

Case Reports

Retroperitoneal Inflammatory Myofibroblastic Tumour in Children: A Case Report and Review of Literature

JH ZHAN, XR LUO, ZW GUAN, XL HU, Y LIU, GQ BAO

Abstract Inflammatory myofibroblastic tumour (IMT) is a kind of rare soft tissue tumour. The most commonly affected organ reported is the lung. However, IMT can occur in any organ of the body. Although abdominal IMTs have been reported, retroperitoneal tumours are exceedingly rare. We here present a case of a thirteen-year old boy with a huge retroperitoneal mass diagnosed as IMT. The tumor was excised completely and no recurrence was found one year after surgery.

Key words Inflammatory myofibroblastic tumour; Retroperitoneal; Surgery

Introduction

Inflammatory myofibroblastic tumour (IMT) occurs commonly in the lung. Extra-pulmonary IMT in children have been described of its present in the mesentery, omentum, liver, bladder, mediastinum, head and neck, extremities, appendix, and kidneys, with the largest tumours occurring in the abdomen and retroperitoneum.¹ Tumours in the retroperitoneal origin with multinodular or ill-defined gross appearance are correlated with a more aggressive clinical behaviour in the form of local recurrence.² However, the definitive diagnosis of IMTs is seldom made before operation. Surgical resection is the primary treatment for IMT and remains the only therapy that can offer a chance of cure. Here, we report a case of retroperitoneal IMT in a

thirteen-year boy in whom the tumour was excised completely with no recurrence. A review of the current literature was also discussed.

Case Report

Our patient was a thirteen years boy with no significant past medical history and initially presented with abdominal distention, anaemia, and poor appetite for one month. Subsequent discussion revealed that the abdominal distension was not associated with nausea, vomiting, or diarrhoea. Physical examination showed a palpable, non-tender mass lesion, measuring 20 cm, in the central and left side of the abdomen.

An ultrasound scan of the abdomen confirmed a large complex mass lesion in the left side of the abdomen, displacing the transverse colon and the small intestines to the right side of abdomen. Chest X-ray did not show any elevation of the diaphragm and active lung lesion.

Complete blood count showed haemoglobin 9.8 g/dL; WBC: $5.17 \times 10^9/L$; HCT: 28.60%; platelet: $838 \times 10^9/L$; red cell indices were hypochromic microcytic anaemia. Liver and renal function tests were within normal limits. CA-199, carcinoembryonic antigen (CEA) and Alpha-fetoprotein (AFP) were also normal.

Abdominal computed tomography (CT) scan was subsequently performed. This showed a $25 \times 18 \times 10$ cm,

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well-defined, heterogeneous mass lesion in the retroperitoneum, with no involvement of the inferior vena cava and the aorta. No significant regional lymphadenopathy was noted (Figure 1).

The patient underwent exploratory laparotomy. Intraoperatively, a large solid mass was identified in the retroperitoneum. Although the tumour was adherent to the left colon, stomach and pancreatic tail, no invasion was noted. Frozen section of the tumour showed spindle-shaped myofibroblast cells, pointing to a probable diagnosis of IMT. Because of this pathologic report (IMT) and the likelihood of local recurrence, part of left colon, stomach and tail of pancreas were also excised together with the tumour. The child recovered well with no post-operative complication. At 12 months after surgery, he remained disease free.

Pathology

The pathological specimen showed a 24×17×10 cm tumour weighing 3.113 kg (Figure 2). Microscopically, tumour was composed of spindle-shaped myofibroblast cells, scanty giant cells, and many inflammatory cells embraced within collagen abundant and thin-wall blood-abundant vessel stroma. Few focal cells with mitoses were noted.

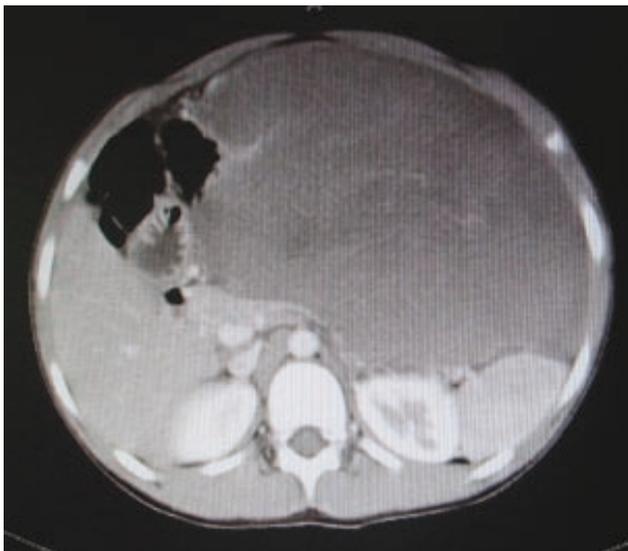


Figure 1 Abdominal computed tomography scan demonstrated a well-defined, heterogeneous mass lesion in the left side of the abdomen. Large solid tumour mass measuring 24×17×10 cm in size, we cut sagittalis, showed grayish white and firm internal appearance. (HE x 100)

Immunohistochemical staining revealed diffuse vimentin positivity with smooth muscle actin in the spindled population (Figure 3). The plump rounded cells showed positivity for desmin. The final histological diagnosis was an IMT of the retroperitoneal invading stomach, left colon and pancreatic tail with clear resection margins.

Discussion

Inflammatory myofibroblastic tumour was first described by Brunn in 1939.³ It has subsequently been reported to



Figure 2 The removed specimen measures 24 cm×17 cm×10 cm in size. It weights 3.113 kg.



Figure 3 Histological section of tumor showing spindle-shaped myofibroblastic cells, scanty giant cells, and many inflammatory cells embraced within collagen abundant and thin-wall blood-abundant vessels stroma. Immunohistochemical staining revealed diffuse smooth muscle actin positivity in the spindled population. (EnVision x 100)

occur in a wide variety of anatomic sites. IMT is now thought to be a true neoplasm, but one that shows a wide spectrum of biological behaviour, varying from common benign lesions to the uncommon varieties that are multifocal with high risk of multiple recurrences.⁴ The most frequent site of occurrence is in the lungs.⁵ For extra-pulmonary IMTs, small bowel mesentery is the most common site of origin.⁶ Retroperitoneal locations are exceedingly rare. Indeed, Medline search revealed only six cases. We summarised these published papers in the Table 1.^{2,7-11}

Currently, it is generally accepted that IMT is indeed a true neoplasm. However, the aetiology of IMT has not yet been clarified. The proposed aetiologies included Epstein Barr virus (EBV), Human herpes virus (HHV8), and over expression of interleukin IL-6. Moreover the recent research suggest that IMT is probably a neoplasm rather than a post-inflammatory process because of cytogenetic clonality, recurrent involvement of chromosomal region 2p23, occasional aggressive local behaviour and metastasis of the tumour.¹² From the recently report, we know the anaplastic lymphoma kinase (ALK) constitutive activation expressed in the IMT and squamous cell carcinomas.¹³ This finding has focused intense interest in inhibiting ALK signaling as an effective molecular therapy against disease with ALK-driven pathways. Butrynski¹⁴ also report that rearrangement correlates well with ALK expression and kinase domain activation, both of which probably predict ALK dependence and crizotinib sensitivity. The dependence of ALK-rearranged tumours on ALK-mediated signaling and suggest a therapeutic strategy for genomically identified patients with the aggressive form of this soft-tissue tumour. However, it is not possible to predict which tumours will

have ALK rearrangement or ALK aneuploidy.

IMTs are seldom diagnosed before operation. Making a correct clinical diagnosis of IMT can be difficult because patients present such diverse symptoms. There are several differential diagnoses of a large retroperitoneal mass including germ cell tumour, rhabdomyosarcoma, neuroblastoma (NB) or neuroganglioblastoma. computed tomography (CT) remains a useful, commonly used modality for the evaluation of retroperitoneal mass. It can demonstrate calcification in almost 85% of NB and IMT. MRI is extremely useful in detecting intraspinal tumour extension.¹⁵ Positron emission tomography/computed tomography (PET/CT) can help surgeon to precisely locate the tumour, IMT is well known for tendency of local recurrence following resection and tumour metastases. On the basis of imaging findings alone, IMT are difficult to differentiate from other retroperitoneal mass, percutaneous needle biopsy can improve our understanding of the disease decision. On the other hand, NB patients excrete vanillylmandelic acid (VMA) which may point to the diagnosis of NB.

Clinical presentation of intra-abdominal IMT depends on its location and growth pattern. In general, the most common initial symptoms and signs are palpable mass, abdominal distention, weight loss, anaemia, and fever. Intraabdominal tumours usually present with an abdominal mass with features of inflammation, such as fever, weight loss and anaemia. Haematologically, there was hypochromic microcytic anaemia in this case.

Typically, IMT is characterised by the expression of vimentin, smooth muscle actin, and cytokeratins, corresponding to that of myofibroblasts along with other

Table 1 The recently case reports of inflammatory myofibroblastic tumour retroperitoneal in the Medline

Reference	Sex/Age (year)	Location	Surgery	Size	Pathology	Follow-up	Chemotherapy
Koirala et al, 2010 ⁷	M/52	Pelvic	Removal	12.5 cm	IMT	1 year	No
Frigui et al, 2009 ⁸	F/14	Left kidney, retroperitoneal	Removal	8 cm	IMT	NA	No
Sugimoto et al, 2008 ⁹	M/61	Right kidney	Removal	3 cm	IMT	3 months	No
Attili et al, 2005 ²	F/46	Left kidney, retroperitoneal	Removal	16 cm	IMT	1 year	No
Tambo et al, 2003 ¹⁰	F/46	Right kidney	Mass with right kidney	NA	IMT	9 months	NA
Esmer-Sanchez et al, 2002 ¹¹	F/55	Attach pancreas	Removal	NA	IMT	NA	NA

NA: Not available; No: Not chemotherapy.

Removal: surgical resection, well-defined margin.

inflammatory markers (CD-68 is an inflammatory marker) (Figure 4).¹⁶ In the present case, Immunohistochemical staining revealed positive for smooth muscle actin and desmin, which are smooth muscle markers and CD-68 is an inflammatory marker, suggesting the tumour to be IMT. Based on the pathological patterns, extra-pulmonary IMTs are histologically categorised into three types:¹⁷ (1) resemble granulation tissue, nodular fasciitis; (2) resemble fibromatosis, fibrous histiocytoma, and smooth muscle neoplasm; (3) resemble a scar or desmoids fibromatosis. These are three different pathologic types of IMT. Based on the above features, we can diagnosis IMT and differentiate from other tumours.

Surgical resection remains the best method of cure. Adjuvant chemotherapy or radiotherapy has been proved to have no or little benefit.¹⁸ Risk of recurrence has been reported to be associated with larger tumours (>8 cm), tumours which originated from the omentum with ill-defined margin, or those presented with pathologically atypia, ganglionlike cells. Thus, close follow-up after primary surgery is necessary.¹⁹

In conclusion, IMT is neoplasm of proliferating myofibroblasts with an associated inflammatory component. Though IMT is uncommon, it should be kept as one of differential diagnosis of retroperitoneal masses. Final diagnosis is based on histological features and immunohistochemical staining. Complete surgical excision should be the aim of curative treatment. Long-term clinical and imaging follow-up are indicated because of the possibility of local recurrence even after many years.

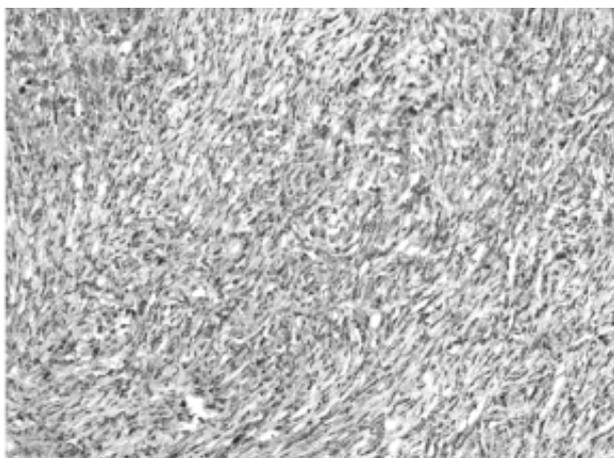


Figure 4 Immunohistochemical staining, the CD-68 (inflammatory marker) showed the focal positivity in the tumour. (EnVision x 100).

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