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CASE REPORT

AN EXTREMELY RARE PRESENTATION OF EXTRAPULMONARY INVOLVEMENT OF INFLAMMATORY MYOFIBROBLASTIC TUMOUR OF SMALL INTESTINE CAUSING INTUSSUSCEPTION

*Dr. Abhijit A. Whatkar, Dr. Bhushan H. Mahamulkar and Dr. Krishna Narayan Tiwari

Department of General Surgery, Noble Hospital, Pune, India

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*Corresponding author: Dr. Abhijit A. Whatkar

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ABSTRACT

Inflammatory myofibroblastic tumour (IMFT) is an uncommon mesenchymal solid tumour commonly documented in children and young adults. It is usually located in lungs however; extrapulmonary involvement has also been reported. IMFT of the small bowel is rare and to our knowledge, only 27 cases have been reported to date in the literature under several names worldwide. Here we report a 28th case of IMFT in a 62 year old man presenting as an ileoileal intussusception. Intussusception in adults presents with non-specific symptoms and classical image signs facilitate preoperative diagnosis. IMFT diagnosis is histopathological which usually implies surgical resection. IMFT must be kept in mind in the differential diagnosis of malignant spindle cell tumours of the small bowel. Immunohistochemical stains are essential for accurate diagnosis. Complete surgical excision, with microscopically clear margins is the mainstay of treatment.

INTRODUCTION

Spindle cell carcinoma or inflammatory myofibroblastic tumor (IMFT) is a rare but real tumor, which is histologically characterized by myofibroblastic spindle cell proliferation with inflammatory infiltrate. It may occur throughout the body in both children and adults, but the lung is the most common affected organ (Höhne *et al.*, 2015; Coffin *et al.*, 1995; Makhoul *et al.*, 2002). Sarcomatoid carcinoma is a rare variant of small intestinal carcinoma (Tsutomer Namikawa *et al.*, 2009). These tumours have been reported in diverse sites including gallbladder (Kim *et al.*, 2003), stomach (Khan *et al.*, 1999), oesophagus (Raza and Mazzara, 2011) and colon (Choi *et al.*, 2011), but are only rarely reported in the small intestine (Reid-Nicholson *et al.*, 2004; Yucel *et al.*, 2011; Moriwaki *et al.*, 2009). Outcome is poor in these cases because patients usually present with large tumour at extended stages (Höhne *et al.*, 2015). Its clinical symptoms and signs are also diverse according to the location of the tumour. However, the tumour in the gastrointestinal tract accompanied by intussusception appears to be rare (Makhoul *et al.*, 2002; Milne *et al.*, 2006; Zuccarello *et al.*, 2006; Ambiru *et al.*, 2009; Salameh *et al.*, 2011; Rezaii Salim *et al.*, 2011; Ntloko *et al.*, 2011; Ida *et al.*, 2013; Gupta *et al.*, 2013; Walia *et al.*, 2014; Appak *et al.*, 2014; Waszak *et al.*, 2015; Dulskas *et al.*, 2016).

CASE REPORT

A 62 year old male presented with pain in the abdomen on and off from the past 3-4 months and had increased in intensity over the past 3 days. The pain was colicky in nature, non-radiating and mostly limited to the right lower quadrant. He also had 3-4 episodes of bilious vomiting and fever since 2 days. The patient complained of recent onset constipation that almost dates back a couple of months and had not passed stools since 3 days. There was a past history of per rectal bleed on and off since 1 year. Patient had experienced around 5 kgs weight loss since the past one month. On examination he was averagely built and had pallor. Abdomen was soft, mildly distended; tenderness was present in the right lower quadrant with no guarding or rigidity. On Per rectal examination there was spasm, evidence of healed fissure at 6 o'clock position and rectum was loaded with faecal matter. X-ray abdomen in erect position showed multiple air fluid levels suggestive of bowel obstruction and an Ultrasound confirmed the findings of small bowel obstruction. A nasogastric tube was inserted for decompression. In view of progressive obstruction a contrast enhanced CT scan was done that revealed an Ileo-ileal intussusception with proximal jejunal obstruction with a mass over the intussuscepted segment. Routine haematological investigations revealed microcytic hypochromic anaemia;

blood cell counts, biochemical and serological investigations were within normal limits. The patient underwent an exploratory laparotomy and intussusception segment was identified almost 40 cm proximal to the ileocaecal junction (Fig.1). Resection of the ileal intussusception and an ileo-ileal anastomosis was performed. The resected specimen was sent for histopathological examination and consisted of a segment of ileum measuring 13 cm in length. The external surface showed dimpling of serosa 5 cm away from one of the cut margin. On cutting open, the mucosa showed a circumscribed globular mass measuring 3.5 x 3.5 x 3 cm in mid-third of ileum (Fig.2). Cut surface showed a firm, greyish white appearance. The surrounding ileal mucosa showed focal thickening, otherwise unremarkable. The closer surgical cut margin was 5.5 cm & the distant cut margin was 9.5 cm away from the mass. Findings were suggestive of a low grade spindle cell neoplasm of the ileum consistent with Inflammatory Myofibroblastic tumour. Mitosis was found to be rare and tumour cell necrosis was absent. Immunohistochemical stains showed immunoreactivity to SMA (Smooth Muscle Actin). No immunoreactivity was found for CD 117, ALK 1, Desmin and PCK. A Final diagnosis of Inflammatory Myofibroblastic Tumour was considered.

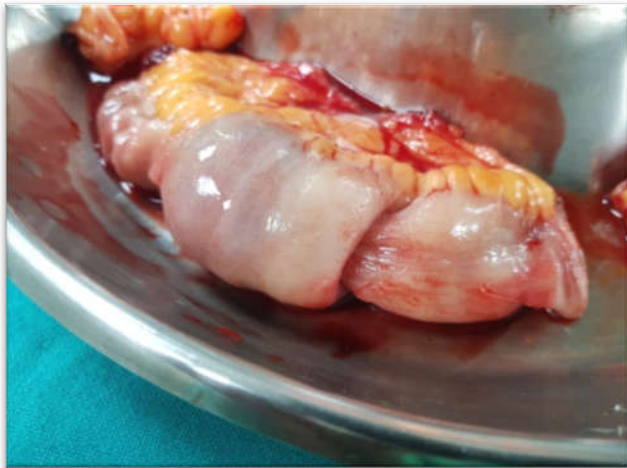


Fig. 1. Resected specimen showing Ileo-ileal intussusception

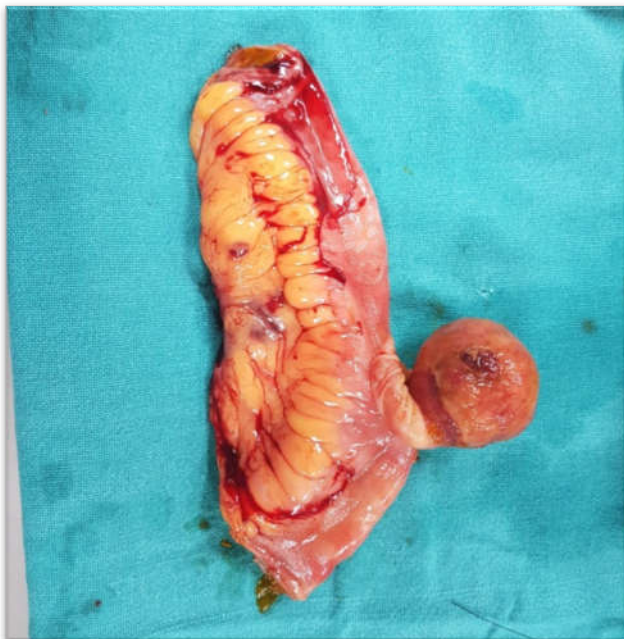


Fig. 2. A globular intraluminal mass as the intussusceptum

DISCUSSION

Intestinal intussusception is a common condition in children that is usually primary and benign. The 3 major causes of secondary intussusception are found to be SchÖnlein-Henoch purpura, Meckel's diverticulum, and polyps (Takeuchi *et al.*, 2012). However, intestinal intussusceptions caused by inflammatory myofibroblastic tumor (IMFT) is rare (Makhlouf *et al.*, 2002; Milne *et al.*, 2006; Zuccarello *et al.*, 2006; Ambiru *et al.*, 2009; Salameh *et al.*, 2011; Rezaii Salim *et al.*, 2011; Ntloko *et al.*, 2011; Ida *et al.*, 2013; Gupta *et al.*, 2013; Walia *et al.*, 2014; Appak *et al.*, 2014; Waszak *et al.*, 2015; Dulskas *et al.*, 2016). Intestinal IMFT can serve as a lead point of intussusception, cause bowel disorders and result in intestinal intussusception. IMFT or Sarcomatoid carcinoma is a controversial and rare tumor that displays both carcinomatous and sarcomatous features. It has variety of names including carcinosarcoma, metaplastic carcinoma, spindle cell carcinoma, and pleomorphic carcinoma (Reid-Nicholson *et al.*, 2004). For the small intestine, sarcomatoid carcinoma was first described by Dikman and Toker (Dikman *et al.*, 2009) in 1973. To the best of our knowledge only less than 30 cases of small intestinal sarcomatoid carcinoma have been reported in the English language literature till date (Reid-Nicholson *et al.*, 2004; Yucel *et al.*, 2011; Moriwaki and Sugiyama, 2009). IMFTs were first well described in the lungs and later became recognized in extrapulmonary locations (Gurzu *et al.*, 2013; Gleason *et al.*, 2008; Kovach *et al.*, 2006; Dalton *et al.*, 2015). 43% of extra-pulmonary involvement affects the mesentery and omentum (Coffin *et al.*, 1995). Gastrointestinal (GI) involvement is rare however there are reports of involvement of any part of GI tract (Coffin *et al.*, 1995; Gurzu *et al.*, 2013; Gong *et al.*, 2015; Unver *et al.*, 2015; Petrovic *et al.*, 2013). It occurs mostly in childhood and young adults but patients of any age and sex can be affected (Gurzu *et al.*, 2013). Clinically, sarcomatoid carcinomas in the small intestine primarily affect middle-aged to older patients, with a mean age of 57 years; with a male to female ratio of 1.5: 1 (Reid-Nicholson *et al.*, 2004).

The clinical presentation is determined by the site of origin and mass effect. A constitutional syndrome consisting of fever, weight loss and malaise is seen in 15–30% of patients (Gleason *et al.*, 2008). IMFT derived from the GI tract presents with clinical symptoms of anemia, loss of appetite or weight, faecal blood positive (Gurzu *et al.*, 2013), abdominal pain, GI obstruction (Petrovic *et al.*, 2013), or intussusception (Milne *et al.*, 2006). Adult patients with intussusceptions have non-specific and often long standing symptoms, usually present for more than a month (Azar *et al.*, 1997). An Intestinal IMFT with intussusception can involve the ileum and colon, the same location as the primary intussusception. The types of intussusceptions include ileoileal, ileocolic, and colocolic intussusception (Makhlouf *et al.*, 2002; Milne *et al.*, 2006; Zuccarello *et al.*, 2006; Ambiru *et al.*, 2009; Salameh *et al.*, 2011; Rezaii Salim *et al.*, 2011; Ntloko *et al.*, 2011; Ida *et al.*, 2013; Gupta *et al.*, 2013; Walia *et al.*, 2014; Appak *et al.*, 2014; Waszak *et al.*, 2015; Dulskas *et al.*, 2016). Our patient presented with a 3 month history of abdominal pain and unintentional weight loss of 5 kgs. He had features suggestive of small bowel obstruction and imaging techniques revealed an underlying ileoileal intussusception. Anaemia in this case may be attributed to a long standing history of per rectal bleed. Abdominal CT scan is currently considered as the most sensitive radiological method to confirm intussusception, with

a reported diagnostic accuracy of 58%–100% (Azar *et al.*, 1997; Honjo *et al.*, 2015). CT signs of intussusception include the “target sign” or shaped soft-tissue mass with a layering effect (Gorospe, 2008); mesenteric vessels within the bowel lumen are also typical. Histologically, IMFTs are characterized by a cellular spindle cell proliferation in a myxoid to collagenous stroma with a prominent inflammatory infiltrate composed primarily of plasma cells and lymphocytes, with occasional admixed eosinophils and neutrophils (Gleason *et al.*, 2008). Three basic histological patterns have been described: 1) Fasciitis-like, with vascular, myxoid, and inflamed stroma, including plasma cells – the inflammatory infiltrate in these areas often contains more neutrophils and eosinophils and fewer plasma cells than in the other two patterns; 2) Compact spindle cell pattern, that is characterized by a cellular proliferation of spindle cells with a fascicular architecture in a collagenous stroma, these foci typically show numerous plasma cells and lymphocytes intimately admixed with the spindle cells; 3) Fibromatosis-like pattern is relatively hypocellular, with elongated rather than plump spindle cells in a densely collagenous background containing scattered lymphocytes, plasma cells and eosinophils, fibrosis and calcification can be seen in the stroma (Coffin *et al.*, 1995).

Pleomorphism is moderate, but mitoses are infrequently seen. Immunohistochemistry analysis showed strong positive for SMA, and negative for ALK-1, Desmin, and CD117 (C-kit), corresponding with the myofibroblastic nature of these cells, and essentially ruled out other gastrointestinal stromal tumors. All of the above mentioned findings and immunohistochemistry were strongly favouring the diagnosis of IMT in our case. Complete surgical excision is the mainstay of treatment and provides the best chance to limit recurrence. When completely resected, with microscopically clear margins, there are virtually no recurrences (Dalton *et al.*, 2015). Chemotherapy and radiation have been used as adjuncts to surgery when the tumour is found not amenable to complete surgical resection (e.g. multiple nodules), when there are positive margins or the tumour is locally aggressive (Kovach *et al.*, 2006; Dalton *et al.*, 2015). Non-steroidal anti-inflammatory drug (NSAID) and steroid have been used in symptom control, usually in conjunction with other treatment modalities (Kovach *et al.*, 2006; Dalton *et al.*, 2015). In patients with ALK rearrangement, targeted kinase inhibition with Crizotinib has been used. This therapy along with surgery has proved useful in cases complicated by local recurrences (Butrynski *et al.*, 2010). IMFTs are classified as tumours of intermediate biological potential by the World Health Organization (WHO) classification, due to a tendency for local recurrence and a small risk of distant metastasis. The recurrence rate varies with anatomical site, from 2%, for tumours confined to the lung, to 25% for extra-pulmonary lesions, usually related with incomplete surgical resection or multi-nodular intra-abdominal tumours (Gleason *et al.*, 2008). The main prognostic indicator is the adequacy of the primary excision (Dalton *et al.*, 2015). In most cases, the prognosis is good, as recurrence is infrequent following complete excision of solitary lesions and metastases are present in less than 5% of cases, usually related with tumours with atypia, ganglion-like cells, TP53 expression, and aneuploidy (Fletcher *et al.*, 2002; Kovach *et al.*, 2006; Dalton *et al.*, 2015). Mortality rate is low, with only few cases described in literature determining IMFT as a cause of death, mostly due to the localization of the tumour, e.g. heart (Coffin *et al.*, 1995). Because IMFTs can recur locally, regular follow-up is necessary even when

surgical resection was performed, especially in the first year, when most recurrences occur (Dalton *et al.*, 2015). There are no guidelines for follow-up of patients with IMFTs, so an individualized approach must be planned. Our patient had a complete excision with clear margins and a low grade tumour with no features of tumour aggressiveness. Thus a symptom based follow-up can be regarded as an appropriate approach.

Conclusion

IMFTs are neoplasms that occur primarily in children and young adults that rarely affect GI tract. It should always be kept in our mind is that IMFT may present with bowel intussusception; hence, a detailed history, physical examination, and imaging studies are necessary for early recognition and diagnosis. It should be noted that the definitive diagnosis is made through a histopathological study. The pathophysiology of this disease is not correctly established yet and this neoplasm is still under study. As small intestinal sarcomatoid carcinoma demonstrates highly aggressive behavior, radical surgery and follow up are recommended.

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