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Case report

A case report of inflammatory myofibroblastic tumor of cecum mimicking malignant wall thickening

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ABSTRACT

Introduction: Inflammatory myofibroblastic tumors (IMT) are an uncommon mesenchymal solid tumor commonly documented in children and young adults (Kim et al., 2012 [1]). Cecum is a rare location of this entity, may simulating a malignant tumor process.

Presentation of case: A 71 year old patient was admitted for pain in the right iliac fossa with chronic constipation evolving for two months associated with weight loss.

As a diagnosic step, an abdominal computed tomography (CT) scan was performed showing a thickening of the cecal wall. There was no regional or distant metastasis. During a colonoscopy, many biopsies have been returned to a non-specific chronic colitis; as a result, the IMT of the cecum was confirmed pathologically after ilio-coecal resection.

Discussion: IMT is difined as a solid mesenchymal tumor basically affects the soft parts as well as the visceral organs. The litterature show that frequent localisations are pulmonary and intra-orbital (Krzysztof Siemion et al., 23 February 2022); therefore, the cecal location is very rare may mimic a malignant tumor (Mauricio Gonzalez-Urquijo et al., January 20, 2020),

It is worth recalling that the imaging outcomes are polymorphic and inconclusive; in addition, Surgical excision is the treatment of choice for IMT (Alireza Mirshemirani et al., Dec 2011) and the histopathology is still required for the final diagnosis.

Conclusion: Radiologic features of IMT present a diagnostic challenge to the radiologist; also, mimic malignancy may lead to unnecessary investigations.

1. Introduction [1]

Inflammatory myofibroblastic tumor (IMT) is an unusual solid tumor involving soft tissue and visceral organs. It occurs mainly in the lung and is rarely present at other extrapulmonary sites.

Clinical manifestations are not specific, depending on the location of the tumor, an inflammatory syndrome may be associated involving fever, weight loss, anemia, hyperplacetosis, polyclonal hypergammaglobulinemia, and high sediment rate.

IMT imaging results are polymorphic and inconclusive. However, diagnosis is based on histopathology. Basically, these tumors are derived from mesenchymal origin and were grouped into a mixture of fibroinflammatory disorders, including the variable spread of inflammatory cells with spindle cells. Surgical treatment remains the preferred treatment, complete excision with a negative margin is less likely to recur.

Our work is focused on the inflammatory myofibroblastic tumor in cecum as a rare site of this entity, which may mimic a malignant tumor, may lead to unnecessary investigations and it follows perfectly the SCARE criteria [2].

2. Case report

A 71-year-old male patient, with a past history of midline hernia surgery in 2017, without occupation, admitted in our structure for right iliac fossa apyretic pain associated to chronic constipation evolving for two months.

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Physical examination objectified a conscious patient, apyretic, with soft abdomen and maximum sensitivity in the right iliac fossa. The Laboratory panel was notable for microcytic anemia (hemoglobin 9.6 g/del, reference 12–17), C-reactive protein 25 mg/del (reference <0.2). Otherwise, other biological tests were normal. An abdominal ultrasound was first performed, but it came back negative due to the digestive gas affecting the scan.

An abdominal CT scan was carried out, showing a thickness of cecal tumor invading the last ileal loop, length of 5 cm with a maximum thickness of 7 mm. The appendix was undamaged. No regional and distant metastasis (Fig. 1). Considering the age of the patient, a malignant origin was evoked, based on that a colonoscopy was performed showed a parietal thickening of cecum, noted at 20 mm from the Bauhin's valve. A multiple biopsies have been made in favour of nonspecific chronic colitis.

The patient was scheduled for exploration and on laparotomy, noticing a wall thickness of cecum for a length of 7 cm; however, there was no evidence of metastatic deposits. Based on histopathological biopsy findings and CT scan data, an Ileococcal resection with lateral anastomosis was done with adequate margins. The post-operative state was normal without immediate or late complications, clinical manifestation resolved.

Histopathology of the specimen revealed a cecal wall infiltrated by a tumor arranged in loose fascicles composed of fibroblasts and myofibroblasts on a fibrous matrix. The tumor cells were mixed with plasma cells, lymphocytes, eosinophils and vascular fan-shaped proliferation. In immunohistochemistry, the spindle cells were negative for CD34 and anaplasic lymphoma kinase1 (ALK1). The profile corresponded to that of an inflammatory myofibroblastic tumor (Fig. 2).

A control CT scan was performed after 2 months, showing fat infiltration in the anastomotic region, with no evidence of residue or tumor recurrence in the short-term follow-up (Fig. 3).

3. Discussion

Inflammatory myofibroblastic tumors (IMT) in the digestive tract are very rare. According to the reported cases, it seems that the stomach and small bowel are the most common sites [3,4]. Regarding the colon, the most frequent site is the right colon and cecum [5]. Children and young adults are more commonly affected. There is no prevalence of gender in the frequency of cases [1].

The mechanism of this tumor is still uncertain; history of abdominal surgery, auto-immune response, infection, particularly the Epstein-Barr virus, and the human herpes virus, have been found in these places [6]. In this case, even if the patient has a previous surgical treatment of the median hernia.

The clinical signs are non-specific, revealing itself either incidentally or after abdominal pain, during an investigation of an occult anemia, or following an occlusive syndrome or bowel obstruction, they also may present fever and malaise [4,5].

Elevated erythrocyte sedimentation rate (ESR), leukocytosis, thrombocytosis, and hypergammaglobulinemia are the laboratory findings that may be noted in some cases due to inflammatory mediators such interleukin 6, interleukin 1b, and cyclin B1 [7]. Our patient had an inflammatory anemia with moderately high C-reactive protein.

The radiological appearance of IMTs is not accurate and they are often misdiagnosed as malignant neoplasms, so a biopsy or surgical procedure is required.

On ultrasound these tumors can be hypoechoic, roughly circumscribed with nonsystemic vascularization. During a CT scan, the most common presentation is that of solid, irregular, and well-defined masses, hypodense, with variable contrast uptake because of the fibrous contingent. It is early and heterogeneous for young fibrosis, cellular inflammatory and edematous, late and homogeneous for mature collagenous fibrosis. The same semiological aspects are found in

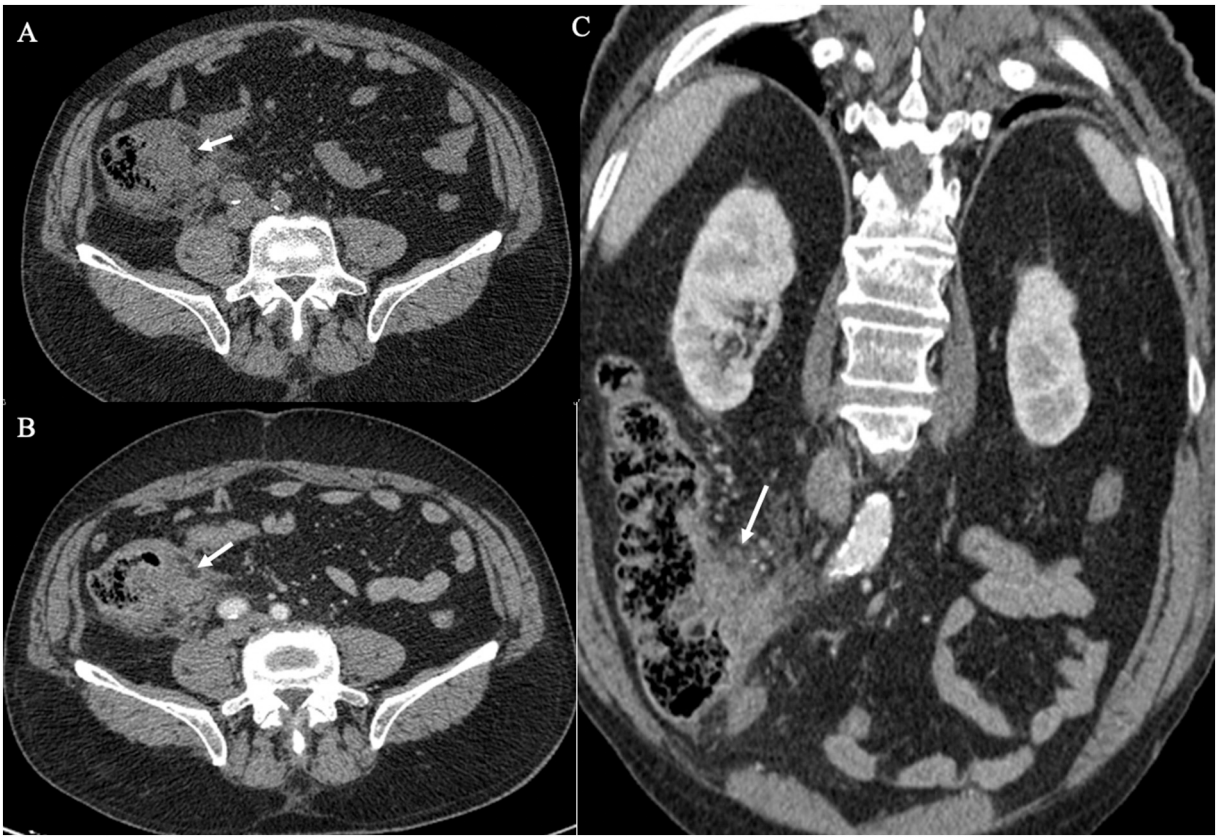

Fig. 1. Abdominal CT scan images in axial (A, B) and coronal (C) views before (A) and after (B, C) injection of contrast agent showing a cecal wall thickness invading the last ileal loop, length of 5 cm with a maximum thickness of 7 mm (arrow). These are an infiltration of the surrounding fat without regional and distant metastasis.

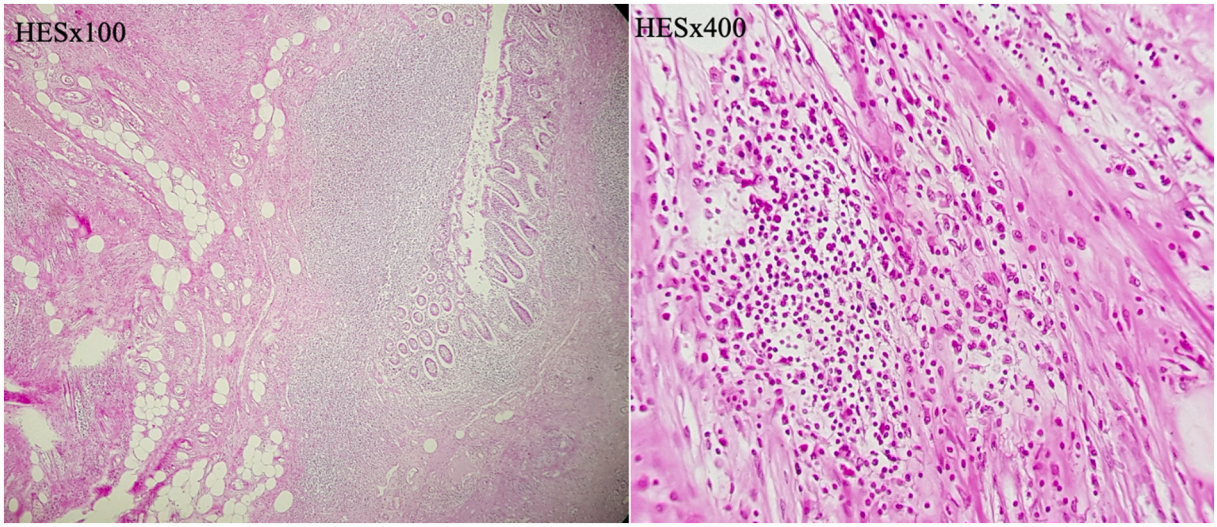

Fig. 2. HES 100: Colonic mucosal dissociated by a proliferation made of activated fibroblasts and myofibroblasts that are arranged on a dense fibrous background. This proliferation is combined with a rich lymphocytic and plasmocytic inflammatory infiltrate.
HES400: Microscopy: fusiform cells mixed with an extensive chronic inflammatory infiltrate of plasmatic cells, lymphocytes and macrophages.

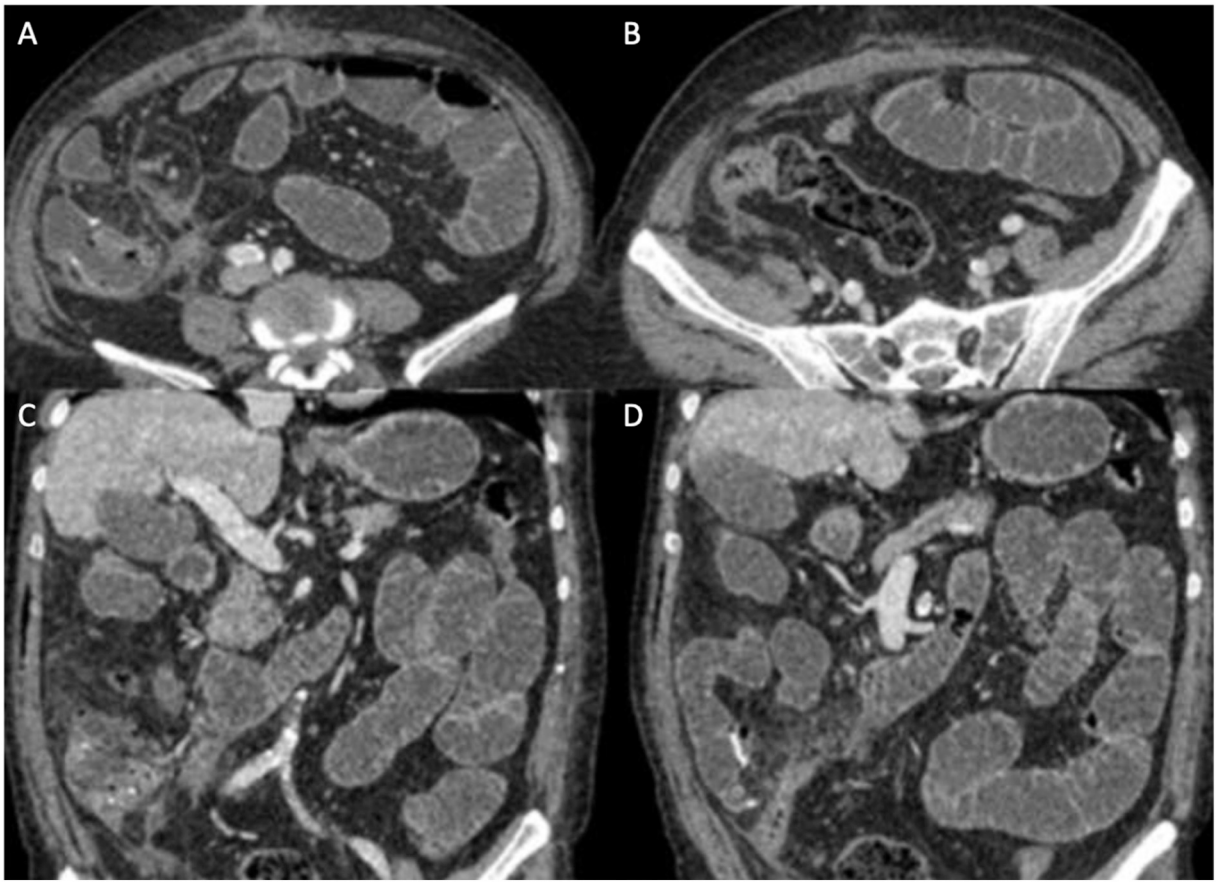

Fig. 3. Control Abdominal CT scan images in axial (A, B) and coronal (C, D) after injection of contrast agent showing post-operative fat infiltration in the anastomosis area, no sign of residue or tumor recurrence in the short-term follow-up.

magnetic resonance imaging (MRI) [8].

The differential diagnosis of IMTs includes malignancy and submucosal tumor from endoscopic findings [1].

Gastrointestinal stromal tumors (GISTs), schwannomas, solitary cell fibroid tumors, desmoid tumors, follicular dendritic cell sarcomas, leiomyomas, and leiomyosarcomas have pathological findings similar to IMTs [1].

The inflammatory fibroid polyp consists of a mixture of small granulation tissue-like vessels, spindle cells, and inflammatory cells (in particular eosinophils). Fibromatoses are made up of spindle or stellate cells, which are arranged in parallel with regularly spaced blood vessels and a collagenous background.

They tend to exhibit mitotic activity. GISTs are marked by CD34 and CD117 (c-kit). Leiomyome and leiomyosarcome are desmin and actin positive and CD117 and CD34 negative. Schwannoma is colored with S-100 (nuclear and cytoplasmic) and is negative for CD117 [1,9].

In immunohistochemical terms, diffuse cytoplasmic reactivity for vimentin is typical of virtually all IMT. Reactivity of smooth muscle actin and muscle-specific actin ranges from a focal to diffuse pattern in the cytoplasm of spindle cells, and desmin is found in many cases [9].

Positivity for ALK1 is demonstrated in about 50% of cases, positivity for ALK1 is still not specific for IMTs [10].

According to an update based on the new World Health Organisation (WHO) classification in 2002, IMTs are considered as an intermediate malignancy with a risk of metastatic spread of less than 5% [10].

Malignant transformation has been interpreted as being due to a chromosome re-arrangement affecting the 2p23 site of ALK gene [5,11].

Multiple treatment modalities have been described for IMT, But Surgical treatment is the therapy of choice; however, more cases of recurrence were reported in those patients with incomplete resection, complete surgical excision with an adequate negative margin has less than 10% chance of recurrence [12].

The adjustment of chemotherapy combined with oral non-steroidal anti-inflammatory drugs, either with or against radiotherapy may be reserved for inoperable tumors, post incomplete resection, at present, there is no evidence of favorable response [8].

4. Conclusion

Inflammatory myofibroblasts tumors are an under-diagnosed entity, and their radiological characteristics suggest malignancy, when in fact they are not. It can be said that only after histological examination can a definitive diagnosis be reached, and surgery remains the preferred and effective treatment for this type of tumor. Last but not least, complete excision with a negative margin is less chance of recurrence of the disease.

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Ethical approval

Our study is exempt from ethical approval.

Registration of research studies

NA.

Declaration of competing interest

The authors report no declarations of interest.

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