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Inflammatory myofibroblastic tumor presenting as ileocecal intussusception—A case report

Cláudia Paiva*, Filomena Soares, Raquel da Inez Correia, Vítor Valente

Department of General Surgery, Hospital de Santo António, Centro Hospitalar do Porto, EPE, Largo Professor Abel Salazar, 4099-001 Porto, Portugal

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ABSTRACT

INTRODUCTION: Inflammatory myofibroblastic tumor (IMFT) is an uncommon mesenchymal solid tumor commonly documented in children and young adults. It is usually located in lungs however, extrapulmonary involvement has also been reported. Here we report a case of IMFT presenting as an ileocecal intussusception.

PRESENTATION OF CASE: A 55-year-old man presented with a two months history of colicky abdominal pain, more intense at the right inferior abdominal quadrant, and unintentional weight loss of 8 kg in the previous four months. Computer tomography showed an image of intussusception at the cecum. Colonoscopy demonstrated a pediculated tubular lesion, with the base near ileocecal valve. We performed a right hemicolectomy. Histopathological examination of the tumor revealed an IMFT.

DISCUSSION: IMFT usually affects lungs with rare gastrointestinal involvement. Clinical presentation is related with location of the neoplasm. Intussusception in adults presents with non-specific symptoms and classical image signs facilitate preoperative diagnosis. IMFT diagnosis is histopathological which usually implies surgical resection. Complete surgical excision, with microscopically clear margins, is the mainstain of treatment, with virtually no local recurrence or metastasis.

CONCLUSION: Gastrointestinal IMFT are rare in the adult population and clinical features depend on its location. Surgical approach with total excision of the neoplasm confirms the diagnosis. Prognosis is good with the main prognostic indicator being the adequacy of the primary excision.

1. Introduction

Inflammatory myofibroblastic tumor (IMFT) is a rare soft tissue neoplasm of unclear etiology composed of myofibroblastic spindle cells accompanied by a dense inflammatory cell component [1]. It occurs primarily in soft tissue and viscera of children and young adults, although the age range extends throughout adulthood [2]. Lung is the site most commonly affected but virtually any site may be involved, including spleen, stomach, intestine, liver, mediastinum, retroperitoneum, bladder, and they can occur as single or multiple tumors [2–5]. IMFT has been synonymously referred to as plasma cell granuloma, plasma cell pseudotumour, inflammatory myofibrohistiocytic proliferation, omental mesenteric myxoid hamartoma, inflammatory pseudotumour [1]. All of these share a key pathologic differentiation: a dominant spindle cell proliferation with a variable inflammatory component. These spindle cells are myofibroblasts, thus, the preferred term for reference should be IMFT reflective of this pathologic feature. Here we describe the case of a cecal IMFT presenting as an ileocecal intussusception.

2. Presentation of case

Our patient was a 55-year-old man with a past history of hypertension and spine surgery. He went to our Emergency Department with a two months history of colicky abdominal pain, more intense at the right inferior abdominal quadrant. He had also unintentional weight loss of 8 kg in the previous four months. Physical examination revealed tenderness in the right inferior abdominal quadrant. Laboratory tests were normal. He was investigated with an abdominal ultrasound which revealed an image of ileocecal intussusception (Fig. 1). The computed tomography (CT) scan revealed an image of intussusception at the cecum, the intussusceptum being a 12 mm hypodense, tubular image that seemed to be the appendix (Fig. 2). He underwent colonoscopy which demonstrated a pediculated tubular lesion, with the base near ileocecal valve, occupying most of ascending colic lumen (Fig. 3). Laparotomy confirmed an ileocecal intussusception with the lead point at a cecal neoplasm that reached the base of the enlarged appendix;

Abbreviations: IMFT, inflammatory myofibroblastic tumor; CT, computed tomography scan; ALK, anaplastic lymphoma kinase; GI, gastrointestinal; NSAID, nonsteroidal anti-inflammatory drugs.

* Correspondence to: Rua Vale Formoso número 62-3°, 4200-507, Porto, Portugal.
E-mail addresses: linhaspaiva@hotmail.com (C. Paiva), filomenasoaresmoreira@gmail.com (F. Soares), dra.inezcorreia@gmail.com (R. da Inez Correia), correiavalente@hotmail.com (V. Valente).

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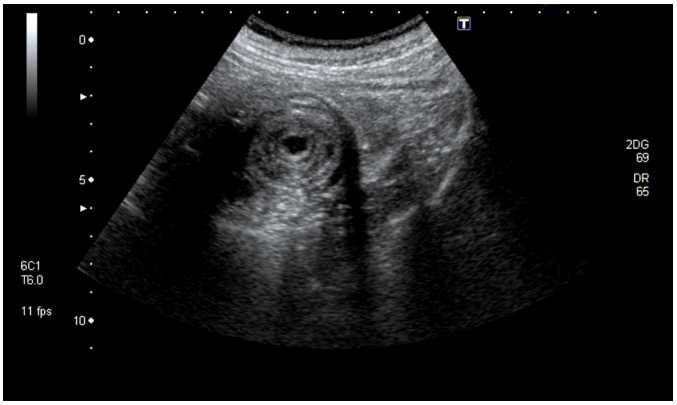

Fig 1. Ultrasonographic image in transverse section: hypoechoic outer layer of edematous bowel wall with echogenic layers, known as the "bulls-eye" or "target" signs, which are characteristic images in intussusception.

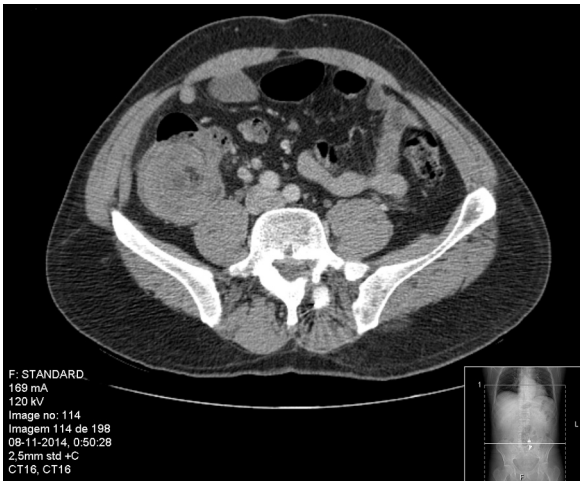

Fig. 2. Axial CT of the abdomen: shows the edematous wall of the intussuscipiens, the "target" sign, suggesting ileocecal intussusception.

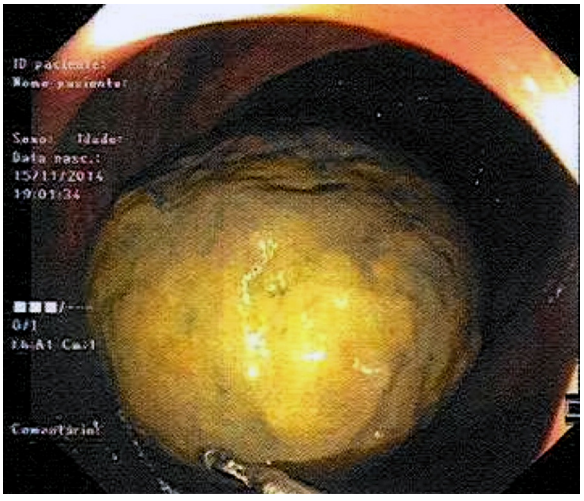

Fig. 3. Endoscopic examination revealed a pediculated lesion, with the base near ileocecal valve, occupying most of ascending colic lumen.

a right hemicolectomy was performed (Fig. 4). Postoperatively, the patient did well, without complication. He was discharged home uneventful at the 7th postoperative day (Table 1).

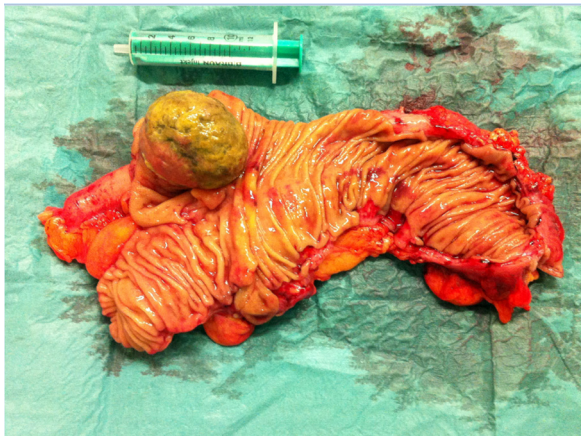

Fig. 4. Bowel wall is cut away, revealing the mass emanating near the base of the appendix.

Table 1
Briefing of the case.

Identification	Male, 55 years old
Medical history	Hypertension; spine surgery
Chief complaint	Abdominal pain, strongest in right inferior quadrant
Physical exam	Tenderness in the right inferior abdominal quadrant
Diagnostic assessment	Laboratory tests: normal. Abdominal ultrasound: image of ileocecal intussusception. Computed tomography scan: image of intussusception at the cecum, the intussusceptum being a 12 mm hypodense, tubular image Colonoscopy: a pediculate tubular lesion, with the base near ileo-cecal valve occupying most of ascending colic lumen
Treatment	Surgery: right hemicolectomy.
Histopathological exam	Complete excision of a low grade inflammatory myofibroblastic tumor
Outcome	No complication. Discharged home at the 7thpostoperative day.
Follow-up	Without clinical recurrence at 7 months

On pathological examination, gross inspection of the specimen revealed a solid, well-defined mass, measuring $5 \times 5 \times 4.5$ cm, near the base of the appendix with exophytic growth into intestinal lumen. It obstructed the appendix, causing a mucocele. Microscopically, the tumor had a proliferation of spindle cells, such as myofibroblasts and fibroblasts, and between these there was a mixed inflammatory infiltrate with eosinophils, and edematous vascularized stroma. There was no nuclear atypia or mitotic activity. Immunohistochemically, it was positive for smooth muscle actin and negative for desmin, CD34, S100, CD31, AE1AE3 and CD117. It also stained negative for ALK. This was compatible with an inflammatory myofibroblastic tumor, low grade tumor/grade 1 in the French Federation of Cancer Centers Sarcoma Group (FNCLCC) classification. The tumor was removed completely. He did well in the last 7 months.

3. Discussion

Different terms for IMFT have been used over the years – plasma cell granuloma, plasma cell pseudotumour, inflammatory myofibrohistiocytic proliferation, omental mesenteric myxoid hamartoma, inflammatory pseudotumour [1]. The term inflammatory pseudotumor has been used for any clinically, macroscopic or microscopic tumor-like lesion caused by an inflammatory or

reactive process. Later, cytogenetic and molecular evidence supported a clonal origin, implying that it is a neoplastic process. A chromosomal rearrangement at band 2p23 is found in about half of cases, which is the site of the anaplastic lymphoma kinase (ALK) gene in the tyrosine kinase locus [3]. ALK mutations are less found in extrapulmonary IMFT. The etiology of IMFT is still unknown. Some authors think that development of IMFT occurs after trauma, surgery or infection, such as Epstein-Barr virus and human herpes virus, related with reactive cytokine production [2–6].

IMFTs were first well described in the lungs and later became recognized in extrapulmonary locations [2–5] – 43% of extrapulmonary involvement affects the mesentery and omentum [7]. Gastrointestinal (GI) involvement is rare however there are reports of involvement of any part of GI tract [2,6–9]. It occurs mostly in childhood and young adults but patients of any age and sex can be affected [2].

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 [3]. IMFT derived from the GI tract presents with clinical symptoms of anemia, loss of appetite or weight, fecal blood positive [2], abdominal pain, GI obstruction [9], or intussusception [10]. Our patient presented with 2 months history of abdominal pain and unintentional weight loss of 8 kg, and imaging techniques revealed it to be an ileocecal intussusception. Adult patients with intussusceptions have non-specific and often long standing symptoms, usually present for more than a month [11]. The classical image signs facilitated the diagnosis, both ultrasonography and abdominal CT scan proved to be useful tools for the diagnosis of intussusception. The classic appearance of an intussuscepted bowel on a sonographic image in a transverse plane is called the "doughnut sign" or a "target lesion" and represents several concentric rings of the bowel. The longitudinal appearance of intussusception usually appears as multiple parallel lines, the so-called "sandwich appearance" or "pseudo-kidney sign"; the lines demonstrate bowel walls and their layers [12]. Abdominal CT is currently considered as the most sensitive radiologic method to confirm intussusception, with a reported diagnostic accuracy of 58%–100% [11,13]. CT signs of intussusception include the "target sign" or shaped soft-tissue mass with a layering effect [14]; mesenteric vessels within the bowel lumen are also typical. CT can be helpful in determining the underlying cause, locating the lead point and evaluating complications. However, both endoscopic and computed tomography examinations do not reveal any differences between an IMFT and other types of neoplasm so it is impossible to differentiate IMFT from a malignant process based on these studies. Diagnosis is based on histopathologic analysis, which usually implies surgical resection. 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 [3]. 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 [7]. Pleomorphism is moderate, but mitoses are infrequently seen.

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 [5]. Chemotherapy and radiation treatment have been used as adjuncts to surgery when the tumor is found not amenable to complete surgical resection (eg, multiple nodules), when there are positive margins or the tumor is locally aggressive [4,5]. Non-steroidal anti- inflammatory drug (NSAID) and steroid have been used in symptom control, usually in conjunction with other treatment modalities [4,5]. 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 [15].

IMFTs are classified as tumors of intermediate biological potential by the World Health Organization 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 tumors confined to the lung, to 25% for extrapulmonary lesions, usually related with incomplete surgical resection or multinodular intra-abdominal tumors [3]. The main prognostic indicator is the adequacy of the primary excision [5]. 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 tumors with atypia, ganglion-like cells, TP53 expression, and aneuploidy [1,4,5]. 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 tumor, e.g. heart [7].

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 [5]. There are no guidelines for follow-up of patients with IMFTs, so an individualized approach must be planned. Our patient had a complete excision and no features of tumor aggressiveness, as such, a symptom based follow-up can be regarded as an appropriate approach.

4. Conclusion

IMFTs are neoplasms that occur primarily in children and young adults that rarely affect GI tract. Diagnosis is histological and the treatment is complete surgical excision. Prognosis is good as they rarely recur locally or metastasize.

Conflicts of interest

None.

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This case-report had no sponsors.

Ethical approval

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Cláudia Paiva – research on bibliographic material; writing of the paper; final approval of the version to be submitted.

Filomena Soares – research on bibliographic material and article preparation; final approval of the version to be submitted.

Raquel da Inez Correia – research on bibliographic material and article preparation; revised critically this case report; final approval of the version to be submitted.

Vítor Valente – revised critically this case report; final approval of the version to be submitted.

Guarantor

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