Case Report

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Mesenteric inflammatory pseudo-tumour of the small intestine presenting with intestinal obstruction in a child: Case report and literature review

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ABSTRACT

We report a case of mesenteric inflammatory pseudo-tumour of the small intestine in a 4-year-old boy admitted with intestinal obstruction diagnosed from histopathology of 8 cm × 7 cm × 5 cm mass resected at laparotomy. We reviewed the literature and recommended complete resection with thorough histopathologic evaluation and long-term follow-up.

Key words: Children, inflammatory pseudo-tumour, intestinal obstruction, mesentery

INTRODUCTION

Abdominal masses in childhood have a very large spectrum of benign and malignant causes. Inflammatory pseudo-tumour (IP) arising from intra-abdominal sites has only rarely been described previously in children. Successful treatment requires careful radiologic and pathologic evaluation to distinguish IP from other lesions and complete resection. To the best of our knowledge, only seven cases of mesenteric IP of the small intestine have been reported in childhood, including our case. Here, we report a case of mesenteric IP of the small intestine presenting with intestinal obstruction and a review of the literature, focusing on diagnosis and treatment.

CASE REPORT

A 4-year-old boy was referred to our hospital with a 2-day history of abdominal pain. On admission, axillary temperature was 36.5°C, heart rate was 128 beats/min, and blood pressure was 98/56 mmHg. On examination, his abdomen was soft and flat, but tender in the lower abdomen. Laboratory investigations were unremarkable, with normal tumour markers. Ultrasonography (US) and computerized tomography (CT) identified an 8 cm diameter irregular heterogeneously enhancing mass in the pelvis that had caused a small bowel volvulus [Figure 1]. At laparotomy, an 8 cm × 7 cm × 5 cm mass was identified arising from the mesentery of the small intestine as the cause of intestinal obstruction [Figure 2]. The tumour and a 10 cm segment of the small intestine about 135 cm distal to the ligament of Treitz were excised, and primary intestinal anastomosis was performed.

Histopathological examination showed a non-malignant mass consisting of predominantly spindle cells containing elliptic and oval nuclei in an oedematous collagenous stroma [Figure 3]. There was an infiltration of varying amounts of inflammatory cells comprising plasma cells, lymphocytes, eosinophils and neutrophils. Immunohistochemically, spindle cells were negative for smooth muscle actin, S-100 protein, CD34 and AK-1. The diagnosis of IP was diagnosed using histopathologic findings. The post-operative course was uneventful, and our case is currently symptom-free 2 years after surgery with normal laboratory and radiologic findings.

DISCUSSION

Brunn first described IP in the lung in 1939, and it was so named by Umiker and Iverson because of its propensity to clinically and radiologically mimic a malignant process.[1] Their classification is controversial and confusing, and a variety of synonyms exists, including inflammatory myofibroblastic tumour and plasma cell granuloma.[2-5] The aetiology and

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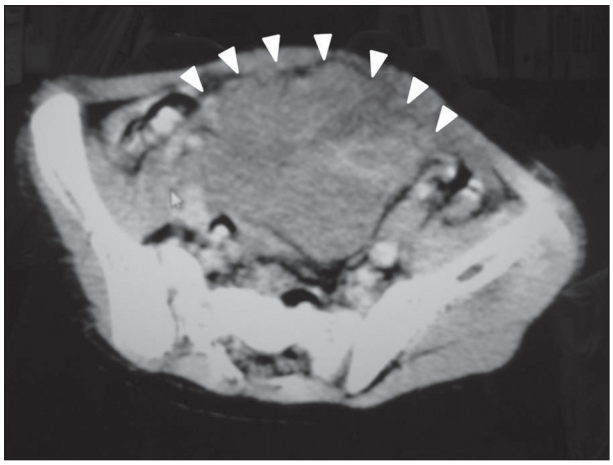


Figure 1: Computerized tomography. Arrowheads indicate the 8 cm irregular heterogeneously enhancing mass in the pelvis

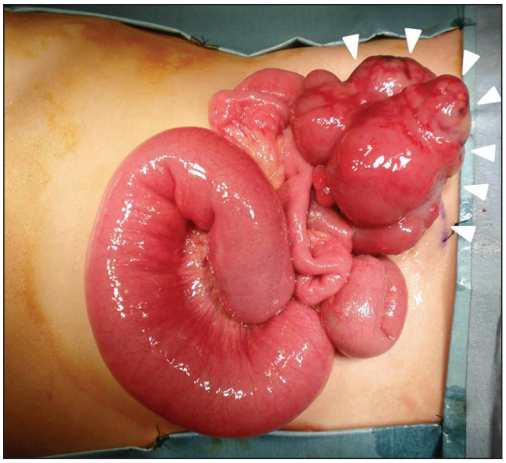


Figure 2: Intraoperative photograph. Arrowheads indicate the 8 cm × 7 cm × 5 cm mass arising from the mesentery of the small intestine

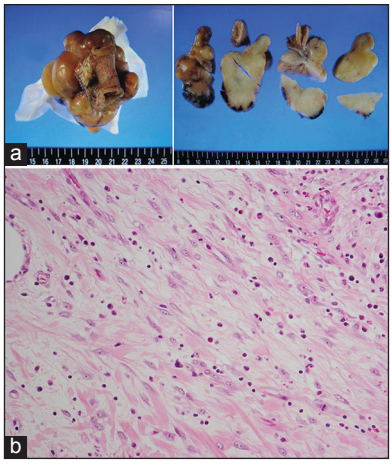


Figure 3: Histopathologic evaluation. (a) Macroscopic: Specimen comprising resected tumour with attached small bowel. The substance of the mass is firm, and yellow-white. (b) Microscopic: A non-malignant mass consisting of predominantly spindle cells containing elliptic and oval nuclei in an oedematous collagenous stroma (H and E, ×40)

cellular origin of IP remain unclear. Some authors have postulated that it may represent an immunologic response to an external agent that may or may not be infectious, while others believe it to be a true neoplasm.[6-9] Several associations have been reported between IP and previous abdominal surgery and lung infection.[6,10-12]

Macroscopically, IP is a firm, well-circumscribed, non-encapsulated, yellow-white mass that commonly adheres to and infiltrates surrounding viscera.[6,13] Microscopically, it is characterized by spindle-shaped cells that are mixed with chronic inflammatory component consisting of plasma cells, lymphocytes, and occasional histiocytes.[6,10] IP can be differentiated from soft tissue sarcomas by the characteristic cellular heterogeneity, presence of mature polyclonal plasma cells,[11] and lymphocytes, and absent or rare anaplasia and mitotic figures.[6,10,12,14]

Clinical presentation of IP depends on the site of the tumour. Mesenteric tumours usually present with a firm, smooth, painless mass associated with an inflammatory response, which is characterized by fever, impaired growth, weight loss, anaemia, elevated erythrocyte sedimentation rate, platelet count and hypergammaglobulinemia.[6,10,12] Recurrences occurred within 1 year of initial surgery and were treated by re-resection.[10,13] It has also been described as being associated with distant metastases although these are rare, and probably more a consequence of multifocal incidence rather than classic malignant metastatic spread.[6,11] Thus, while IP is generally accepted to be benign, multiple local recurrences, rarely encountered distant metastases, and tumour or complicated therapy-related deaths suggest there is a spectrum of activity that lies somewhere between inflammation and neoplasia,[6,12] although there have been no reports of IP undergoing malignant change.[6,12-16]

Of particular clinical importance is the differential diagnosis of mesenteric IP from malignant mass lesions. Unfortunately, there are no specific or characteristic clinical/laboratory findings, including tumour markers and imaging modalities that can distinguish IP from other neoplasms,[11] with IP appearing on US and CT as a well-circumscribed mass of homogeneous echogenicity with soft tissue density displacing or invading adjacent tissues.[17] Soft tissue sarcomas such as leiomyosarcoma, fibrosarcoma and malignant fibrous histiocytoma must be excluded and require careful histopathologic evaluation.[6,10,12,14] In other words, if there is the

Table 1: Mesenteric intestinal pseudo-tumour of the small intestine in children

Author	Date	Age (years)	Sex	Clinical presentation	Size (cm)	Procedure	Recurrence
Day[3]	1986	5	Male	Fever, frequent urination	10	TR	—
		9	Female	Abdominal pain	7	TR	—
		7	Female	Fever	8	TR	—
Souid[10]	1993	2	Male	Fever, constipation	20	TR	—
Ciftci[11]	1998	9	Male	Fever, abdominal pain	9	TR	—
Vaughan[18]	2005	15	Male	Abdominal pain	7	TR	—
Our case	2011	4	Male	Abdominal pain	8	TR	—

TR: Total resection

absence of cellular atypia, storiform pattern, frequent mitoses and bizarre tumour cells then a diagnosis of IP can be made.[11]

To the best of our knowledge, only seven cases of mesenteric IP of the small intestine have been reported in childhood, including our case [Table 1]. Mean age at diagnosis was 7.3 (range: 2-15 years); male to female ratio was 5:2. Mean tumour size was 9.8 cm (range: 7-20 cm). Total resection was performed in each case and considered curative with no further management such as immunomodulation or chemotherapy planned. No case died from neutropenic sepsis. The most frequent presenting symptom was abdominal pain (n = 4, including our case) and fever (n = 4).

Our case emphasized the importance of including mesenteric IP in the differential diagnosis of intestinal obstruction associated with an abdominal mass in a child. Because IP is a benign tumour with rarely encountered malignant characteristics such as local recurrence, and potential to 'metastasize', radical and unnecessary surgical procedures such as pelvic exenteration or amputation,[19] and potentially harmful therapy (chemo or radiotherapy) should be avoided because total resection would appear to be the treatment of choice according to reports in the literature.[6,10,12,19]

Post-operative management of IP cases is not covered in reports in the literature, and the appropriate duration of follow-up is unknown. Most recurrences occurred within 1 year of initial surgery although later recurrences have been described.[20,21]

Clinically indistinguishable from highly malignant neoplasms on diagnostic imaging, IP can be treated by complete resection and careful histopathologic evaluation will clarify the diagnosis through exclusion. Because of a known risk for recurrence, prolonged follow-up is mandatory, but outcome is expected to be good. With longer follow-up, the full clinical course of this condition will become more apparent. Meanwhile, awareness of IP will enhance the care and management of children presenting with an abdominal mass with acute signs.

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