Abstract:
We report a patient with a jejunal leiomyosarcoma who presented with fever of unknown origin. Resection of the tumour resulted in resolution of his symptoms. Jejunal leiomyosarcoma is a rare tumour consisting of smooth muscle cells. Patients with this type of smooth muscle cell tumour generally present with vague symptoms such as abdominal pain, chronic anaemia, diarrhoea and because of this and the low sensitivity of most diagnostic studies - for example, enteroclysis, computed tomography, and echography - jejunal leiomyosarcoma is notoriously difficult to diagnose at an early stage.

Histologically, differential diagnosis between leiomyoma and leiomyosarcoma is not clear cut, as it is based on a number of differentiating features with a wide overlap. The tumour typically spreads by direct invasion and haematogenously to the liver, lungs, and bone. The only proved treatment is resection, with a five-year survival of 40-60 per cent.

Keyword: LEIOMYOSARCOMA, PERFORATION

Case Report: A 38-year-old man was referred to this hospital because of fever of unknown origin and abdominal pain. His symptoms had begun one week earlier with episodes of rigor and a temperature followed by general fatigue. He had vomiting for 1 day. In addition he complained of anorexia and weight loss of 4 kg in the previous six months. While the patient admitted some episodes of melaena, but no haematemesis or haematuria. On examination...
pt was febrile with signs of an acute abdomen. Chest, abdominal radiographs were normal. A computed tomogram of the abdomen showed a large (8 x 5.5 cm) mass in the right iliac fossa that seemed to be in contact with the small intestine. There was no sign of retroperitoneal lymph node enlargement, and the mass showed central necrosis. A laparotomy was performed, with resection of a necrotic perforated small intestinal tumour at a distance of 100 cm from the ileocaecal valve, on the antimesenteric aspect. The tumour was adherent to the urinary bladder and the sigmoid. There were no signs of liver or peritoneal cavity metastases. Postoperative progress was normal. In the subsequent 6-month follow-up with CT thorax, abdomen there was no evidence of recurrence or metastasis. The macroscopic examination showed a nodular, fleshy white-gray mass with a diameter of 6.5 cm and a central cyst formation that contained brown sterile fluid. Microscopic tumour examination showed well differentiated smooth muscle cells with slightly pleomorphic, hyperchromatic nuclei and abundant eosinophilic cytoplasm. The number of mitoses was > 5 per 10 high power field. There was invasion of the mucosa as well as the serosa with local perforation and signs of localised peritonitis. The edges of the resected small bowel fragment consisted of normal intestinal tissue. The resected lymph nodes were normal. It was reported as leiomyosarcoma. Discussion A leiomyosarcoma of the jejunum presenting with bouts of spiking fever has not been described recently in the published reports. The tumour usually presents with melaena, because of ulceration, or with abdominal cramps, signs of subacute obstruction, or volvulus. Chronic diarrhea or, rarely, spontaneous perforations also occur. In our patient central necrosis with localised peritonitis as a consequence of a small perforation probably caused the presenting symptom - fever.

REVIEW OF LITERATURE We found, in published reports, references to leiomyosarcomas or leiomyomas of other organs presenting with fever - surgical removal of an uterine myoma, a leiomyosarcoma of the lung with fever, cough, and dyspnoea; a renal leiomyosarcoma with signs of pyelonephritis; and a leiomyosarcoma of the spleen presenting with fever. In the gastrointestinal tract there were reports of a large bowel leiomyosarcoma, a stomach leiomyosarcoma that caused fever and anaemia, and an oesophageal leiomyosarcoma that presented with fever, cough, pyrosis, and dysphagia. The most sensitive examination is reported to be a small bowel enteroclysis, which commonly shows an extraluminal mass with a crater like defect. The extraluminal growth is a possible explanation for the absence of any sign on our small intestinal x-ray examination. An abdominal computed tomogram is useful for detecting large tumour masses, as in our case, but it may also miss smaller lesions. An abdominal ultrasound examination may also show large lesions, but echographic examination of the small intestine is often negative because of artefacts caused by air in the intestine. Selective angiography is useful for delineating subserosal tumours and those that are actively bleeding. The only proved treatment for leiomyosarcomas of the jejunum is wide excision with 5 cm margins of the tumour with all areas of invasion. There are no conclusive reports that adjuvant radiotherapy or chemotherapy is helpful. The five year survival after en bloc resection.
shows only slight variation between most series: 50% to 55%.
The spread of small intestinal leiomyosarcomas is primarily by direct invasion, as well as by the haematogenous route. Most metastases occur in lungs, liver, and bone. Only a few cases of lymphatic spread have been reported. In our case, the routinely resected lymph nodes did not show any sign of tumour, nor did we find any indication of metastases. The margins were free. Histologically, the distinction between benign and malignant tumours is difficult to make and classification of malignancy is unreliable. However two of the strongest pathologic predictors of malignant behaviour are size (generally > 5 cms) and mitotic count (generally > 5 mitoses per high power field).

Conclusion Thus all GSTs should be considered a low grade malignancy with a small risk of recurrence and metastasis. Local excision with negative histological margins is recommended because of the low malignant Patients with a very large GST (> 5 cms) should have life long follow up including periodic CT scan, due to risk of recurrence and metastasis. These principles should be followed whether the lesion is termed a GST or leiomyoma/leiomyosarcoma.