Inflammatory myofibroblastic tumor (IMT), also called inflammatory pseudotumor, is a rare disease of mesenchymal origin, first described in 1937. IMT is characterized by a dense inflammatory cell component and amid myofibroblastic proliferation. Its histopathologic nature is benign, but it may not be differentiated from a malignant tumor because of its local invasiveness, aggressive behavior, and its tendency to recur and metastasize. In 2003 was classified as an intermediate neoplasm in the current World Health Organization histologic typing. It was initially described in the lung, which is also the site where it is most commonly presented. Subsequently, it was seen that it can also be presented in soft tissue, the mesentery, the omentum, and retroperitoneum; thenceforth, it was recognized that any anatomic location can be involved, making IMTs of the gastrointestinal tract extremely rare, being only 13 confirmed cases of appendicular origin. Most of these reported cases of IMT of the appendix are adolescents or young adult men. Herein, we present a case of a 65-year-old man who presented to our emergency department on septic shock and acute abdomen secondary to visceral perforation. The patient referred inability to pass flatus or to evacuate for the past 24 hours. On physical examination he was tachycardic and hypotensive. He presented with diffuse abdominal tenderness, abdominal pain, nausea, vomiting, and abdominal distension. Digital rectal examination revealed an empty rectum, without abnormalities.

Complete blood count confirmed the presence of leukocytosis of 17.9 × 10^3 with hemoglobin of 18.1 g/dL and platelet count of 207 × 10^3. Blood chemistry revealed acute renal failure, with a serum creatinine of 3 mg/dL, blood urea nitrogen 60 mg/dL, and urea of 210 mg/dL.

Simple abdominal radiographs displayed dilated loops and no signs of gas in the rectum, suggesting bowel obstruction, and no free air was seen. A noncontrast computed tomography (CT) of the abdomen was ordered, revealing a thick and irregular fibrous capsule at the base of the appendix with central low-attenuation necrotic component. Surrounding inflammatory changes were present with periappendiceal reactive nodal enlargement, pneumoperitoneum, and dilated large bowel. The appendix was enlarged with a perforation at the tip (Figure 1).

A preoperative diagnosis of bowel perforation with peritonitis was obtained, on the basis of clinical and radiological findings. The patient’s blood pressure could not be stabilized despite reanimation with crystalloids; therefore, the patient was transferred to the operating room where the attending surgeon and the surgical resident performed an emergency laparotomy. Massive bowel dilatation was encountered and umbilical hernia repaired presented to the emergency department. He arrived complaining of a 5-day history of diffuse abdominal pain, nausea, vomiting, and abdominal distension. The patient referred inability to pass flatus or to evacuate for the past 24 hours. On physical examination he was tachycardic and hypotensive. He presented with diffuse abdominal tenderness, suggestive of peritoneal irritation and severe distension. Digital rectal examination revealed an empty rectum, without abnormalities.

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approximately 3 L of purulent intraperitoneal fluid was aspirated. A mass measuring 6 cm × 6 cm was seen at the tip of the appendix with a perforated area of about 1 cm in diameter. The base of the cecum was healthy, and an appendectomy was successfully performed (Figure 2). The patient was transferred to the surgical intensive care unit for 48 hours and had an adequate postoperative outcome, referring only moderate pain during the initial days. Oral intake was reintroduced on the third day, without complications. On the postoperative day 7, the patient was discharged home, with good oral intake and with normal evacuations. The patient was seen in the ambulatory clinic at 1 and 6 months after surgery for a follow-up examination with a satisfactory evolution with normal renal function.

Histological examination of the specimen dyed with hematoxylin and eosin, revealed a mass showing fibroblastic proliferation accompanied by a dense inflammatory infiltrate in the mucosa and a clear thickening of the submucosa. A few areas showed myxoid changes with spindle cells, alternating with polyclonal plasma cells and lymphocytes (Figure 3). A perforation was identified in the section from the tip of the appendix lined by fibrinoid exudates. The immunohistochemistry of the paraffin section was positive for vimentin and smooth muscle actin and negative for anaplastic lymphoma kinase (ALK) (Figure 4). Final diagnosis was inflammatory pseudotumor with appendiceal perforation.

Discussion
Inflammatory myofibroblastic tumor is an atypical pseudosarcomatous, inflammatory, and histopathologic entity that most often affects children and young adults. When we performed a comprehensive search of English and Spanish literature, no more than 13 cases reported were found. Of note, most of the reported cases are in pediatric patients and young adult men. The first case report was published in 1984 and describes an 8-year-old boy who presented with symptoms of acute appendicitis, and the most recent report, published in 2018 by Bashir et al., described a 14-year-old boy with the same preoperative diagnosis. Our patient constitutes the oldest patient who has presented with this appendicular tumor, making it the 14th case of this entity and also the first to present with septic shock secondary to perforation. It is most similar to a case reported by Kumar in which the patient was also a male adult who presented with peritonitis.
A variety of substitutes have been attributed to these lesions, such as plasma cell granuloma, mast cell granuloma, histiocytoma, xanthomatous pseudotumor, fibroxanthoma, fibromyxomatous
tumor, pseudosarcomatous myofibroblastic proliferation, inflammatory fibrosarcoma, pseudosarcomatous fibromyxoma
tous tumor, and reactive pseudosarcomatous response.3,7 The
etiology of IMT remains unclear, and in most cases no causa
tive agent is evident. Its development has been described after
trauma, neoplasia, and surgery.9 There are reports of an associa
tion with Castleman disease, Hodgkin disease, peptic ulceration,
Behçet disease, and chronic infections (Campylobacter
jejuni, Helicobacter pylori, Escherichia coli, Epstein–Barr virus,
Coxiella burnetii, Klebsiella pneumoniae).3,5,8 We do not know
the exact origin of our patient’s appendiceal pseudotumor;
however, we do not consider acute appendicitis to be an etio-
logical factor for developing this entity. Nonetheless, we believe
that appendicitis occurred secondary to obliteration of
the appendiceal lumen by the tumor, leading to perforation.
Another rare presentation of this tumor was described by
Majumdar et al8 describing a case with concomitant mucosal
dysplasia, first impression of which was pseudomyxoma
peritonei.

Surgery remains the most effective treatment for this pseu-
dotumor1,3; however, it must be noted that IMTs present with
inconstant biological behavior that ranges from benign lesions
(most frequent) to more aggressive variations. Whether com-
plete surgical resection can be achieved depends on several fac-
tors such as the location of the tumor, multinodularity, and
proximity to neighbor structures.3,4 For multiple and invasive
lesions, chemotherapy with cisplatin, doxorubicin, and metho-
trexate may be considered, as well as radiotherapy for local
recurrence or incomplete removal of the tumor.1,3

The clinical presentation of IMT varies markedly, depend-
ing on the site at which the tumors originated. Most of the
patients with appendiceal IMT presented with abdominal pain
in the right lower quadrant, nausea, and vomiting, mimicking
acute appendicitis.7,8,10,12 In some patients, imaging studies
were ordered suspecting this last diagnosis, thus revealing an
appendicular mass.1,5,9,13,14 Despite detection during preopera-
tive screening methods, its radiological appearance is nonspe-
cific and insufficient to make a specific diagnosis. Therefore,
the final diagnosis is made by histopathologic examination.7

In our case, our patient had abdominal tenderness to palpa-
tion and presented with septic shock. We therefore ordered a
CT of the abdomen preoperatively, which reported changes
consistent with complicated appendicitis and bowel obstruc-
tion, leading to an emergent laparotomy.

The microscopic descriptions of IMT are relatively uni-
iform. In general, these tumors are composed of a dominant-
spindle cell proliferation with inflammatory polyclonal
mononuclear infiltrates, including plasma cells, lymphocytes,
and histiocytes, arranged in a collagenous and slightly myxoid
background. Our patient’s tumor presented these histologic
changes. Three histological patterns are described: fibromyxoid
or vascular, proliferating, and sclerosing. Immunostains on
spindle cells have often been found positive for vimentin,
smooth muscle actin, muscle-specific actin, desmin, and focally
for cytokeratins in most of the cases. Specific immunostaining
techniques can help differentiate these tumors, with a favorable
prognosis.5,7,10 The expression of ALK, may be a specific
marker for IMT. Immunostaining for ALK produced fibrillar

Figure 4. (A) Vimentin stain shows a strong positive stain to cytoplasm with mesenchymal differentiation. (B) Smooth muscle actin shows positive stain to
to myofibroblast. (C) Anaplastic lymphoma kinase stain results negative to cytoplasmatic and nuclear staining.
| REFERENCES      | COUNTRY     | AGE/GENDER | PRESENTATION       | IMAGING                                    | PROCEDURE                        | MICRO                                                                 | IHC                                      |
|-----------------|-------------|------------|--------------------|--------------------------------------------|----------------------------------|------------------------------------------------------------------------|------------------------------------------|
| 1 Narasimharao  | Japan       | 8/M        | Intermittent fever | US: 7 cm × 5 cm mass in right hypochondrium | Laparotomy, simple appendectomy  | Destruction of the appendicular mucosa and replacement of the wall by diffuse inflammatory cells, predominantly plasma cells | NA                                       |
| 2 Yamagiwa et al | Japan       | 41/M       | Abdominal pain     | US: tumorous mass in the appendix          | Laparotomy, simple appendectomy  | Eosinophilic cell and fibroblastic infiltrations                       | NA                                       |
| 3 Jougon, 1991  | France      | NA         | NA                 | NA                                         | NA                               | NA                                                                     | NA                                       |
| 4 Bonnet et al  | France      | 15/M       | Fever, anorexia,   | US: 4 cm retrovesical soft-tissue, noncalcified mass | Laparotomy, simple appendectomy  | Cellular proliferation with plasma cells, lymphocytes, histiocytes, and mesenchymal cells in a poor collagenous stroma | Positive: vimentin Negative: desmin, leukocyte common antigen, and S-100 |
| 5 Khoddami et al | Iran        | 29/M       | RLQ pain           | CT and US: 10.5 cm × 2.5 cm paracecal mass in the right lower quadrant | Laparotomy, right hemicolecotomy and ileo-transverse colon anastomosis   | Extensive spindle cell proliferation with mixed inflammatory cells and lymphoid follicle formation | Positive: Vimentin, desmin, and muscle-specific actin |
| 6 Vijayaraghavan | India       | 34/M       | RLQ pain, fever    | US: appendicular mass                      | Laparoscopic appendectomy        | Spindle-shaped myofibroblasts and inflammatory cells consisting of eosinophils, lymphocytes, plasma cells, and neutrophils | Positive: vimentin, smooth muscle actin Negative: neuron-specific enolase, S-100, desmin, CD34, and CD117 |
| 7 Uludag et al  | Belgium     | 20/M       | RLQ pain, nausea, | NA                                         | Conventional appendectomy        | Fibroblasts with pleomorphic swollen nuclei and a mixed type inflammation formed by histiocytes and dispersed neutrophil leukocytes, plasma cells, and small lymphocytes | Positive: vimentin, smooth muscle actin, and CD68 |

(Continued)
### Table 1. (Continued)

| REFERENCES       | COUNTRY       | AGE/GENDER | PRESENTATION                        | IMAGING                        | PROCEDURE                             | MICRO                                                                 | IHC                                      |
|------------------|---------------|------------|-------------------------------------|-------------------------------|----------------------------------------|------------------------------------------------------------------------|------------------------------------------|
| 8 Majumdar et al⁸ | India         | 41/M       | RLQ pain, palpable growing mass,    | US: 7 cm × 5 cm mass          | Laparotomy, Right hemicolecotomy and   | Dysplasia of mucosal lining, stroma rich in collagen, intense myofibroblastic proliferation, and a polymorph infiltrate comprising plasma cells, lymphocytes, histiocytes | Positive: smooth muscle actin and desmin | Negative: S-100                          |
|                  |               |            | fever, anorexia, weight loss        | involving the appendix       | ileo-transverse colon anastomosis       |                                                                        |                                          |                                         |
| 9 Eunji et al, 2014¹⁷ | Korea        | 85/M       | History of gastric cancer           | Incidental appendicular mass  | Conventional appendectomy               | Proliferation of spindle cells in a collagenous and slightly myxoid background with scattered blood vessels | Positive: vimentin and smooth muscle actin | Negative: S-100, ALK, c-kit, desmin, CD21, CD23, and CD35 |
| 10 Schoonjans et al, 2016¹⁸ | Belgium      | 42/F       | RLQ pain, nausea, fever             | CT: appendicular mass of 2.3 cm × 1.8 cm × 1.9 cm | Laparoscopic appendectomy               | Spindle cells, accompanied by a prominent inflammatory infiltrate, composed of plasma cells and lymphocytes | Positive: Vimentin and cytokeratin AE1/AE3 | Negative: CD68, desmin, CD117, and ALK |
| 11 Kumar³        | India         | 50/M       | RLQ pain, vomiting, anorexia        | Chest X-Ray with free gas under the right dome of the diaphragm | Laparotomy, simple appendectomy        | Fibroblastic proliferation accompanied by a prominent infiltrate of chronic inflammatory cells | Positive: vimentin, smooth muscle actin, and CD68 | Negative: ALK                           |
| 12 Henrique et al¹⁰ | Brazil        | 33/F       | RLQ pain, vomiting                  | CT: enlarged appendix with appendix mucocele | Conventional appendectomy               | Fusocellular pattern permeated by inflammatory cells rich in plasma cells and lymphoid aggregates | Positive: Vimentin, CD138, CD45, and myeloperoxidase | Negative: ALK-1                         |
| 13 Bashir et al¹⁰ | Saudi Arabia  | 14/M       | RLQ pain, nausea, vomiting          | US: 3.1 cm × 2.6 cm mucocele of the appendix | Laparotomy, simple appendectomy        | Spindle-shaped myofibroblastic cells in an edematous myxoid background with proliferating blood vessels and an infiltrate of plasma cells, lymphocytes, and eosinophils | Positive: desmin | Negative: CD117, CD34, DOG-1, and ALK-2 |

Abbreviations: ALK, anaplastic lymphoma kinase; CT, computed tomography; IHC, immunohistochemistry; NA, not available; RLQ, right lower quadrant; US, ultrasonography.
or granular cytoplasmic staining in the neoplastic cells, sometimes with cell membrane accentuation. This marker is implicated selectively in younger patients and is generally associated with a favorable outcome.15

Because of the presence of spindle cells in bundles, soft-tissue sarcomas such as malignant fibrous histiocytoma, rhabdomyosarcoma, and fibrosarcoma should be kept in mind in the differential diagnosis of IMT; nonetheless, these tumors will express mitotic activity.3 Final diagnosis of our case was diagnosed as benign IMT because there was eosinophilic infiltration, with no mitotic activity, and because of the occurrence of dense inflammatory cells and fibroblasts with reactive swollen nuclei.

The incidence of local recurrence has been reported to be 15% to 37% in a large series of children presenting with IMT of mesentery and retroperitoneum; furthermore, infrequent distant metastases also have been suggested to occur.4 Predictors of a potentially malignant behavior include cellular atypia, ganglion-like cells, necrosis, nucleolar prominence, atypical or increased mitotic figures, expression of p53, and DNA aneuploidy. However, a unique and consistent chromosomal abnormality has not been identified.7,8 Nonetheless, none of the 13 appendicular IMT reported cases had an aggressive behavior. They are summarized and detailed in Table 1.

Conclusions
This study constitutes the 14th confirmed case report of an appendicular IMT. It is important to include IMT in differential diagnoses of appendicular masses to avoid excessive resections; however, appendectomy should be sufficient for this relatively indolent lesion. Although IMT is a benign tumor, which rarely presents malignant behavior, long-term clinical and radiological follow-up is recommended due to the lack of scientific data of this particular tumor at this specific anatomical site.

Author Contributions
MGU: Conceptualization, data curation, writing original draft
ARD: Data curation, formal analysis, investigation
SEK: Project administration, supervision
RGR: Supervision and editing
GG: Validation, Writing-review & editing.

Ethical approval
All procedures performed in studies involving human participants were in accordance with the ethical standards of Tecnologico de Monterrey ethics committee and institutional review board number 122 and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Informed 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.

ORCID iD
Mauricio Gonzalez-Urquijo https://orcid.org/0000-0001-5101-1541

REFERENCES
1. Henrique P, Castronovo V, Mumm L, Ar P, Mq S. Inflammatory myofibroblastic tumor of appendix—case report. J Pediatr Surg. 2017;52:1656-1658.
2. Zhao J, Jing J, Fang Y, et al. Intra-abdominal inflammatory myofibroblastic tumor: spontaneous regression. World J Gastroenterol. 2014;20:13625-13631.
3. Kumasaka I. An interesting case of inflammatory pseudotumor appendix presenting as appendiceal perforation with peritonitis. J Pediatr Surg. 2016;51:1253-1255.
4. Karaman M, Semček ME, Čifčić AO, et al. Inflammatory myofibroblastic tumor in children: diagnosis and treatment. J Pediatr Surg. 2003;38:908-912.
5. Vajvazraghavan R, Chandrashekar R, Belagavi CS. Inflammatory myofibroblastic tumor of appendix. J Clin Pathol. 2006;59:999-1000.
6. Fragoso AC, Eloy C, Estaveco-Costa J, Campos M, Farinha N, Lopes JM. Abdominal inflammatory myofibroblastic tumor: a clinicopathologic study with reappraisal of biologic behavior. J Pediatr Surg. 2011;46:2076-2082.
7. Uludag M, Citgez B, Polat N. Inflammatory pseudo-tumor of the appendix and acute appendicitis: a case report. Acta Chir Belg. 2008;108:451-453.
8. Majumdar K, Sakhija P, Singhpreet K, Archan K, Gondal R, Agarwal A. Inflammatory myofibroblastic tumor appendix with concomitant mucosal dysplasia, simulating pseudomyxoma on preoperative aspiration cytology. J Cancer Res Ther. 2012;8:317-319.
9. Oh E, Ro JY, Gaskoer JM, Kim JW, Jung W, Yoon SO. Inflammatory myofibroblastic tumor of the appendix arising after treatment of gastric cancer: a case report and review of the literature. Am J Surg Pathol. 2014;38:657-659.
10. Bashir MR, Al Sobhani MO, Al-Rikabi AC. An unusual case of inflammatory myofibroblastic tumor of the appendix masquerading as acute appendicitis. Oman Med J. 2011;56:250-252.
11. Narasimharao KL, Malik AK, Mitra SK, Pathak IC. Inflammatory pseudotumor of the appendix. Am J Gastroenterol. 1984;79:32-34.
12. Yamagiwa YT, erada N, Hashimoto O, Ino T. An inflammatory pseudotumor of the appendix. Jpn J Cancer Clin. 1990;36:1059-1062.
13. Bonnet JP, Bassett T, Djoussé D. Abdominal inflammatory myofibroblastic tumors in children: report of an appendiceal case and review of the literature. J Pediatr Surg. 1996;31:1311-1314.
14. Khodadadi M, Sanaye S, Nikhoo B. Rectal and appendiceal inflammatory myofibroblastic tumors. Arch Iran Med. 2006;9:277-281.
15. Chan JK, Cheuk W, Shimizu M. Anaplastic lymphoma kinase expression in inflammatory pseudotumors. Am J Surg Pathol. 2001;25:761-768.
16. Jougon J, Amar A. Inflammatory pseudotumor of the appendix. Apropos of a case. Review of the literature. J Gastroenterol. 1991;26:86-88.
17. Oh E, Ro JY, Gaskoer JM, Kim JW, Jung WH, Yoon SO. Inflammatory myofibroblastic tumor of the appendix arising after treatment of gastric cancer: a case report and review of the literature. Acta Pathol Microbiol Immunol Scand. 2014;122:657-659.
18. Schoonjans C, Caluwé G, Bronckaers M. Appendiceal inflammatory myofibroblastic tumor: a rare postoperative finding. Acta Chirurgica Belgica. 2016;116(4):243-46. doi:10.1080/00015458.2016.1139940.