ECRONICON OPEN ACCESS

EC CLINICAL AND MEDICAL CASE REPORTS

Case Report

Inflammatory Myofibroblastic Tumour of the Small Bowel - A Case Report

Kailas Bhandarkar P[1,2]*, Hemanshi Shah[3], Shallika Jayaswal[4], Sudhir Sighvi[5], Keshava Murthy[5] and Omprakash Makhija[5]

[1]Department of Paediatric Surgery, Evelina London Children's Hospital, UK

[2]Department of Pediatric Surgery, Topiwala National Medical college and B.Y.L. Nair Charitable Hospital, Mumbai, Maharashtra, India

[3]Professor and Head, Department of Pediatric Surgery, Topiwala National Medical college and B.Y.L. Nair Charitable Hospital, Mumbai, Maharashtra, India

[4]Associate Professor, Department of Pediatric Surgery, Topiwala National Medical college and B.Y.L. Nair Charitable Hospital, Mumbai, Maharashtra, India

[5]Senior Registrar, Department of Pediatric Surgery, Topiwala National Medical college and B.Y.L. Nair Charitable Hospital, Mumbai, Maharashtra, India

***Corresponding Author:** Kailas Bhandarkar P, Department of Paediatric Surgery, Evelina London Children's Hospital, UK and Department of Pediatric Surgery, Topiwala National Medical college and B.Y.L. Nair Charitable Hospital, Mumbai, Maharashtra, India.

Received: October 23, 2018; **Published:** June 25, 2019

Abstract

Inflammatory myofibroblastic tumours (IMT) are rare tumours and occur infrequently in children. Abdominal sites are less commonly involved. Complete surgical excision is needed to prevent local recurrence. Adjuvant therapy may be needed in cases where complete excision is not possible

Keywords: *Inflammatory Myofibroblastic Tumour; Abdominal; Recurrence; Adjuvant Therapy*

Introduction

IMT is a rare tumour and is usually solitary. IMTs occurring in intra-abdominal sites have rarely been described in children and may present with signs and symptoms of bowel obstruction. They are characterised by rapid growth, local invasiveness and recurrence. We report a 6 year old girl with abdominal IMT who presented with extensive bowel involvement and anaemia.

Case Report

A 6 year old girl was admitted with a history of abdominal lump since 6 months. There was no history of abdominal pain, vomiting or diarrhoea, bowel symptoms or fever. She was diagnosed at a private hospital as abdominal tuberculosis and commenced on anti-tuberculosis medications and then referred to us.

Clinical examination showed an anaemic young girl, pyrexial (38.4 C). Abdominal examination revealed a firm, mobile, non-tender mass in the umbilical region measuring 8 by 6 cm. Laboratory tests revealed a Hb of 6 g%. Ultrasound abdomen showed a mass which was not separate from bowel loops with calcifications. CT abdomen was performed to delineate the anatomy further. It showed that the mass was arising from the terminal small bowel encasing the distal small bowel. Calcifications were noted within the mass (Figures 1 and 2). The proximal small bowel was not dilated.

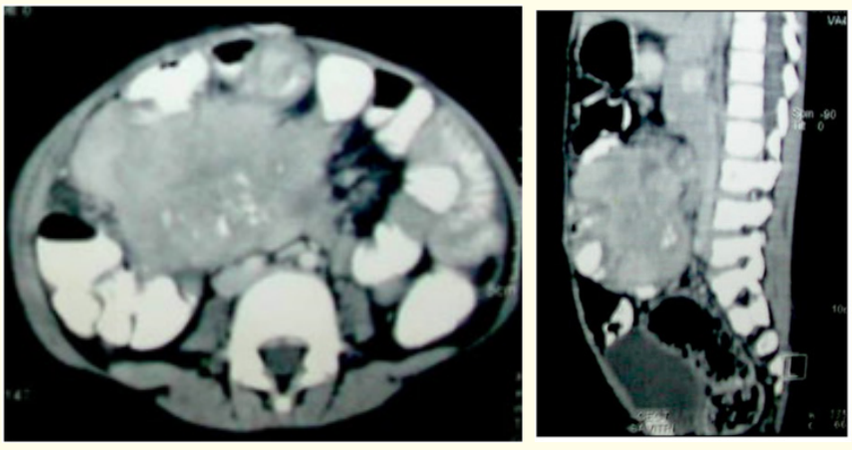

Figure 1 and 2: *CT scans of the abdomen showing a mass encasing the small bowel with calcification.*

Surgical exploration revealed a mass arising from the terminal ileum. Resection of the tumour mass along with the involved terminal ileum was performed (Figure 3) followed by an ileo-transverse anastomoses Histopathology revealed that the mass composed largely of spindle cells, diffusely invading the muscularis, submucosa and the lamina propria. There were abundant inflammatory cells composed of lymphocytes and plasma cells but no abnormal mitoses (Figure 4). She is on regular follow up and is without recurrence four years since her operation. Non-Steroidal anti-inflammatory medications (NSAIDs) were not used either before or after the surgery.

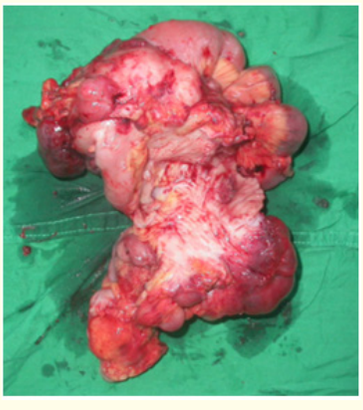

Figure 3: *Gross resected specimen-tumour mass with involvement of the small bowel mesentery.*

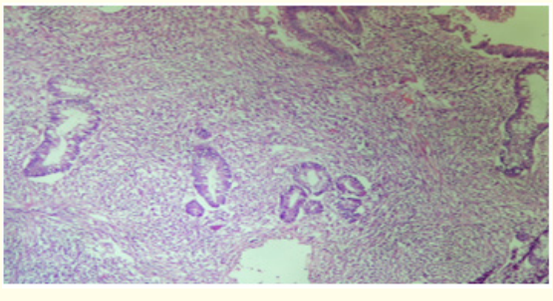

Figure 3: *Gross resected specimen-tumour mass with involvement of the small bowel mesentery.*

Discussion

IMTs are uncommon and may arise within numerous organs [1]. They are classified as tumours of intermediate biological potential by the most recent World Health Organisation classification. This is due to a tendency for local recurrence and a small risk of distant metastasis [2]. Several terms have been used to describe these tumours. They include inflammatory pseudotumour, inflammatory fibroid, pseudo-sarcomatous fibromyxoid tumor and inflammatory fibrosarcoma [3]. IMTs are more common in women than in men. Most studies quote an age range of 3 months to 46 years [4].

Common sites of presentation include lung, mesentery, liver, and spleen. Abdominal tumours are rare and may present with anaemia, intermittent abdominal pain, fever, weight loss, diarrhoea, dysphagia [5] or a palpable abdominal mass [6]. A study involving large series of patients showed that most arose from the abdomen, retroperitoneum and the pelvis [4].

These tumours can rarely present symptoms due to local invasiveness, recurrence or with distant metastasis [6]. Immunohistochemistry have shown IMTs are positive for anaplastic lymphoma kinase in approximately 50%, with positivity ranging from 36% to 71% in most large series [2].

Management includes complete surgical resection, observation, chemotherapy and radiation. Adjuvant treatment may be needed especially in cases of local aggressiveness, positive margins or if location of tumour is not amenable to surgical resection [1]. Complete surgical resection and close monitoring at follow-up are necessary to diagnose and treat recurrences early. The optimal management of locally aggressive and recurrent forms should be decided individually for each patient [6]. Local recurrences have been noted especially in those who had partial resection and did not receive adjuvant therapy. Tumour recurrence is unusual following complete surgical resection or organ-preserving combined modality therapy [1].

The recurrence rate varies according to anatomical site, from less than 2% for tumours confined to the lung, to 25% for extra-pulmonary lesions [7]. Recurrences are particularly common among intra-abdominal tumours and those in delicate anatomical locations such as the airway, possibly reflecting the difficult nature of complete surgical resection in these areas. However, recurrence is infrequent following complete excision of a solitary lesion [7]. Successful surgical management is possible even in cases of multiple recurrences [5]. Small study groups have reported successful treatment with NSAID. Non-responders were treated with surgical excision [8].

Conclusion

IMTs are rare group of locally invasive tumours. Those arising from the bowel are difficult to differentiate from more common types of bowel tumours. Complete surgical excision in needed for cure and to prevent local recurrence. In view of the potential for local recurrence, long term follow up is mandatory.

Bibliography

1. Kovach SJ., *et al*. "Inflammatory myofibroblastic tumors". *Journal of Surgical Oncology* 94.5 (2006): 385-391.

2. C Gleason and J L Hornick. "Inflammatory Myofibroblastic tumour: where are we now?" *Journal of Clinical Pathology* 61.4 (2008): 428-437.

3. Coffin CM., *et al*. "Inflammatory myofibroblastic tumor, Inflammatory fibrosarcoma, and related lesions: an historical review with differential diagnostic considerations". *Seminars in Diagnostic Pathology* 15.2 (1998): 102-111.

4. Coffin CM., *et al*. "Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases". *American Journal of Surgical Pathology* 19.8 (1995): 859-872.

5. Sanders BM., *et al*. "Inflammatory pseudotumor of the alimentary tract: clinical and surgical experience". *Journal of Pediatric Surgery* 36.1 (2001): 169-173.

6. Karnak I., *et al*. "Inflammatory myofibroblastic tumor in children: diagnosis and treatment". *Journal of Pediatric Surgery* 36.6 (2001): 908-912.

7. Cerfolio RJ., *et al*. "Inflammatory pseudotumors of the lung". *Annals of Thoracic Surgery* 67.4 (1999): 933-936.

8. Su W., *et al*. "Treatment of pseudotumors with nonsteroidal anti-inflammatory drugs". *Journal of Pediatric Surgery* 35.11 (2000): 1635-1637.

Volume 2 Issue 4 July 2019