International Journal of Surgery Case Reports 84 (2021) 106091

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

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

Montserrat Del Carmen Valencia Romero [a], Adrián Morales Cárdenas [b,*], José Enrique Cabrales Vázquez [c], César Felipe Ploneda Valencia [a]

[a] Hospital Civil Dr. Juan I. Menchaca, Mexico
[b] Surgical Oncology, Regional Hospital IMSS 46, Mexico
[c] Institute Jalisciense of Cancerology, Guadalajara, Mexico

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 mesentery 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

* Corresponding author.
E-mail address: adrianmd05@gmail.com (A.M. Cárdenas).

https://doi.org/10.1016/j.ijscr.2021.106091
Received 17 April 2021; Received in revised form 21 May 2021; Accepted 4 June 2021
Available online 8 June 2021

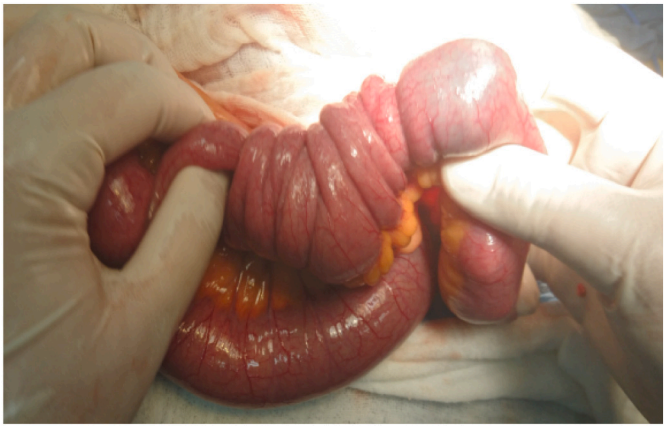

Fig. 1. Tumor 80 cm from the ileocecal valve.

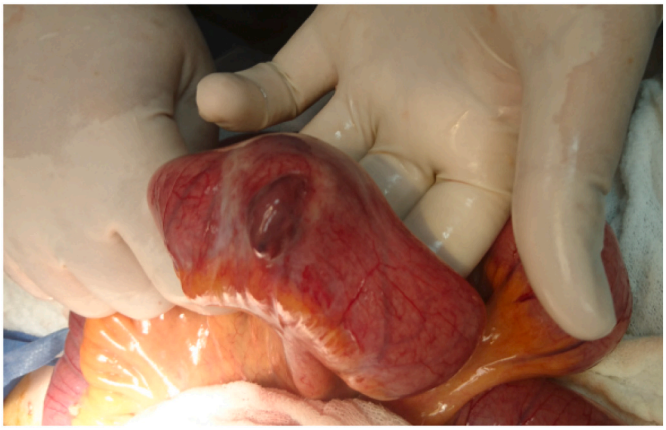

Fig. 2. Tumor obstructing 80% of lumen in proximal ileum.

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. Histopathological 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.

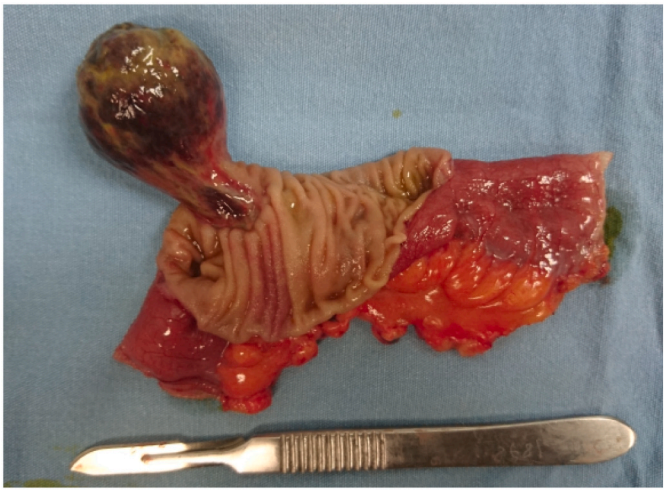

Fig. 3. Intraluminal tumor, pedunculated 5 cm.

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 myxioid changes, with a size of 2 to 15 cm. In the histological analysis, 3 patterns are observed: firstly, myxioid, 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	Size (cm)	Anatomic site	ALK	Follow	Recurrence
Yung-Sung 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
Naeem Liaqat [17]	2019	Female	14 m	10	Jejunum	Pos	6 months	SR
Rina Harada [18]	2020	Male	18 m	3	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

Immunohistochemical analysis reveals inflammatory cells that express benignity, as opposed to malignant cells that are polyclonal. Cells positive for actin, smooth muscle, vimentin and desmin, in 33–67% of cases the cells are positive for ALK, which is present in some malignant lesions [17,28]. Ultrastructurally, a myofibroblastic component with abundant rough endoplasmic reticulum, cytoplasmic filaments and dense bodies is demonstrated [29].

IMT generally has a benign behavior, the local recurrence rate is approximately 2 to 25%, the risk of distant metastasis is 5%. When there are metastases, they occur mainly in the lung, abdomen, mesentery, retroperitoneum, mediastinum, liver, pelvic bones, neck and forearm [29,30].

The possibility of malignancy is mainly due when the myofibroblasts present atypia, even some authors consider them as low-grade or intermediate-grade fibrosarcomas and even a pseudosarcoma rather than a pseudotumor [29,30].

The treatment of choice is complete resection, since this is curative in most cases, once the tumor is resected, the possibility of recurrence is very low. When an incomplete resection is performed, there may be a recurrence years later, most commonly within the first year [31]. Other treatments have been described such as corticosteroids, anti-inflammatories, radiotherapy, colchicine, which are reserved for cases in which radical resection cannot be performed [28–31]. Our team opted for a wide intestinal resection with end-to-end anastomosis. After 5 years of follow-up, the patient did not present data of recurrence.

4. Conclusions

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

Provenance and peer review

Not 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.

José Enrique Cabrales Vazquez. Data collection, drafting, revision and approval of final manuscript.

Cesar Felipe Plonea Valencia. Data collection, drafting, revision and approval of final manuscript.

Consent

Written informed consent was obtained from the patient for the 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.

Guarantor

Adrian Morales Cardenas.

Declaration of competing interest

Nothing to declare.

References

[1] E. Contreras, I. Gallardo, S. y Espejo, P. Seguí, A propósito de un caso: tumor miofibroblástico inflamatorio de localización inusual, Radiología 52 (5) (2010) 473–476.
[2] H. Makhlouf, L. Sobin, Inflammatory myofibroblastic tumors (inflammatory pseudotumors) of the gastrointestinal tract: how closely are they related to inflammatory fibroid polyps? HumPathol 33 (2002) 307–315.
[3] S. López, C. Claderón, D. Carrasco, V. Ocampo, Tumor miofibroblástico inflamatorio gástrico en un niño de ocho años de edad, Acta Pediatr. Mex. 32 (2011) 332–336.
[4] C. Coffin, L. Dehner, Meis-Kindblom. Inflammatory myofibroblastic tumor, inflammatory fibrosarcoma and related lesions: an histological review with differential diagnostic considerations, Semin. Diagn. Pathol. 15 (1998) 102–110.
[5] American Joint Committee on Cancer, AJCC Cancer Staging Manual, Chapter 45, Eighth edition, Springer, 2018, pp. 539–545.
[6] M. Oscoz, J. Olascoaga, E. Rúperez, T. y cols Molins, Tumor miofibroblástico en la edad pediátrica. A propósito de 3 casos, An. Pediatr. (Barc) 71 (4) (2009) 331–335.
[7] Riaz A. Agha, Thomas Franchi, Catrin Sohrabi, Ginimol Mathew, Ahmed Kerwan, for the SCARE Group, The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[8] J. Attard, A. MacLean, Adhesive small bowel obstruction: epidemiology, biology and prevention, Can. J. Surg. 50 (2007) 291–300.
[9] T. Van Oudheusden, B. Aerts, I. Hjt de Hingh, M. Luyer, Challenges in diagnosing adhesive small bowel obstruction, World J. Gastroenterol. 19 (2013) 7489–7493.
[10] L. Narla, S. Spottswood, S. Narla, R. Kolli, Inflammatory pseudotumor, Radiographics 23 (2003) 719–729.
[11] I. Pinilla, Y. Herrero, M. Torre, M. Nistal, M. Pardo, Myofibroblastic inflamatory tumor of the lung, Radiologia 49 (2007) 53–55.
[12] Osnaya M, Zaragoza T, Escoto A y cols. Tumor miofibroblástico inflamatorio (Pseudotumor inflamatorio) ocasionando abdomen agudo. Rev. Chil. Cir. Vol 66 - N° 3, Junio 2014; pág. 264–268.
[13] T. Lazure, S. Ferlicot, F. Gauthier, et al., Gastric inflammatory myofibroblastic tumor in children: an unpredictable course, J. Pediatr. Gastroenterol. Nutr. 34 (2002) 319–322.
[14] Y.-S. Yeh, C.-J. Ma, S.-F. Yang, et al., Inflammatory Myofibroblastic tumor of the ileum causing an unusual Ileocecal intussusception, Foolyn J. Health Scl 2 (1) (2010) 36–39.

[15] A. Amouei, F. Ehsani, M. Vaghefi, et al., Inflammatory myofibroblastic tumor of the small intestine: a case report, Int. J. Surg. Case Rep. 22 (2016) 44–45.
[16] E. Raffaeli, L. Cardinali, M. Fianchini, et al., Inflammatory myofibroblastic tumor of the transverse colon with synchronous gastrointestinal stromal tumor in a patient with ulcerative colitis: a case report, Int. J. Surg. Case Rep. 60 (2019) 141–144.
[17] N. Liaqat, S. Tasneem, R.-M. Imram, et al., Inflammatory myofibroblastic tumor of jejunumin a child, J Pediatr Surg Case Rep 51 (2019), 101313.
[18] R. Harada, M. Ohtaki, N. Hashizume, et al., Inflammatory myofibroblastic tumor of the small intestine with intussusception in a child: case report and literature review, J Pediatr Surg Case Rep 101 (56) (2020) 423.
[19] Y. Braham, A. Migaou, M. Njima, Inflammatory myofibroblastic tumor of the lung: a rare entity, Resp. Med. Case Rep. 31 (2020), 101287.
[20] R. George, D. Swerdloff, M. Akgul, et al., A rare case of urachal inflammatory myofibroblastic tumor, Urol. Case Rep. 36 (2021), 101575.
[21] A.S. Ivanov, P.A. Antonov, Z.R. Chitalov, Renal inflammatory myofibroblastic tumor: a case report, Urol. Case Rep. 37 (2021), 101620.
[22] B. Gleason, J.L. Hornick, Inflammatory myofibroblastic tumours:where are we now? J. Clin. Pathol. 61 (2008) 428–437.
[23] M.R. Wick, O. Nappi, Inflammatory sarcomatoid carcinoma of the lung: report of three cases and clinicopathologic comparison with inflammatory pseudotumor in adult patients, Hum. Pathol. 26 (1995) 1014–1021.
[24] S. Treissman, D.A. Gillis, C.L. Lee, M. Giacomantonio, L. Resch, Omental-mesenteric inflammatory pseudotumor. Cytogenetic demostration of genetic changes and monoclonalityin one tumor, Cancer 73 (1994) 1433–1437.
[25] S.J. Kim, W.S. Kim, J.-E. Cheon, et al., Inflammatory myofibroblastic tumors of the abdomen as mimickers of malignancy:imaging features in nine children, AJR AMJ Roentgenol. 193 (2009) 1419–1424.
[26] N.E. New, P.W. Bishop, M. Stewart, M. Harris, Inflammatory pseudotumor of lymph nodes, J. Clin. Pathol. 48 (1995) 37–40.
[27] I. Karnak, M.E. Senocak, A.O. Ciftci, et al., Inflammatory myofibroblastic tumor in children: diagnosis and treatment, J. Pediatr. Surg. 36 (2001) 908–912.
[28] C. Fisher, Myofibroblastic malignancies, Adv. Anat. Pathol. 11 (2004) 190–201.
[29] T. Tang, A.D. Segura, H.W. Oechler, et al., Inflammatory myofibrohistiocytic proliferation simulating sarcoma in children, Cancer 65 (1990) 1626–1634.
[30] G. Hedlund, J.F. Navoy, C.A. Galliani, W.H. Johnson Jr., Aggressive manifestations of inflammatory pulmonary pseudotumor in children, Pediatr. Radiol. 29 (1999) 112–116.
[31] A. Csendes, P. Pereira, M. Zamorano, y cols. Tratamiento médico o quirúrgico de la obstrucción intestinal alta, Rev. Chil. Cir. 68 (3) (2016) 227–232.