels, fibrous tissue and nerves.^{7,8} At the early stage, this tumour is unable to be detected, and the symptoms of these tumours mainly are asymptomatic, or with a palpable mass, abdominal distension and pain.⁸ The leiomyosarcoma involving the gastrointestinal tract will be presented with gastrointestinal bleeding and pain.⁹ Yannopoulos and Stout first reported a case of mesenteric leiomyosarcoma as they described that most of these tumours tend to be asymptomatic and grow to a large-sized intraperitoneal cavity without any intestinal

Figure 4 Caldesmon immunohistochemistry showed positive staining.

Figure 5 Desmin immunohistochemistry revealed positive staining.

obstruction.¹⁰ Leiomyosarcoma exhibits high mitotic activity, anaplasia and cellular differentiation that influenced the malignant behaviour of these tumours. Ranchod and Kempson showed frequency of mitoses to be the most useful indicator of malignant potential that determined tumour highly to develop local recurrences and distant metastasis involving peritoneal, hepatic and lung.¹¹ Hence, the careful postoperative follow-up is a must to detect an early stage of recurrence or metastasis.

Lee suggested that the surgeons should attempt wide radical margin removal of all smooth muscle tumours which is at least 4 inches of healthy tissue on all sides of the tumour involving mesentery.¹² Hence, the oncological resection is achieved to improve overall survival and reduced local recurrence. Patients with leiomyosarcoma of mesentery generally display a poor prognosis. Five-year survival rate is only 20%–30% and the complete surgical resection will favour the outcome. Case series and literature review by Kato *et al* in 2016 stated that since 1998, it was 13 cases which have been reported so far by proper immunohistochemistry.¹³ Kato *et al* added that the recurrence can occur within 5 years, and periodic monitoring is recommended using imaging modality to monitor the progression following the surgical resection.

Figure 6 Smooth muscle actin strongly stained positive result.

Figure 7 CD34 staining highlighted.

Hepatic metastasis is common in duration of few months to several years after initial surgery, and there is also one case of cutaneous metastasis reported after 3 months of tumour resection.¹⁴ The surgery is a mainstay of treatment for this type of tumour as it is notoriously resistant to radiotherapy and chemotherapy.¹⁵ Earlier detection by CT together with complete oncological resection may improve long-term prognosis in patients with primary leiomyosarcoma of mesenteric origin.¹⁶ Due to its rarity and low incidence, the standard chemotherapy regimen for recurrence and metastasis is not yet established.^{14 17}

Learning points

- ▶ The primary mesenteric leiomyosarcomas are extremely rare tumour in gastrointestinal malignancy.
- ▶ Complete oncological surgical resection is mandatory and the diagnosis is concluded by the histopathological examination especially by immunohistochemistry staining.
- ▶ Adjuvant chemoradiotherapy had a limited role in patients with poor prognosis.
- ▶ The postoperative surveillance is a must due to its possibility of local recurrence and metastasis tendency.

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