Case report

Inflammatory myofibroblastic tumor of the proximal ileon in a patient with complicated umbilical hernia: A case report and literature review

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ARTICLE INFO

Keywords:
Inflammatory myofibroblastic tumor
Low-grade sarcoma
Intestinal obstruction

ABSTRACT

Introduction: Inflammatory myofibroblastic tumors are neoplasms that occur infrequently, mainly affects children and young adults. It is an intermediate grade fibrotic multinodular neoplasm.

Description of the case: We present the case of a 47-year-old female patient, who underwent emergency umbilical hernioplasty, later developed intestinal obstruction secondary to an inflammatory myofibroblastic tumor.

Discussion: In 1939 Brunn described it for the first time, later in 1954 Umiker named it “Inflammatory Myofibroblastic Tumor”. The symptoms are nonspecific. In 15 to 40% of patients they are asymptomatic. Cells positive for actin, smooth muscle, vimentin and desmin, in 3367% of cases the cells are positive for ALK, which is present in some malignant lesions. The recommended treatment is radical resection.

Conclusion: The diagnosis is established by histopathological study, surgery is the cornerstone of treatment.

1. Introduction

Inflammatory myofibroblastic tumors are neoplasms that occur infrequently, most frequently affect children and young adults, however it can occur at any age [1–4].

It is an intermediate grade fibrotic multinodular neoplasm, which originates from the soft tissue or the viscera, the most common sites of involvement are the lung, the mesentry and the omentum, it can present as a multifocal disease within the abdomen, it can increase in size in 5 to 10 years [5].

This type of tumor presents highly variable clinical characteristics, evolution, location and prognosis; the definitive diagnosis is made through histopathological study, currently surgery constitutes the definitive treatment with low levels of recurrence when resection is complete [6].

2. Case description

We present the case of a 47-year-old female patient with abdominal pain in the epigastrium of 19 h of evolution, which is why she decided to go to the emergency department. Important antecedents: genetic burden for diabetes mellitus, arterial hypertension of 7 years of evolution, drugs enalapril 10 mg twice daily, previous surgeries; umbilical hernioplasty 4 years ago. No known genetic history.

Relevant data from the physical examination; Tachycardia, distended abdomen, decreased peristalsis, non-reducible postincisional umbilical hernia, painful on palpation, discoloration changes. Presurgical exams within normal parameters. She is taken to emergency hernioplasty for our surgical team, hernioplasty is performed with mesh placement without complications, hospital stay without complications, she is discharged after 24 h.

On the fifth postoperative day, she returned to the emergency department due to intestinal obstruction, an abdominal tomography was performed with a transition site near the ileocecal valve, small bowel loops greater than 3 cm, interase edema and absence of air in the rectal ampulla.

Our surgical team took the patient to an exploratory laparotomy, finding distended small bowel loops with a transition site in the proximal ileum at 80 cm from the ileocecal valve, an intraluminal tumor was palpated (Fig. 1) which causes an 80% obstruction of the intestinal lumen (Fig. 2), we resected with a proximal and distal margin of 5 cm with manual anastomosis. We explored the surgical specimen, finding...
an intraluminal tumor of approximately 5 cm. (Fig. 3).

We started clear liquids at 24 h and advanced the diet at 72 h without complications, the patient was discharged on the fourth day. Histo-pathological report: A segment of the small intestine measuring 10 by 2.5 cm when sectioned, a pedunculated nodule measuring 4.5 by 2.5 cm is identified, which is located 3 and 7 cm from the lateral surgical limits with a smooth gray-brown external surface and a soft consistency. When cut, the surface of the nodule has a grayish gelatinous appearance, with focal necrotic-hemorrhagic areas and a soft consistency, macroscopically in continuity with the serosa. The rest of the intestinal mucosa has light brown edematous folds with an elastic consistency. The wall with an average thickness of 0.4 cm. Conclusion 4 cm diameter inflammatory myofibroblastic tumor of the small intestine.

During the follow-up the patient is satisfied with the treatment, at the moment no recurrence data [7].

3. Discussion

In 1939 Brunn described it for the first time, later in 1954 Umiker named it “Inflammatory Myofibroblastic Tumor”. Since then it has received multiple names, for example, inflammatory pseudotumor, plasma cell granuloma, histiocytoma, xanthoma, fibroxanthoma [1–3]. At first it was believed that these tumors were secondary to an inflammatory response caused by tissue damage, however today it is classified as a neoplasm since it presents a more aggressive evolution than an inflammatory process also has the ability to invade blood vessels and recur [4–6].

Inflammatory myofibroblastic tumors are infrequent tumors with a behavior most of the time benign, can occur between 2 and 16 years old, in a recent review two incidence peaks were observed, one in pediatric age, the second between 50 and 60 years [8–10]. In our case, the diagnosis was made at 47 years of age, which corresponds to the second peak of incidence.

Any part of the body that is affected, the main reported sites are the lung, the mesenterium and the omentum, less frequently the ileum and colon. Other sites reported even less frequently are the orbit, meninges, heart, thyroid glands and kidney [11–13]. Table 1 shows reports from the literature, with different sites of involvement.

The etiological factors have not yet been clearly established, some authors have associated it with an immune response generated by pathogens such as; Mycobacterium avium intracellullare, Campylobacter jejuni, Corinebacterium equi, Bacillus sphaerius, Coxiella burnetti, Ebstein-Barr, Escherichia coli or in those with a history of abdominal surgeries, radiotherapy, taking corticosteroids [22, 23]. In our patient, the only important antecedent was umbilical hernioplasty 4 years earlier.

It has been reported that patients present aberrations in chromosome 2p23 and 9p, which suggests that genetic factors are relevant in the formation of this type of tumor [24]. In our study, the etiology was not fully determined; the finding was incidental and unrelated to the initial clinical presentation.

The symptoms are nonspecific, when it is abdominal there is pain and systemic manifestations such as fever, anorexia, weight loss, but there may be others such as dysphagia, occlusion, constipation and rectal bleeding. In 15 to 40% of patients they are asymptomatic. When it is located in the small intestine, it generally presents as a surgical emergency [25–27]. In our case, the patient presented obstructive symptoms after the previously performed umbilical hernioplasty, which generated confusion since we consider that this condition was due to a complication of the procedure.

Laboratory studies are not very specific. Imaging studies are taken for another pathology and can be found as incidental tumors, which suggest a benign process, using ultrasound and tomography, encapsulated hypodense lesions can be observed [25–27]. In our case, the office studies did not reveal the presence of a tumor, the diagnosis was made once the histopathological study was completed.

The WHO defines IMTs as lesions composed of myofibroblastic spindle cell populations accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils. Macroscopically, they are observed as a well circumscribed, encapsulated, firm tumor with non-infiltrating edges and focal myxoid changes, with a size of 2 to 15 cm. In the histological analysis, 3 patterns are observed: firstly, myxoid, vascular changes and inflammatory areas, secondly spindle cells, compact and intermixed inflammatory cells, and thirdly a predominance of dense collagen matrix [28,29].
Table 1
Reports from the literature, with different sites of involvement.

| Case report | Year | Genero | Age (y) | Size (cm) | Anatomic site      | ALK | Follow | Recurrence |
|-------------|------|--------|---------|-----------|--------------------|-----|--------|------------|
| Yung-Song Yeh [14] | 2010 | Female | 50y     | 4         | Distal ileon       | NR  | NR     | NR         |
| Abdolhamid Amouei [15] | 2016 | Male   | 5y      | 15        | Ileocecal          | NR  | NR     | NR         |
| Our case    | 2016 | Female | 47y     | 4.5       | Ileon              | NR  | 4 years| SR         |
| Eugenia Raffaeli [16] | 2019 | Female | 59y     | 2.8       | Transverse Colon   | Neg | 6 months| SR         |
| Naem Lisap [17] | 2019 | Female | 14 m    | 10        | Jejunum            | Pos | 6 months| SR         |
| Rina Harada [18] | 2020 | Male   | 18 m    | 8         | Ileon              | Pos | 1 year | SR         |
| Yosra Braham [19] | 2020 | Female | 46y     | 2         | Lung               | NR  | 14 month| SR         |
| Rose George [20] | 2021 | Female | 27y     | 6.6       | Uracho             | Pos | NR     | NR         |
| A.S. Ivanov [21] | 2021 | Male   | 59y     | 3.8       | Kidney             | Neg | NR     | NR         |

Abbreviations: cm; centimeter, y; year, m; months, NR; not reported, Neg; negative, SR; no recurrence, Pos; positive

4. Conclusions

Myofibroblastic tumors occur infrequently, in most cases they are incidental findings both in complementary studies or found inoperatively, so it is important to make a correct diagnosis and thus provide adequate treatment, reducing the rate of recurrence.

Provenance and peer review
No commissioned, internally reviewed.

Funding
None.

Ethical approval
None required.

Sources of funding
None.

Author contribution
Adrian Morales Cardenas. Concept and design, data collection, drafting, revision and approval of final manuscript.

Montserrat del Carmen Valencia Romero. Concept and design, data collection, drafting, revision and approval of final manuscript.

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Declaration of competing interest
Nothing to declare.

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