

Case Report

Gigantic jejunal leiomyosarcoma

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ABSTRACT

Jejunal leiomyosarcoma is a rare neoplasm. We report a case of gigantic leiomyosarcoma of the jejunum in a 30-year old man who presented with abdominal distension, pain and constipation. Computerized tomography scan revealed a giant tumor filling the entire abdominal cavity. Fine needle aspiration cytology was suspicious. At laparotomy, a huge jejunal leiomyosarcoma measuring 30 x 25 x 19 cm and weighing 13 kg was completely excised. To the best of our knowledge, this is the first case of a giant jejunal leiomyosarcoma.

Keywords: Jejunal leiomyosarcoma, giant.

Saudi Medical Journal 2001; Vol. 22 (4): 369-371

Gastrointestinal leiomyosarcomas are rare tumors. They arise most frequently from the ileum, the jejunum being the 2nd most common site of involvement. They are slow growing tumors that rarely metastasize. Ultrasound and computerized tomography guided biopsy are useful diagnostic techniques, and the best treatment is wide surgical excision with free margins. Adjuvant therapy is indicated for a selected group of patients. We have completely excised a huge jejunal leiomyosarcoma measuring 30 x 25 x 19 cm and weighing 13 kg.

Case Report. A 35-year old man presented with a slowly growing abdominal mass, which had been present for one year. It was associated with loss of weight, loss of appetite, constipation and dyspnea. There was no vomiting. Systemic review was unremarkable. On examination, the patient looked cachectic. There was no pallor, jaundice or lymphadenopathy. Abdominal examination revealed a huge pelvi-abdominal mass involving the whole abdomen. It was mildly tender on deep palpation and cystic in consistency. The rest of the

examination was irrelevant. Basic laboratory investigations (Complete blood cell count, urea, creatinine, electrolytes, liver function tests, clotting profile, chest x-ray and abdominal x-ray) were within normal limits. Barium enema showed smooth, narrowed left colon, which was displaced medially and downwards. Intravenous urogram (IVU) was normal. Contrast computerized tomography (CT) scan of the abdomen and pelvis demonstrated a huge multi-septated cystic mass extending from the left hypochondrium to the pelvis (Figure 1). Fine needle aspiration cytology revealed suspicious cells. Laparotomy was carried out and showed a huge cystic mass measuring 30 x 25 x 19 cm filling the entire abdomen and displacing the left colon inferiorly. A loop of jejunum was adherent to the mass (Figure 2). There were no disseminated peritoneal or liver metastases. Dissection was facilitated by aspiration of 4 litres of dark red fluid. The whole mass was completely excised with excision of the adherent loop of jejunum. There was no residual disease. Intestinal continuity was restored by end-to-end jejuno-jejunostomy. The

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Received 19th September 2000. Accepted for publication in final form 10th December 2000.

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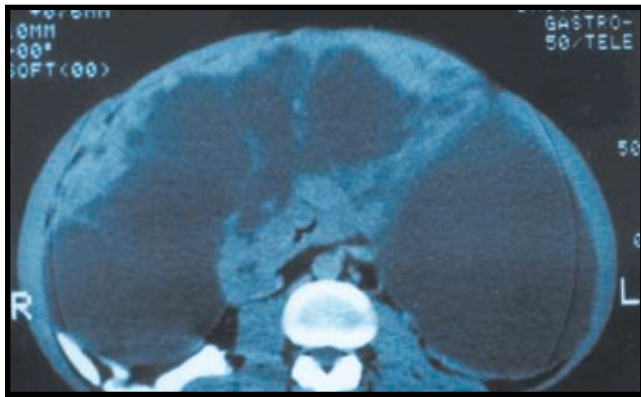


Figure 1 - Computerized tomography of abdomen revealing a huge multi-septated mass involving the whole abdomen and pushing the bowel to the right.

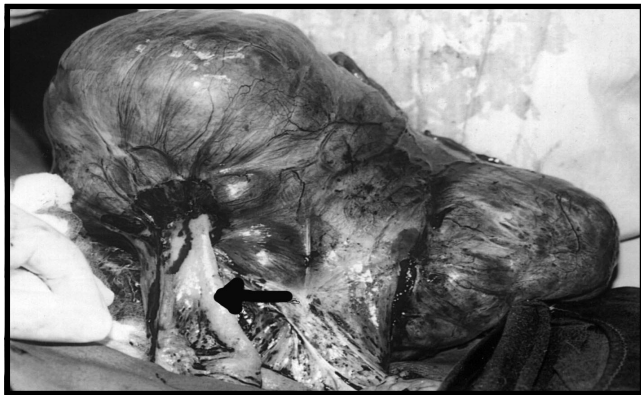


Figure 2 - Huge jejunal leiomyosarcoma which is highly vascular and arising from a loop of jejunum (arrow).

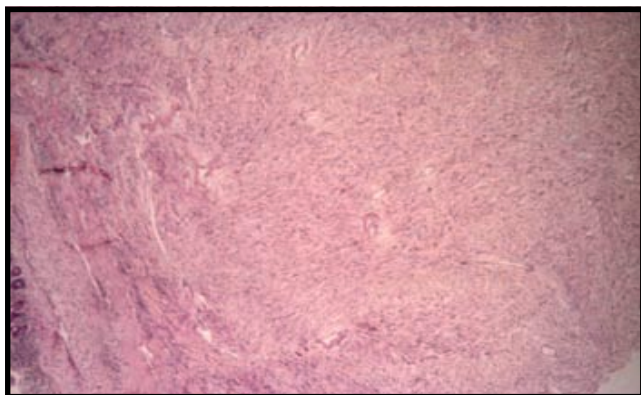


Figure 3 - Low-grade jejunal leiomyosarcoma showing focal ulceration of the mucosa with presence of spindle cell tumor in muscularis extending to the serosa (Hematoxylin and Eosin stain x 25).

patient had an uneventful post-operative recovery. No adjuvant chemotherapy or radiotherapy was given. Histopathological examination revealed a spindle cell tumor extending from the musculosa to the serosa of the jejunum. There were hypercellular interlacing bundles of smooth muscle cells with ample eosinophilic cytoplasm, large atypical nuclei with the presence of multinucleated giant cells. Mitosis was less than 5/10 high power field. These features are compatible with low-grade leiomyosarcoma of the jejunum weighing 13 kg, with free surgical margins (Figure 3).

Discussion. Leiomyosarcoma arise from malignant smooth muscle elements. They are rare small intestinal tumors.¹ Leiomyosarcomas are the most common intestinal sarcomas and represent 10-20% of all malignant tumors of the small bowel.² They often exhibit protracted, slow growth and rarely metastasize.¹ The symptoms of these tumors are variable and non-specific and depend on their size, location and histology. These include abdominal pain, abdominal distension, intestinal obstruction, gastrointestinal hemorrhage or even hemoperitoneum.² The jejunum being the 2nd most common site of involvement after the ileum. Leiomyosarcomas usually grow in an intramural location with serosal extension and the neoplasms often become large masses that invade outside the intestine. They often develop ischemic necrotic centers as they frequently outgrow their vascular supplies. This can lead to fistula or abscess formation. Frequently their size is large with more than 75% being larger than 5 cm in diameter.³ Tominaga⁴ et al reported a jejunal leiomyosarcoma measuring 30 x 25 x 18 cm and weighing 4.934 kg.⁴ In our patient, the tumour was measuring 30 x 25 x 19 cm and weighing 13 kg. To our knowledge, this is the first case reported of jejunal leiomyosarcoma of such a huge size and weight. Ultrasonic guided biopsy or needle aspiration cytology are helpful.^{4,5} Computerized tomography guided core needle biopsy is a safe and accurate procedure and is indicated in selected patients where endoscopic biopsy is not possible.⁶ Surgical resection should always be carried out with complete macroscopic and microscopic tumor resection.⁷ The tumor grading is the major prognostic determinant.⁸ No definite role of adjuvant chemotherapy or radiotherapy has been established. However, surgery alone is inadequate for high-grade tumours and adjuvant therapy should be considered in these patients.⁸

In conclusion, we report a case of a giant jejunal leiomyosarcoma that was successfully resected. Complete surgical excision with free surgical margins is the optimal therapy.

Acknowledgment. We would like to express our great thanks to Mrs. Joy A. De Silva.

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