

# Surgical case report of uterine leiomyosarcoma metastasising to the pancreas resected by enucleation

Daniela Zammit, Charles Cini, Jonathan Cutajar

## Abstract

A 64-year old lady presented with worsening abdominal pain, vomiting and constipation. She had diffuse abdominal tenderness with peritonism, requiring an emergency laparotomy as a result of a perforated sigmoid tumour. An incidental hard lump was identified on the anterior surface of the pancreas and was removed by enucleation. It was later diagnosed as metastatic leiomyosarcoma based on histology and from her history of uterine malignancy. No recurrence is reported up to this day.

Uterine leiomyosarcomas are aggressive malignant tumours with a high predisposition to metastasis, commonly to the lungs, liver, brain and bone. Metastasis to the pancreas is a rare occurrence and considered highly unusual, which can typically present with non-specific symptoms and signs. Imaging can pick up a pancreatic lesion and a radiologically-guided FNA as a pre-operative attempt is acceptable in order to differentiate the lesion before undertaking any major surgery. However, in view of only a few case reports found in the literature, surgical management of pancreatic metastases is not clearly defined with a questionable long-term prognosis. Most cases are managed by elective radical excision with good result.

Review of the literature shows other more radical surgical approaches were used. This is the first report of metastatic uterine leiomyosarcoma to the pancreas being managed by enucleation, with a successful follow-up and no recurrence.

## Keywords

Uterine leiomyosarcoma, Pancreatic metastasis, Emergency surgery, Enucleation, Malignancy

## Presentation of case

A 64-year old lady has a past history of high-grade uterine leiomyosarcoma for which she had undergone total abdominal hysterectomy & bilateral salpingo-oophorectomy three years prior to this presentation. At operation, no obvious metastases were noted and it was confirmed as confined to uterine wall. Following surgery, the patient underwent radiotherapy.

In August 2015, the patient presented with a 1-week history of worsening abdominal pain associated with nausea and vomiting, weight loss and anorexia. She also admitted to passing small amounts of loose stools with a 10-day history of constipation. On examination, she had diffuse

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## Case Report

abdominal tenderness with lower abdominal peritonism. Blood investigations revealed a normocytic, normochromic anemia, leukocytosis, neutrophilia, thrombocytosis and hypoalbuminemia. An initial Computed Tomography (CT) scan showed thickening with perforation of the rectum and two lung lesions were noted measuring 0.4 and 0.6cm in the right lung base. She was taken for emergency laparotomy and a Hartmann's procedure was performed. A loop of small bowel invaded by the tumour (which also had palpable lymph nodes in its mesentery) was resected and side-to-side ileal anastomosis was done. The anterior wall of the lesser sac (i.e. the gastro-colic ligament) was opened and a hard lump was noted on the anterior aspect of the pancreas. This was locally removed by

enucleation and local haemostasis achieved. No further abdominal deposits were noted.

Gross morphological assessment of the lump removed from the anterior aspect of the pancreas showed a firm nodule measuring 24mm x 20mm x 16mm. On sectioning, it had a white whorled cut surface. Histology of the lesion showed the appearance of a spindle cell tumour with marked nuclear pleiomorphism and bizarre nuclei, with brisk mitotic activity, adjacent to normal pancreatic tissue. The tumour cells resulted as smooth muscle actin (SMA), desmin and myosin positive on immunohistochemistry, consistent with a diagnosis of leiomyosarcoma (*Figure 1 & 2*).

**Figure 1:** (H&E x400 magnification). Spindle cell tumour with a fascicular arrangement, showing marked pleiomorphism and numerous bizarre giant cells. The histology of the lesion is identical to that of the uterine leiomyosarcoma diagnosed in the same patient three years prior during hysterectomy.

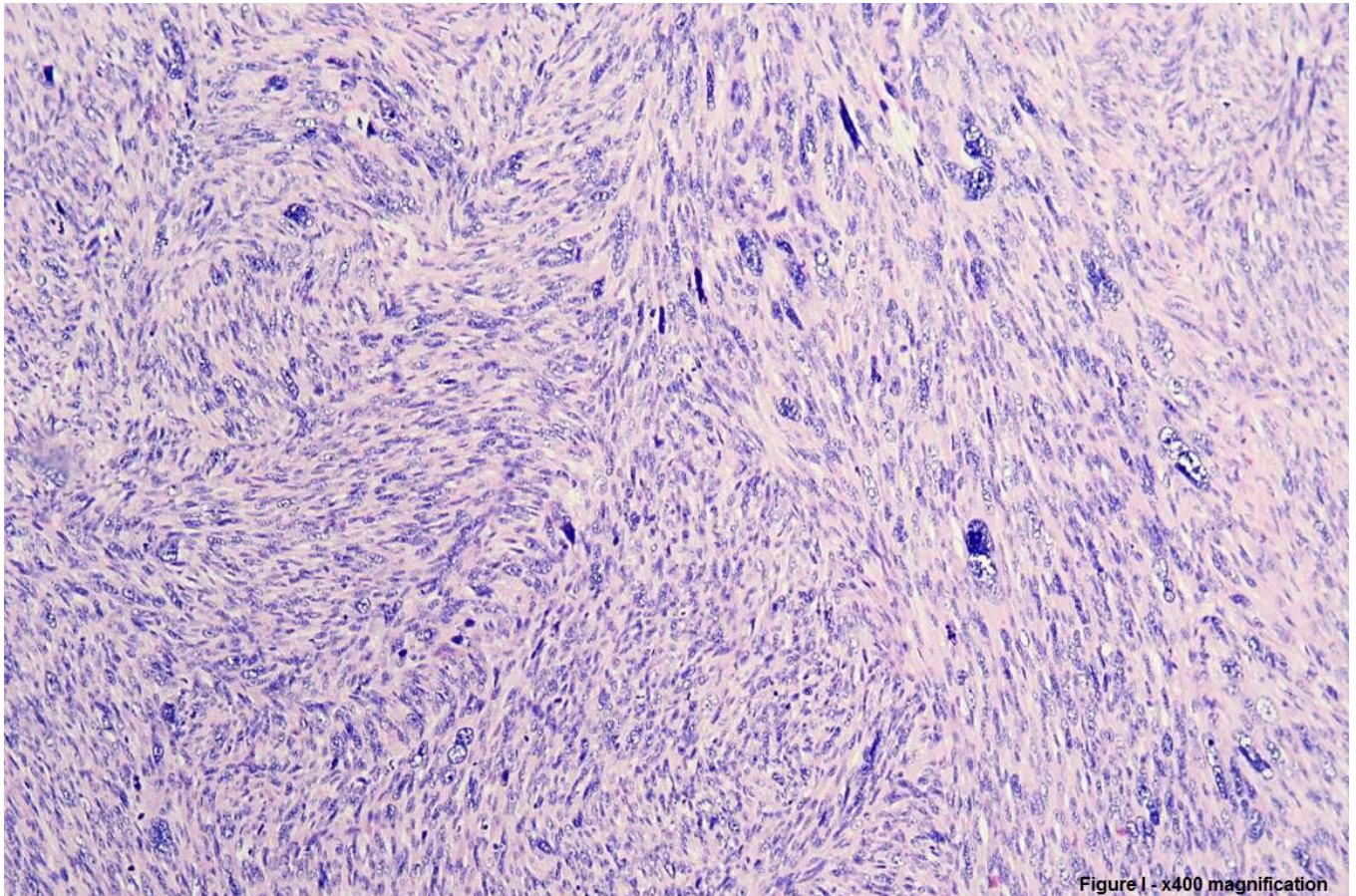


Figure 1 - x400 magnification

## Case Report

**Figure 2:** (H&E x100 magnification) A thin fibrous capsule separates the leiomyosarcoma in the top right half of the image from the normal pancreatic tissue in the bottom left corner, which allowed the lesion to be locally excised out.

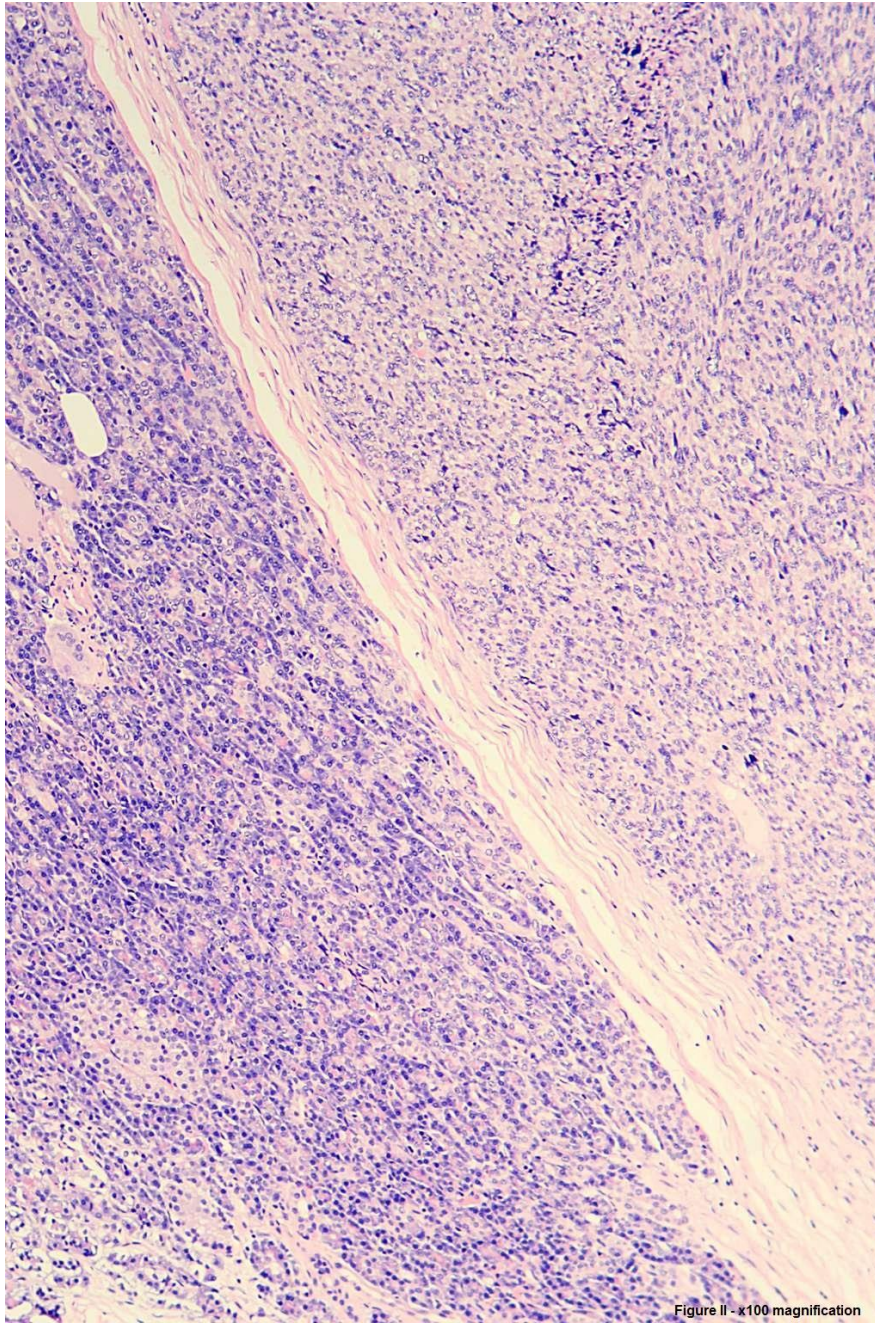


Figure II - x100 magnification

The intestinal specimens on the other hand showed metastatic intestinal adenocarcinoma over small bowel mesentery, together with moderately-to-poorly differentiated intestinal adenocarcinoma in the large bowel, reaching the radial margin. Significantly enlarged lymph node metastases and perinodal infiltration (11 out of 20 lymph nodes positive for metastasis) were also present (pT4b N2b M1).

During a multi-disciplinary discussion with

the pathologist and oncologist, it was agreed that the patient was too frail to undergo further radical surgery to address the pancreas and was continued to be followed-up from the intestinal adenocarcinoma point of view.

### Discussion

Uterine leiomyosarcoma is a malignant tumour arising from the myometrial smooth muscle.

It has an aggressive course which requires extensive therapy, necessitating total abdominal hysterectomy with bilateral salpingo-oophorectomy together with regular follow-up in view of its high risk of recurrence. The most common sites of metastasis include the lung, kidney and the liver.<sup>1-2</sup> Apart from the uterus, leiomyosarcomas can also arise from the ovaries, veins, spermatic cord, intestine, retroperitoneum and soft tissue.<sup>3</sup>

Leiomyosarcomas to the pancreas may present as large cystic lesions on imaging. They can be easily mistaken for a pancreatic pseudocyst rather than a malignant lesion. However, the latter are usually more prevalent in men and with a history of gallstones or pancreatitis.<sup>4</sup> In this particular case, the patient is a female with no relevant past history of gallstones or pancreatitis. Moreover, one must keep in mind her past gynaecological history of uterine leiomyosarcoma.

The pancreas is an uncommon site for metastasis to occur, even more so for metastatic leiomyosarcoma of the uterus which is considered as a rare occurrence. Only a few cases have been reported in the literature. Autopsy studies by Nakamura *et al.* have shown that the most common pancreatic secondary tumours are carcinomas, specifically adenocarcinomas. The stomach is the most common primary tumour site, followed by the lung and extra-hepatic bile duct.<sup>5</sup> On the other hand, Adsay *et al.* suggested that lung cancer is the most common source of metastasis to the pancreas.<sup>6</sup>

In the literature such metastatic lesions are typically diagnosed on routine follow-up CT scans, and then excised during elective surgery. They had resorted to radical operations, including pancreaticoduodenectomy, distal pancreatectomy, segmental resection and if not possible to preserve pancreatic tissue, total pancreatectomy has also been reported. In this article, we reviewed a case report where the lesion was noted during emergency surgery and resected by enucleation.

Macroscopically, these lesions show a white whorled surface. Microscopically, they are characterised by interlacing spindle-shaped cells. The mitotic count would give an indication of the level of aggressiveness of the tumour. Other features to look out for include cellularity, degree of atypia and pleiomorphism and presence of myofibrils.<sup>7-8</sup> It is believed that leiomyosarcomas of the pancreas arise from smooth muscle of the pancreatic blood vessels or pancreatic ducts.<sup>9</sup>

In view of lack of standardised guidelines for the management of pancreatic metastases, one must use clinical judgement after taking into consideration the patient's co-morbidities and discuss it in a multi-disciplinary setting. A wide negative resection margin during surgery would ensure complete excision and increase the chances of survival and quality of life.<sup>10</sup> In this particular case, a less radical approach of removing the tumour by enucleation was adopted, and to date, no further recurrence has been reported.

### Conclusion

Review of the literature shows other more radical surgical approaches were used. This is the first report of metastatic uterine leiomyosarcoma to the pancreas being managed by enucleation, with a successful follow-up and no recurrence.

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## Case Report

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