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Inflammatory Myofibroblastic Tumor of Appendice - Case Report

Henrique P[1], Falcão CAF[1]*, Lopes HMM[2], Paz AR[3] and Sousa MG[4]

[1]Medical Student of the Faculdade de Medicina Nova Esperança - Paraíba, Brazil
[2]Medical Science by the Medical Science College of Paraíba and Postgradute in Medical Clinic by the Municipal Server of Paraíba – Paraíba, Brazil
[3]Doctor graduated in Medicine from UFPB. Residency in Pathology by Unicamp, Specialist in Pathology SBP / AMB, Brazil
[4]Doctor and Master's degree program by the Discipline of Gastroenterology Surgical of UNIFESP/EPM-SP, Post Doctoral of UNIFESP/SP, Brazil

***Corresponding author: Falcão CAF,** Email: camillafranca@hotmail.com

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Abstract

First described in 1937, the inflammatory myofibroblastic tumor (IMT) is a rare disease of mesenchymal origin and unknown etiology. IMTs of the gastrointestinal tract are quite rare, the majority of cases originating from the appendix affects teenagers or young adult males. The manifestations depend on the affected organs, but in general, the clinical-radiological presentation is non-specific, and histopathological confirmation is needed for diagnosis. Surgical treatment is the approach of choice. However, by mimicking malignant tumors they are often resected more aggressively and unnecessarily. The relevance of this case to the scientific community lies in the fact that it is a rare entity with just a fill reported cases in appendix which represents a difficult diagnosis and can evolve aggressively. Case report: A 33-year-old young woman complaining of abdominal discomfort associated with episodes of chills and vomiting. A computed tomography scan of the abdomen showed an enlarged appendix, with an expansive lesion inside, suggestive of an appendix mucocele. An appendectomy was performed with mesoappendice resection. The histopathological examination showed mesenchymal neoplasm, with an appearance suggestive of inflammatory myofibroblastic tumor. Conclusion: The IMT can manifest several symptoms which makes it difficult to distinguish from other neoplasms, histopathological evaluation through biopsy is the adopted method. Immunohistochemistry and molecular techniques are essential, especially positive ALK whichis related to the patient's prognosis as age and location. Surgical therapy is the most accepted approach. Since it is rare and able to mimic malignant tumors, sometimes it is surgically treatedin a more aggressive way without being necessary. Therefore, this report is of great value for the medical community because the knowledge of this entities' existence as a differential diagnosis of nonspecific appendicular masses at clinical and radiological examination may reduce excessive resections and post-operative morbidity.

Introduction

First described in 1937, inflammatory myofibroblastic tumor (IMT), also known as inflammatory pseudotumor (ITP), is a rare disease of mesenchymal origin commonly found in children and adolescents [1-3]. It has been identified as the first site of the lung, since several other regions have been discovered as possible primary focus 6. IMT of the gastrointestinal tract is even rarer [4,5] with only 10 confirmed cases of appendix involvement by 2012, according to the latest Surveys. Most of these reported cases of appendix IMT are adolescents or young adult males [1,3].

The etiology of IMT is still uncertain [4,6], but probably there is an association with inflammation, trauma, viral infection, chromosome translocation and gene fusion [1,8]. Although TMI has been described as a benign lesion, there are reports of aggressive behavior by extensive local growth, recurrence and metastasis [1]. Histologically, it is characterized by myofibroblastic cells and inflammatory component which comprises in particular plasma cells, lymphocytes and histiocytes [3-5]. Since it is able to originate in different sites, the IMT can manifest several symptoms and produce distinct laboratory resultsaccording to the affected organ and tumors' size, which make it difficult to distinguish from others neoplasms. Radiological appearance does not provide great information because it is nonspecific and, therefore, insufficient to provide the diagnosis [1,3,8].

The definitive diagnosis can be obtained through the histopathological evaluation, performed from the percutaneous biopsy or withdrawal of samples directly from the lesion during diagnostic laparoscopy. Complementation with immunohistochemistry and molecular techniques are important in the investigative process [1,3,7]. The treatment of the myofibroblastic inflammatory tumor consists in resection of the tumor mass, especially in the symptomatic patients due to mass effect, and in those in which the diagnosis was not possible only with biopsy [1,2,8].

Because they have a wide variety of clinical and radiological presentations, IMT represent an important diagnostic challenge for appendicular masses, since they are, sometimes, only discovered through the histopathology of the resected piece. This fact results in more radical surgical procedures and with extensive margins without necessity, which increase, therefore, the morbidity of the operative act [3].

Case Report

A 33-year-old woman with complaints of abdominal discomfort in the right hypochondrium associated with episodes of chills and vomiting. At physical examination showed good general condition, stained, anicteric, with nonspecific pain in the right hypochondrium. No complaints of altered bowel habits or weight loss. She denied comorbidities such as diabetes mellitus and systemic arterial hypertension. After ultrasonography (USG) of the gallbladder and biliary tract showing an acalculous cholecystitis, the investigation was continued with cholangio-resonance, which didn't show any alteration. In the upper digestive endoscopy, atrophic gastritis, non-erosive esophagitis, duodenitis bulb and a positive urease test for H.pylori were identified for which the treatment was performed. Due to the persistence of abdominal pain, the USG of bile ducts was repeated, and the report described a normal gallbladder, which excluded acalculous cholecystitis. Upon physical examination of the abdomen, she presented pain in the right iliac fossa, with no other findings in the other quadrants.

An abdominal tomography scan showed an enlarged appendix image with an expansive lesion in the interior, suggestive of an appendix mucocele. We chose to perform appendectomy with resection of mesoapendice. The result of the anatomopathological study of the piece was mesenchymal neoplasia, four centimeters, with a fusocellular pattern permeated by inflammatory cells rich in plasma cells and as rare lymphoid aggregates, located in the muscular wall with focal ulceration of the mucosa and extension to the subserosus; without lymphatic or perineural invasion. An aspect suggestive of inflammatory myofibroblastic tumor.

Immunohistochemistry showed positivity for Vimentin, CD138 (in plasma cells), CD45 (in lymphocytes), Myeloperoxidase (in rare polymorphonuclear cells) and negative for ALK-1. This pattern

confirmed the hypothesis of inflammatory Pseudotumor. The patient evolved without postoperative complications and remained in outpatient follow-up.

Discussion

The IMT is a rare benign neoplasm of mesenchymal origin and unknown etiology [1,2,4], initially described in 19372. Due to its variety of presentations, it can mimic, clinically and radiologically, malignant tumors, especially sarcoma [5,6]. Despite being able to develop in any anatomical location [1,2,5], the gastrointestinal tract is an uncommon site of IMT and, even more so, when the primary focus is the vermiform appendix. When we approach this topic in the literature, it is noticed that most of the reported cases are in adolescents and young adult males [1,3] what makes the discussion of this pathology more relevant when affecting a young adult woman, as in the case studied.

Although IMT is considered a benign condition, there are reports of aggressive behavior by extensive local growth, recurrence and metastasis [1]. The sites may influence the prognosis of these tumors - hepatic lesions have a more favorable outcome; mainly when compared to other sites of the abdomen, pelvis and retroperitoneum, which tend to present a more aggressive behavior and poor prognosis. Otherassociations found with the location of these masses are age and gender. The hepatic locus is usually identified in middle-aged and elderly men, also this population is mostly related to cases of tumors' regression [1]. Histologically, inflammatory pseudotumors are characterized by myofibroblastic cells forming areas of myofibroblasts in the form of a spindle and an inflammatory component comprising in particular plasma cells, lymphocytes, eosinophils and histiocytes [3-5,8].

Based on the location, IMT can manifest several symptoms.If the lesionis at the appendix, it may present acute appendicitis, intestinal obstruction, abdominal pain, abscesses.Also the laboratory results are incapable of distinguish this entity from other neoplasms [1,3,7]. Image exams do not provide great information due to the multiplicity of presentations, such as cavities, abscesses and more commonly solid, irregular mass with well-defined contours associated or not with central necrosis [9]. Such characteristics can be attributed to several pathologies, which makes the findings unspecific and insufficient to provide definitive diagnosis [1,3,7]. The histopathological evaluation performed by percutaneous biopsy or removal of lesion's fragments during videolaparoscopy is the adopted method to diagnose IMT. Immunohistochemistry and molecular techniques are an important step of the investigative process to distinguish this neoplasm from other soft tissue tumors [1,7,8].

Studies have shown that most cases of IMT are positive for actin, vimentin and keratin to immunohistochemistry, as well as the importance of ALK expression at the molecular level as a possible specific marker for this neoplasm [1,3]. Since it can present positivity in 36% to 60%, with some articles referring up to 100% of the cases depending on the place of origin of the tumor; In addition, the expression of ALK can be considered a prognostic factor because it's able to estimate the aggressiveness of inflammatory pseudotumors [1,8].

The presented case presents a good prognosis, since the tumor was resected with free margins and the immunohistochemistry, the main marker of aggressiveness of this neoplasm, the ALK, was negative. However, because localization is also a prognostic factor, outpatient follow-up is part of the management for these tumors.

Surgical resection remains the most effective form of treatment of inflammatory myofibroblastic tumor [4,5,7,8]. Although there are reports of spontaneous resolution and regression of the tumor mass through the use of antibiotics, corticosteroids and nonsteroidal anti-inflammatory drugs, this conservative approach should only be taken into consideration if the lesions are unresectable to the imaging exams as Computed tomography or diagnostic videolaparoscopy [1].

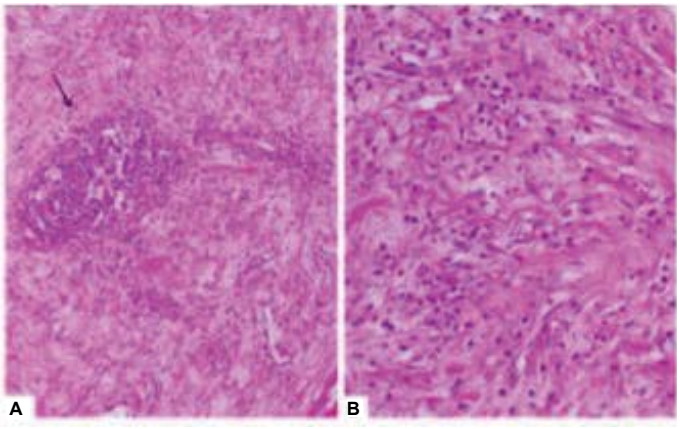

Figure 1: Photomicrograph.
(A) showing lymphoid agregates (arrow) collagenization and intense myofibroblastic proliferation (HE stain, 200x); (B) polymorphic infiltrate comprising of plasma cells, lymphocytes, histiocytes and few neutrophils in stromal fibrosis (HE stain, 200x).

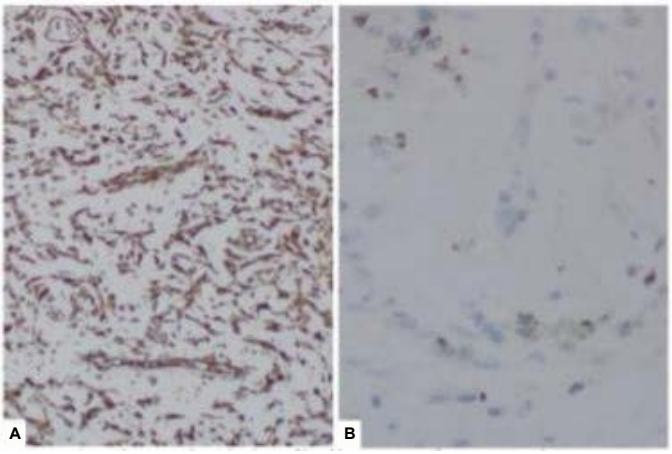

Figure 2: Immunohistochemistry (400x).
(A) myofibroblasts positive for vimentin. (B) CD138 positivity in plasma cells.

The IMTs located in the gastrointestinal tract have a high rate of recurrence in relation to the other sites, so surgical therapy is the most accepted approach and should be adopted in all symptomatic patients by mass effect and in those in whom the diagnosis was not possible only with Biopsy [1,2]. For multiple and invasive lesions chemotherapy may be considered [2] and adjuvant radiotherapy for local recurrence or incomplete removal of the lesion8. However, the diagnosis should be prioritized as early as possible to avoid the adoption of more aggressive surgical margins as necessary (Figure 1 and 2).

Conclusion

The present study is of great relevance to the scientific community for reporting a very rare case of appendicular IMT in young adult women. The inflammatory pseudotumors have a variable prognosis according to age, location and positivity of ALK. Surgery remains the most appropriate treatment; however, since they present non-specific clinical signs and symptoms similar to malignant tumors, they are commonly approached aggressively, adding more morbidity to the operative event. Therefore, it is necessary to include IMT in differential diagnoses of appendicular masses in order to reduce excessive resections and is important to carry out the long-term follow-up in these patients to evaluate recurrence ormetastases- despite the low risk.

Citation: Henrique P, Falcão CAF, Lopes HMM, et al. Inflammatory Myofibroblastic Tumor of Appendice - Case Report. J Hepatol Gastroenterol 2017; 1:006.

References

1. Zhao JJ, Ling JQ, Fang Y, Gao XD, Shu P, Shen KT, et al. Intra-abdominal inflammatory myofibroblastic tumor: Spontaneous regression. World J Gastroenterol. 2014; 20:13625-13631.
2. Zhang C, Li CJ, Zong WK, Xu ML, Fan GW. One case of inflammatory myofibroblastic tumor - a case report. J Cancer Res Ther. 2015; 11 Suppl 1:C131-133.
3. Majumdar K, Sakhuja P, Kaur S, Rastogi A, Gondal R, Agarwal A. Inflammatory myofibroblastic tumor appendix with concomitant mucosal dysplasia, simulating pseudomyxoma on preoperative aspiration cytology. Journal of Cancer Research and Therapeutics. 2012; 8:317-319.
4. Schoonjans C, Caluwé G, Bronckaers M. Appendiceal inflammatory myofibroblastic tumor: a rare postoperative finding. Acta Chir Belg. 2016; 116:243-246.
5. Uludag M, Citgez B, Polat N. Inflammatory pseudo-tumour of the appendix and acute appendicitis: a case report. Acta Chir Belg. 2008; 108:451-453.
6. Bonnet JP, Basset T, Dijoux D. Abdominal inflammatory myofibroblastic tumors in children: report of an appendiceal case and review of the literature. J Pediatr Surg. 1996; 31:1311-1314.
7. Vijayaraghavan R, Chandrashekar R, Belagavi CS. Inflammatory myofibroblastictumour of appendix. J Clin Pathol. 2006; 59:999–1000.
8. Cantera JE, Alfaro MP, Rafart DC, Zalazar R, Muruzabal MM, Barquín PG, et al. Inflammatory myofibroblastic tumours: a pictorial review. Insights Imaging. 2015; 6:85-96.

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