A Case of Rare Small Bowel Tumor in a Child and Review of Literature

J. Deepak^{1,2} · K. L. Aravind¹ · Gowrishankar¹ · S. Ramesh¹

Received: 22 November 2013 / Accepted: 19 May 2015 / Published online: 3 June 2015
© Indian Association of Surgical Oncology 2015

Introduction

Small bowel tumors are amongst the rarest types of childhood malignancies, and accounts for only 2 % of all gastrointestinal (GI) cancers across all age groups. An overwhelming 98 % of malignant neoplasms of small bowel are comprised of adenocarcinomas, carcinoid tumors, lymphomas or sarcoma/gastrointestinal stromal tumors (GISTs). The less common benign small bowel tumors include hyperplastic polyps, adenomas, GISTs, lipomas, hemangiomas and those associated with Peutz Jeghers syndrome. We encountered a child with rare variant of small intestinal tumor involving the jejunum. We present this case along with a review of the literature.

Case Report

A two and half years old female child presented with recurrent episodes of colicky abdominal pain and bilious vomiting since 3 months. On clinical examination, a small mobile intra-

abdominal mass in the peri-umbilical region was felt. The rest of the examination was normal. Ultrasonography (USG) abdomen revealed a well-defined hypo echoic mass, measuring 3.7×2.3 cm in the right iliac fossa with multiple enlarged mesenteric lymph nodes and mild ascites. Complete blood count showed hemoglobin (Hb) of 9.5gm/dl and erythrocyte sedimentation rate (ESR) of 24 mm/hr, with normal leucocyte and platelet counts. The renal and liver function tests were normal.

Contrast Enhanced Computed Tomography (CECT) abdomen showed 4×2.7 cm mass arising from small bowel with subtle central calcification (Fig. 1).

A working diagnosis of small bowel tumor with a high probability of GIST was made and the child was taken up for surgery. Fine needle aspiration biopsy (FNAC) or trucut needle biopsy was not attempted as the lesion was small, mobile, well circumscribed and easily resectable en bloc on imaging review.

At laparotomy, a 5×4 cm well encapsulated mass was seen arising from the ante-mesenteric surface of the proximal jejunum with a few mildly enlarged lymph nodes in the adjacent mesentery (Fig. 2). The tumor was resected completely with a 5 cm margin of normal intestine on either sides, along with the adjacent mesentery and enlarged lymph nodes, followed by a primary end to end bowel anastomosis. Postoperative recovery was uneventful.

Pathological examination revealed a well circumscribed, firm, globular mass measuring 4×4×2 cm arising from the serosal aspect of the antimesenteric border of intestine extending upto the underlying mucosa. Cut section of the tumor showed uniformly distributed greyish white tissue. Microscopic examination was suggestive of spindle cell neoplasm, showing plenty of spindle cells, with adjacent areas of collagenisation and calcification. There was no evidence myxoid area or fascicular pattern or necrosis or haemorrhage.

✉ J. Deepak
drjdeepak@yahoo.co.in

K. L. Aravind
draravindkl@gmail.com

Gowrishankar
bcgshankar@gmail.com

S. Ramesh
doctorsramesh@gmail.com

¹ Department of Pediatric Surgery, South Hospital Complex, Dharmaram College Post, Indira Gandhi Institute of Child Health, Near NIMHANS, Bangalore 560029, India

² No.1247, 8th cross, Chandra layout, Vijayanagar Bangalore 560040, India

Fig. 1 Contrast Enhanced Computed Tomography (CECT) abdomen showing the mass arising from small bowel with subtle central calcification

Spindle cell nucleus showed coarse chromatin with no evidence of mitosis or nuclear atypia or hemorrhage. Inflammatory cell infiltrate rich in plasma cells was seen throughout the specimen (Fig. 3). Resected intestine margins were free from tumor. All the six mesenteric nodes were negative for tumor cells.

Immunohistochemistry (IHC) study was done to predominantly differentiate between 2 principal differential diagnoses including intestinal GIST and intestinal IMT. It revealed spindle cells positive for smooth muscle actin (SMA) (Fig. 4) and S100 and IHC was negative CD34, CD117 and DOG1 pointing to the diagnosis of IMT. SMA positivity is found in >90 % of IMT and CD34, CD117 and DOG1 is usually negative in IMT. Whereas CD34, CD117 and DOG1 is usually positive in GIST. Thus the diagnosis of intestinal GIST was ruled out by IHC.

Fig. 2 Globular mass arising from the ante-mesenteric border of proximal jejunum

Fig. 3 Hematoxylin & Eosin staining [100×], showing myofibroblasts with infiltrates of plasma cells and lymphocytes ("cart wheel appearance")

The need for further chemotherapy and radiotherapy was not considered by medical oncologist in view of complete tumor resection with 5 cm normal intestinal margin, tumor having no mitotic activity or nuclear atypia, and tumor being confined to single site with size less than 5 cm and having no distant or lymph node metastases. However a 3 monthly follow up with clinical examination and abdominal sonography is done regularly. Child is doing well at 18 months follow up visit. Abdominal sonography and CBC were normal at follow up. There is no evidence of any recurrence of tumor on follow up.

Discussion

IMT is a rare solid tumor of spindle cell origin that often affects children and young adults [1, 2]. The variety of terms

Fig. 4 Immunohistochemistry (IHC) [100×], showing myofibroblasts staining positive for smooth muscle actin [SMA]

used to describe this entity includes inflammatory pseudotumor, cellular inflammatory pseudotumor, plasma cell granuloma, inflammatory myofibrohistiocytic tumor and inflammatory fibrosarcoma [3, 4]. The etiology and cellular origin of IMT remains unclear. Some authors believe that this entity represents an immunologic response to an infectious agent or a non-infectious agent, whereas others believe that IMT is a true neoplasm. Several associations are reported between IMT and previous abdominal surgery, lung infection, and *Campylobacter jejuni* infection. IMT can virtually affect every anatomic region and organ of the body. According to World Health Organisation classification IMT is a soft tissue tumor belonging to fibroblastic/myofibroblastic group of tumors [1]. Further it is classified in the subgroup of intermediate (rarely metastasizing) myofibroblastic tumor [1]. Most common site of this tumor is lungs [2]. Among the extrapulmonary sites, IMT is commonly reported in stomach, mesentery and greater omentum [4].

Abdominal IMT often presents as palpable mass with pain abdomen, fever and weight loss. Intestinal obstruction may be the presentation, when mass causes external encasement, luminal compromise due to large mass from the bowel wall and rarely because of bowel intussusceptions [4, 5]. The child reported in this study presented with abdominal mass and subacute intestinal obstruction secondary to luminal compromise from the tumor arising from jejunum. Initial imaging modality is USG abdomen. Contrast enhanced CT scan is helpful to define the mass and often shows areas of central necrosis and calcification. Differential diagnoses include gastrointestinal stromal tumor (GIST), lymphoma, adenocarcinoma, desmoid tumor and carcinoid tumor [5].

Multiple studies have shown expression of p80 and the clonal rearrangement of the anaplastic lymphoma kinase (ALK) gene on chromosome 2p23, leading to over expression of the oncoprotein in the spindle cell components of some of these tumors. Abnormalities in chromosome 2p is seen in up to 60 % of patients younger than 10 years of age [5]. DNA aneuploidy and association of the lesion with oncogenic viruses such as Epstein-Barr virus, Human Herpes virus type 8 and over expression of IL-6 have also been demonstrated.

Laboratory abnormalities seen in minority of patients are suggestive of chronic inflammatory response like elevated erythrocyte sedimentation rate (ESR), anaemia, thrombocytosis, hypergammaglobulinemia. These often resolve after resection of the tumor. Our patient had anaemia and elevated ESR preoperatively which resolved at 6 month follow up.

Histopathologically, IMT is characterised by spindle cell proliferation with areas of myxoid change, hypocellularity and collagenous background. Chronic inflammatory infiltrate is seen with polyclonal plasma cells, lymphocytes, eosinophils and rarely foamy macrophages [1].

On immunohistochemistry, IMT cells frequently positive for SMA (>90 %), desmin (10 to 70 %) and keratin [2]. IMT is usually negative for CD117, DOG1 and CD34 [2]. Complete en-bloc surgical resection is the treatment of choice [4]. During initial resection a thorough inspection is done to detect any smaller lesions, as these may be the cause for early recurrence.

Recurrences are documented in 18 to 40 % of cases [6, 7]. Pseudotumors confined to a single organ at the time of original presentation recur infrequently (6 %), whereas IMTs found outside a single organ at the time of original presentation recur frequently (35 %). Further, IMTs confined to the lung recur infrequently (1.5 %) as do extra pulmonary IMTs confined to a single organ (8 %). Therefore, although containment within a single organ is of great prognostic significance, complete removal with adequate margins is of equal importance [8].

Surgical excision is the treatment for recurrent lesions. Other modalities of treatment for recurrent lesions are adjuvant chemotherapy (cisplatin, doxorubicin and methotrexate), radiotherapy, steroid and non-steroidal anti-inflammatory drug therapy. NSAIDs are proposed to act by antiangiogenesis via down-regulation of VEGF, direct suppression of proliferating endothelial cell activation via COX-2, and an anticytokine effect with suppression of the inflammatory process [9]. However the role of NSAIDs is not well established. The role of targeted inhibitor of ALK such as Crizotinib in IMTs harbouring ALK translocations is also being evaluated [10].

Conclusion

In summary, IMT is a rare neoplastic variant of small bowel tumor with a tendency for local invasion and recurrence. This entity should be considered in the list of the differential diagnoses of small bowel tumors especially in the pediatric population.

Conflict of Interest All the four authors (Author A, B, C and D) declare that they have no conflict of interest.

References

1. Coffin CM, Fletcher JA (2013) Inflammatory myofibroblastic tumor. WHO classification of tumors of soft tissue and bone. IARC, Lyon
2. Coffin CM, Hornick JL, Fletcher CD (2007) Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. *Am J Surg Pathol* 31:509–520
3. Kamak I, Senocak ME, Ciftci AO, Çağlar M, Bingöl-Koçulu M, Tanyel FC et al (2001) Inflammatory myofibroblastic tumor in children: diagnosis and treatment. *J Pediatr Surg* 36:908–912

4. Chen SS, Liu SI, Mok KT, Wang BW, Yeh MH, Chen YC, Chen IS (2007) Mesenteric inflammatory myofibroblastic tumors in an elder patient with early recurrence: a case report. *World J Gastroenterol* 13(26):3645–3648
5. DiFiore JW, Goldblum JR (2002) Inflammatory myofibroblastic tumor of the small intestine. *J Am Coll Surg* 194(4):502–506
6. Coffin CM, Watterson J, Priest JR, Dehner LP (1995) Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor). A clinicopathologic and immunohistochemical study of 84 cases. *Am J Surg Pathol* 19:859–872
7. Gleason BC, Hornick JL (2008) Inflammatory myofibroblastic tumours: where are we now? *J Clin Pathol* 61:428–437
8. Janik JS, Janik JP, Lovell MA, Hendrickson RJ, Bensard DD, Denver BSG (2003) Recurrent inflammatory Pseudotumors in children. *J Pediatr Surg* 38:1491–1495
9. Applebaum H, Kieranb MW, Cripe TP, Coffin CM, Collinse MH, Kaipainenf A, Laforme A, Shamberger RC (2005) The rationale for nonsteroidal anti-inflammatory drug therapy for inflammatory myofibroblastic tumors: a Children's Oncology Group study. *J Pediatr Surg* 40:999–1003
10. Mosse YP, Lim MS, Voss SD, Wilner K, Ruffner K, Laliberte J et al (2013) Safety and activity of crizotinib for paediatric patients with refractory solid tumours or anaplastic large-cell lymphoma: a Children's Oncology Group phase 1 consortium study. *Lancet Oncol* 14:472–480