DUODENAL LEIOMYOSARCOMA: A REPORT OF A RARE AND AGGRESSIVE TUMOUR

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ABSTRACT

A 45-year -old Malay lady who presented with intermittent abdominal pain and a left hypochondrial mass was found to have a 10x8x5 cm duodenal tumour without local invasion at laparotomy. En bloc resection of the tumour with adequate margin of clearance was done and histopathological diagnosis of low grade leiomyosarcoma was made. Fourteen months later, she returned with multiple metastases in the liver and needed palliative chemotherapy for pain relief. Duodenal leiomyosarcomata are very rare tumour. Their prognostic indicators include biological grading, tumour size and presence of metastases. Recognition of its high malignant potential calls for close surveillance calls even after apparent curative surgery.

Keywords: Duodenum, leiomyosarcoma, liver metastases

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INTRODUCTION

Retrospective studies over three decades involving over 10,000 patients with gastrointestinal malignancies showed that only 1.6% to 2.4% of them originated from the small bowel[1,2]. The rare occurrence of small bowel malignancies, in contrast to the adjacent stomach, esophagus and large bowel is puzzling. Several theories had been put forward to explain its rarity. Among them are rapid transit time through the small bowel, fluidity and alkalinity of its content, its relative sterility and presence of local detoxifying enzymes such as benzpyrene hydrolase and protective factors like secretory IgA[2-4]. The latter theory, suggesting the presence of protective immune system in the small intestine, is supported by the observed increased incidence of small bowel tumour in patients treated with immunosuppressive drugs and the reported increased incidence of second tumours associated with small bowel malignancies[1,3,4]. Leiomyosarcoma which makes up only 9-20% of all the small intestinal tumours is thus exceedingly rare[1,2,5].

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CASE REPORT

A 45-year-old Malay lady who presented with six months history of intermittent abdominal pain was found to have a mobile left hypochondrial mass measuring 6x8 cm clinically. There was no history or appetite or weight loss or history suggestive of gastrointestinal disturbance or bleeding.

She had undergone a total abdominal hysterectomy for uterine leiomyoma a year before. Except for the presence of pallor, physical examination was otherwise unremarkable.

Blood investigations revealed hypochromic microcytic anemia of 8.8 gm/dl, subsequently confirmed to be due to iron deficiency. Her liver function tests and chest radiograph were normal. Ultrasound examination of the abdomen showed a cystic mass 8.1x6.3 cm in the region of the tail of the pancreas which was separate from the spleen and the left kidney.

At laparotomy, a large vascular, soft tumour measuring 8x10 cm was found arising from the fourth part of the duodenum. It was adherent to the body of the pancreas and the transverse colon but no invasion was noted. The duodenal tumour was excised en bloc with adequate margin of clearance. Postoperative recovery was uneventful and she was discharged well.

Histopathologic examination revealed a circumscribed tumour 10x8x5 cm in size at the fourth part of the duodenum which was made up of spindle shaped cell (Fig 1) and arose from the muscular layer of the duodenum. Occasional mitoses of 2 per 10 high power fields was seen. Vessels were free of infiltration and surgical margins were clear. A final diagnosis of low grade leiomyosarcoma was made.

The patient was followed up six monthly. Fourteen months after surgery she returned with severe right hypochondrial pain associated with vomiting for one day. Clinical examination revealed a grossly enlarged liver which was nodular, hard and non-tender. Ultrasonography showed two masses; one is echogenic measuring 5 cm and the other cavitating measuring 8 cm, in the right lobe of the liver (Fig 2). Computed tomography of the liver confirmed the presence of these masses. Ultrasound guided biopsy of the echogenic mass revealed neoplastic spindle cells proliferation with occasional mitotic figures adjacent to fibrous connective tissue stroma confirming the clinical suspicion of metastatic leiomyosarcoma. Palliative chemotherapy comprising Cis-platinum, Adriamycin and Vinblastine was administered for continued severe right

Fig 1 - Duodenal leiomyosarcoma. This highly cellular tumour shows spindle cell appearance and mitotic activity. (H.E x 400)

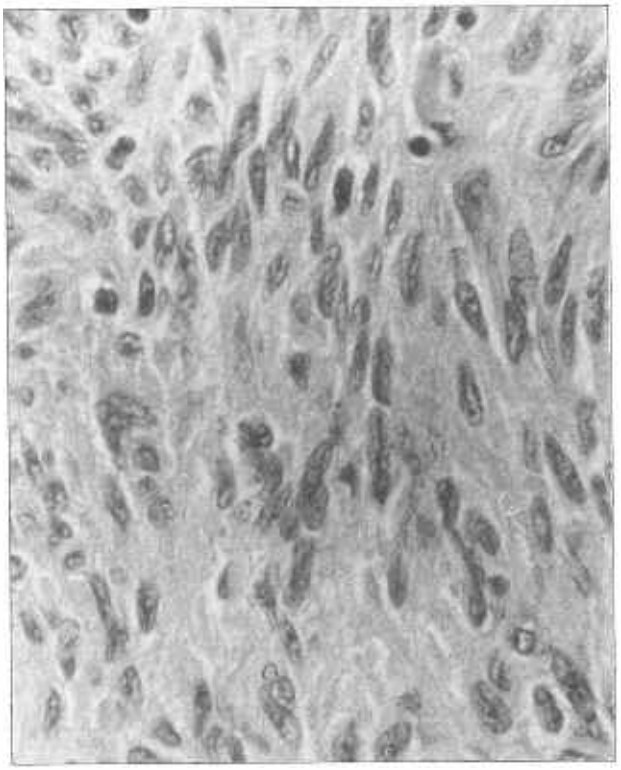
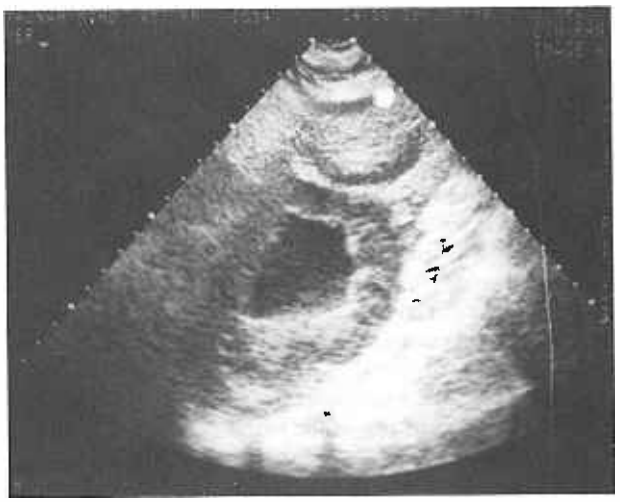


Fig 2 - Ultrasound of the liver showing an echogenic and a cavitating mass in the right lobe of the liver



hypochondrial pain in the context of inoperable multiple liver metastases and worthwhile palliation was achieved.

DISCUSSION

Intestinal leiomyosarcoma commonly presents as an abdominal mass, with abdominal pain or gastrointestinal bleeding[2-10]. Almost 60% of the gastrointestinal leiomyosarcoma reported was larger than 8 cm in diameter and over 40% were larger than 10 cm [6]. Surprisingly, in spite of the large size of many of these lesions, only 6-20% of them were reported to present with intestinal obstruction [6,7,10]. A probable explanation is their submuscosal origin and their extraluminal mode of growth [6-8]. As a result, this malignancy is often asymptomatic in the early stages and rather difficult and rather difficult to diagnose [6]. Our patient demonstrated the typical presentation of this rare malignancy and underlined the high frequency of diagnosis made only at operation.

Histopathologic grading of gastrointestinal leiomyosarcoma was characterized in terms of differentiation, cellularity, anaplasia or atypia and mitotic index[5]. Besides histological grading, the other prognostic indicators of leiomyosarcoma of the small intestine include presence or absence of the metastases and tumour size[6].

The overall 5-year survival rate of leiomyosarcomas of the small intestine had been reported to be 40-50%[5,10-13]. Other reports, however, suggest poorer prognosis[2,4,8].

It has also been reported that in those who survived complete resection of the tumour, 46% of them had recurrent disease at a median interval of two years[5]. It therefore suggests that this tumour is probably more aggressive than previously believed to be.

The relationship of the uterine myoma in this patient to her small intestinal leiomyosarcoma is unclear. Considering the high incidence of uterine myoma in the population, its coexistence with any disease is not unusual. However, multipotential connective tissue metaplasia and multiple primary smooth muscle tumours have been reported[14].

In conclusion, we report here a patient who came with the typical presentation of an extremely rare malignancy which had behaved more aggressively than previously suggested. Recognition of its high malignant potential calls for close surveillance even after apparent curative surgery.

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