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Intestinal leiomyosarcoma in childhood

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Abstract. A 4-month-old boy with leiomyosarcoma of the ileum presented to us with complete intestinal obstruction. Small gut resection was done. The child was well 9 months after surgery with no evidence of a recurrence.

Key words: Leiomyosarcoma – Intestinal tumour in childhood

Introduction

Intestinal leiomyosarcoma in childhood is rare. Angerpointner et al. reported a case in 1981 and on review of the medical literature, they could only find 14 more cases reported previously [1]. To this series, Faldella et al. added another patient in 1983 [2]. Recently we saw a 4-month-old boy with leiomyosarcoma of the ileum. This is the second case of leiomyosarcoma of the ileum so far reported and the first case that has occurred in a Chinese patient.

Case report

A 4-month-old boy was admitted to our hospital with abdominal distension and repeated vomiting during 1 day. He was well until about 1 month prior to admission when he developed spasmodic low grade fever, diarrhoea and had poor feeding. By the time of admission his size and body weight were below the third percentile. He was dehydrated and emaciated. The abdomen was grossly distended and a mass about 6 cm in diameter was felt in the lower abdomen. The rectum was empty with no 'red currant jelly' stool. A plain abdominal radiogram revealed an intestinal obstruction with a dilated small bowel and multiple fluid levels.

The child was rehydrated. At laparotomy, a fleshy tumour, 6 cm in diameter, was found originating from the wall of the ileum and growing almost totally in an extraluminal direction [3]. This tumour created an acute bend in the ileum causing a complete intestinal obstruction 10 cm from the ileo-caecal valve (Fig. 1). It also infiltrated outwards to involve the mesentery and the serosa of the jejunum at two sites. The mucosa of the small gut was free. There were multiple enlarged lymph nodes at the mesentery but there was no intra-abdominal metastasis. Small gut resection was done, about 70 cm jejunum and 10 cm ileum were left behind and a primary enteroenterostomy was done. Post-operatively the child developed diarrhoea on oral feeding. He was treated with intravenous feeding and later oral feeding was gradually resumed and the child tolerated it well. He was followed up regularly and was well with no recurrence 9 months after surgery.

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Pathology

Grossly the tumour was bosselated and fleshy. The cut surface was greyish-white with soft semitranslucent areas towards its periphery. There were some areas of haemorrhage and necrosis.

Microscopically the tumour was cellular and composed of spindle-shaped cells. The cells were arranged in interlacing bundles (Fig. 2), sometimes in whorls. The cells and the shape of the nuclei were irregular in some places. Mitotic figures varied in frequency in different areas. In some the mitosis was zero and in others it was up to 15 in ten high power fields. The peripheral areas showed granulation type tissue, with many blood vessels and inflammatory cell infiltration. It is this granulation type tissue that invaded the jejunal serosa. The enlarged lymph nodes showed reactive changes only.

On electron microscopy, the main bulk of the tumour showed cells of smooth muscle origin with varying degrees of

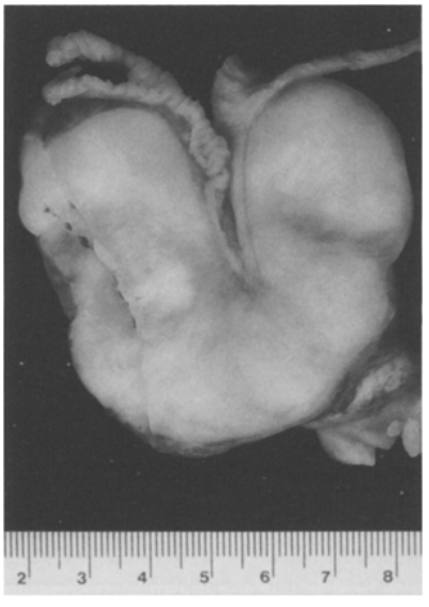

Fig. 1. An apple-shaped tumour on cut surfact showing areas of necrosis and haemorrhage

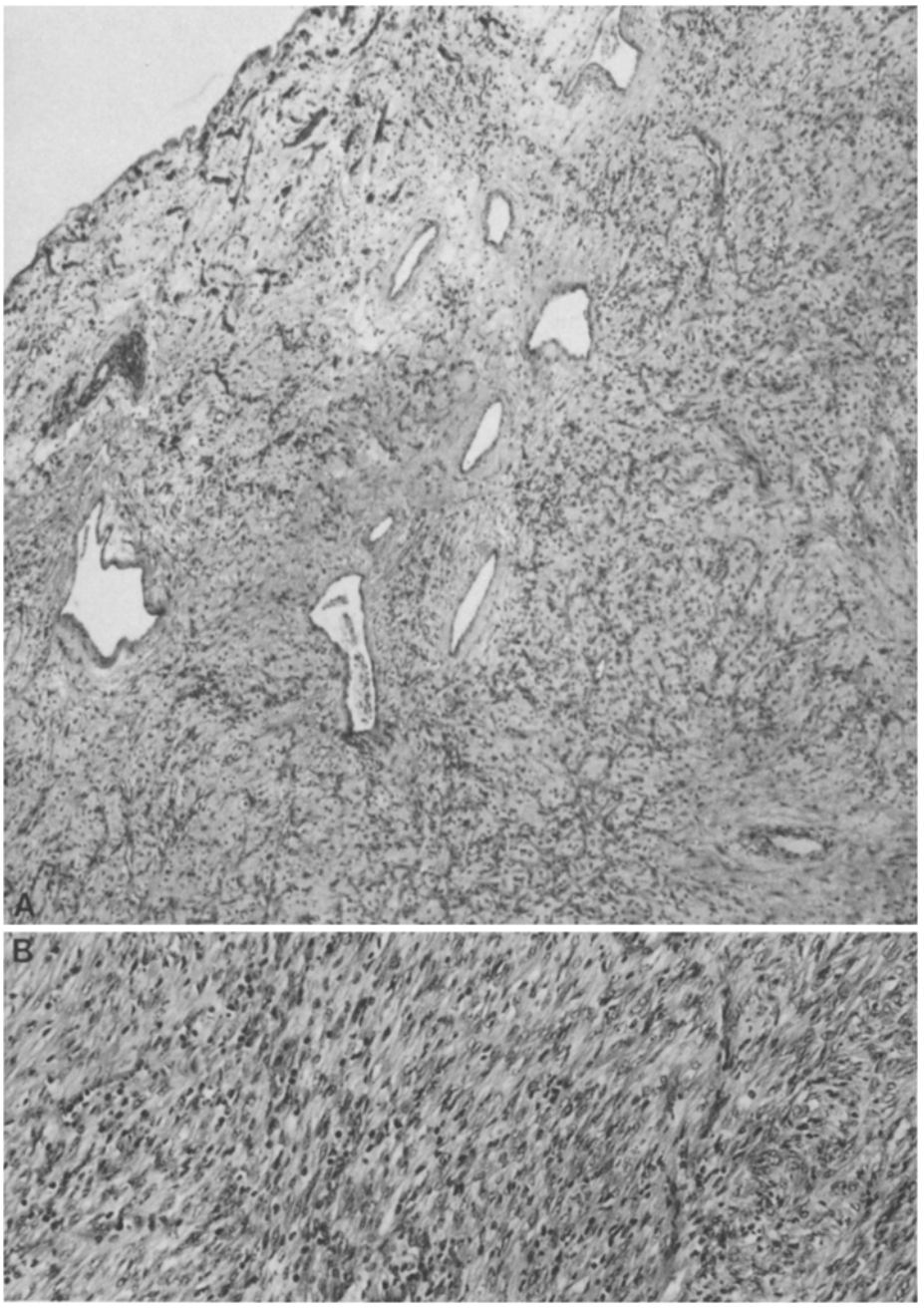

Fig. 2. A Granulation type tissue forming the peripheral of the tumour (H and E, × 10). **B** High power view of a more cellular area showing typical interlacing pattern of a spindle cell population of the tumour (H and E, × 40)

maturation; the peripheral granulation type tissue had neovascularisation, primitive smooth muscle cells and myofibroblasts.

Discussion

Intestinal leiomyosarcoma in childhood is rare. The age of discovery ranges from the first days of life up to 12 years and there is a marked female predominance in contrast to the reverse in adulthood [1, 2]. Addition of our male case to the series does not alter the position. Patients present with abdominal masses [1], intestinal obstruction [2], and in two cases the tumours presented with a jejuno-jejunal intussusception [4, 5]. This tumour has been reported to occur in the duodenum once, the jejunum six times, the ileum once, the Meckel's diverticulum once, the colon five times and the rectum twice [1, 2]. In our patient, it occurred in the ileum and this is the second case so far reported at this site. The first occurred in a 12-year-old girl in whom gut resection and anastomosis was done and the patient was well after 5 years [8].

Skandalakis and his associates stressed the difficulty in distinguishing malignant leiomyosarcoma from benign leiomyoma [8]. For the histopathologic diagnosis of a malignant leiomyosarcoma we use the criteria described by Ranchod and Kempson [6], namely frequency of mitotic figures, necrosis, size, cellularity, cellular atypia and evidence of invasion of surrounding structures. The electron microscopy of these leiomyosarcomas in young persons has been reported only in two instances [2, 7], and our case concurs with them in the main findings. However, local invasion by granulation type tissue in our case appears unique and worth a special mention.

There is agreement among all authors that the therapy of choice is radical surgery and tumours were resected in every case. Whether adjuvant chemotherapy or radiotherapy is necessary is still debatable [1, 9]. Because of the small number of intestinal leiomyosarcomas in childhood and the very few follow-up studies, little can be said about prognosis [1].

In our patient we decided not to give any adjuvant chemotherapy or radiotherapy because (a) radical surgery was done, (b) the effect of chemotherapy or radiotherapy on leiomyosarcoma is not yet proven in childhood, and (c) the other previous case with leiomyosarcoma of the ileum has had a long term survival.

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Even in momentous times, when everything is at stake, you do go on with your daily life as if nothing is happening.

Johann Wolfgang von Goethe (1749–1832) in "Elective Affinities"