

Multiple intrabdominal fibromatosis: finding during appendectomy. Case report

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ABSTRACT

A male patient with mixed skin, 32 years old, with a health history, was seen in the Surgical Guard Corps of the Hospital Héroes del Baire, Isla de la Juventud, Cuba, due to acute abdominal pain, which was interpreted as acute appendicitis and underwent emergency surgery. An appendectomy was performed. During the intraoperative finding, an irregularly shaped, mamelonating tumor was found in the mesentery of the terminal ileum and alleged multiple tumor "seeds" in the greater omentum. The primary tumor and the greater omentum were resected and sent for biopsy, which reported the diagnosis: multiple intra-abdominal fibromatosis. The patient had a satisfactory post-surgical evolution.

Keywords: Mesenterium, Terminal ileum, Major omentum, Multiple Intra-bdominal Fibromatosis

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Fibromatosis múltiple intrabdominal: hallazgo durante apendicectomía. Presentación de caso

RESUMEN

Paciente masculino, de piel mestiza y 32 años de edad, con antecedentes de salud, quien fue visto en el Cuerpo de Guardia de Cirugía del Hospital Héroes del Baire, Isla de la Juventud, Cuba, por dolor abdominal agudo, que fue interpretado como apendicitis aguda e intervenido quirúrgicamente de urgencia. Se realizó apendicectomía y durante el transoperatorio se encontró, como hallazgo, un tumor de forma irregular, mamelonante, en el mesenterio del íleon terminal y supuestas «siembras» tumorales múltiples en el epiplón mayor. El tumor primario y el epiplón mayor fueron resecados y enviados a biopsia, que informó como diagnóstico: fibromatosis múltiple intrabdominal. El paciente tuvo una evolución posquirúrgica satisfactoria.

Palabras clave: Mesenterio, Íleon terminal, Epiplón mayor, Fibromatosis Múltiple Intrabdominal

Fibromatose intra-abdominale multiple: découverte lors d'une appendicectomie. Présentation de cas

RÉSUMÉ

Patient masculin, à peau mixte, âgé de 32 ans, avec des antécédents médicaux, qui a été vu dans le Corps de Garde Chirurgical de l'Hospital Héroes del Baire, Isla de la Juventud, Cuba, en raison de douleurs abdominales aiguës, interprétées comme une appendicite aiguë et nécessitant une intervention chirurgicale d'urgence. Une appendicectomie a été réalisée et lors des constatations peropératoires, une tumeur mamelonnante de forme irrégulière a été trouvée dans le mésentère de l'iléon terminal et de multiples «graines» tumorales présumées dans le grand omentum. La tumeur primitive et le grand omentum ont été résequés et envoyés pour une biopsie qui a rapporté le diagnostic: fibromatose intra-abdominale multiple. Le patient a eu une évolution post-opératoire satisfaisante.

Mots clés: Mésentère, Iléon terminal, Omentum majeur, Fibromatose intra-abdominale multiple

Fibromatose intra-abdominal múltipla: achado durante apendicectomia. Apresentação de caso

RESUMO

Paciente masculino, de pele mestiça e 32 anos de idade, previamente saudável, foi atendido no Corpo de Guarda de Cirurgia do Hospital Héroes del Baire, Isla de la Juventud, Cuba, devido a dor abdominal aguda, interpretada como apendicite aguda, sendo submetido a uma apendicectomia de urgência. Durante o transoperatório, foi encontrado, como achado intraoperatório, um tumor de forma irregular, mamelonado, no mesentério do íleo terminal, além de possíveis implantações tumorais múltiplas no epíplon maior. O tumor primário e o epíplon maior foram ressecados e enviados para biópsia, cujo diagnóstico foi: fibromatose intra-abdominal múltipla. O paciente teve evolução pós-operatória satisfatória.

Palavras -chave: *Mesentério, Íleo terminal, Epíplon maior, Fibromatose intra-abdominal.*

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INTRODUCTION

Fibromatosis was initially proposed by Scout¹, a rare tumor disease resulting from the proliferation of fibroblasts and/or myofibroblasts¹⁻⁴. Its histology is benign^{1,3,5}, although it invades locally^{3,4}, and it can have several locations, with the intra-abdominal location being the least common^{6,7}, where the mesentery of the small intestine is the most affected^{1-4,7,8}, followed in frequency by the retroperitoneal, the mesocolon², the cecal appendix, and others⁷.

Its etiology has not yet been fully clarified⁴. Some authors suggest that it is more common in women than in men^{3,5-7,9,10}, while others deny significant differences in sex or race⁸. The low frequency of its appearance and the *sui generis* characteristics of the presentation of this case motivated us to produce this publication.

PATIENT INFORMATION

A male patient with mixed skin, 32 years of age, who is personally and family healthy and has a healthy lifestyle, attended the Surgery ward of the "Héroes del Baire" General Teaching Hospital on the Island of La Juventud, Cuba. 24 hours before, he had begun with acute abdominal pain, which started as periumbilical discomfort, which later migrated to the right iliac fossa, where it progressively intensified. This was accompanied by nausea, vomiting, loss of appetite, and low-grade fever, the latter of which appeared more recently.

CLINICAL FINDINGS

During the physical examination, an involuntary contracture in the right iliac fossa was found in the abdomen. It was painful on palpation, sudden decompression, percussion, and pain when coughing, which was interpreted as a peritoneal reaction. He also tended to tachycardia since his heart rate was found to be 96 beats/minute. Added to all this was a temperature of 37.5° C.

CALENDAR

Symptom	Appearance time
Abdominal pain	≈ 24 hours
Nausea	≈ 24 hours
Vomit	≈ 24 hours
Loss of appetite	≈ 24 hours
Low fever	≈ 3 hours

DIAGNOSTIC EVALUATION

The clinical picture made up of the interrogation that provided the symptoms, as well as their sequence of appearance, plus the signs found in the physical examination that denoted a peritoneal reaction, in addition to the complementary indicated ones, in which only the Leukogram was notable, which showed mild leukocytosis with shift to the left; a diagnosis of peritoneal syndrome was made, the most probable etiology of which turned out to be acute appendicitis, and as such he was prepared to undergo emergency surgery.

THERAPEUTIC INTERVENTION

During the emergency laparotomy, acute suppurative appendicitis was confirmed, so an appendectomy was performed without complications. However, as an intraoperative finding, a tumor of around 10 (ten) centimeters in diameter, with an irregular mamelonated multilobulated surface was found, protruding at the level of the mesentery of the terminal ileum, which seemed not to infiltrate any neighboring organs (photos 1 and 2).

In a subsequent review, it was observed that the greater omentum presented multiple similar lesions but considerably smaller in size, showing tumor "seeds" (photos 3 and 4). No other lesions were found in the liver or the rest of the abdominal cavity. The primary tumor and the entire greater omentum were resected and sent for histological study to the Department of Pathological Anatomy of this hospital, the result of which was confirmed by the National Reference Center for Pathological Anatomy (CENRAP) at the Hermanos Ameijeiras Hospital, Havana.

The final histological result was Multiple Fibromatosis, with the following immunohistochemical tests expressed: CD 117, α Actin and S 100 (negative), and Vimentin (positive). The patient presented a satisfactory post-surgical evolution in the ward without complications and was discharged a few days later.

MONITORING AND RESULTS

A few days later, he was evaluated in a postoperative consultation and was in perfect condition. Another type of treatment and follow-up was impossible for us to carry out since the patient moved outside the country shortly after.

DISCUSSION

Fibromatosis, also known as a desmoid tumor, comprises only 0.03% of all tumors^{2-4,9,10}, most frequently oscillating from 1:1,000,000⁶ to 5:1,000,000 per year^{1,3,7,8,10}. It has benign characteristics^{1,3,5} and can be single or multiple^{1,8}. It can be increased slowly^{1,3,9} and repeated following surgical extirpation^{4,9} it is invasive in a local form^{2,4,9} but does not create metastasis^{1-4,6,8-10}. The most common appearance interval is variable.

Authors such as Palacios-Fuenmayor place it between 25 and 35 years old³, just as our patient, but Velázquez, Sabilón y Molina place it between 40 and 60 years old⁶. Although its etiology is not entirely known, the following are proposed as risk factors: previous abdominal surgeries or traumas, pregnancy, female, estrogen treatments^{8,10}, family history of the condition, familial adenomatous polyposis⁹⁻¹¹, Gardner's syndrome^{1-5,7,9,11} and some genetic alterations such as trisomies 8 and 20^{1,2,6,10}, contrary to our case, which was male and did not have any history mentioned in other publications.

Its location can be extra-abdominal (60%), abdominal wall (25%), or intra-abdominal (8-15%)^{6,7}, which is the most aggressive due to its ability to infiltrate pelvic or abdominal organs⁷. The most common location within the abdomen is the mesentery of the small intestine^{2-4,7,8}, just like in this case, in which the surgeons' attention was drawn to the presence of multiple "seeds" in the greater omentum, an element of which the authors could not find reference in the literature consulted.

Its imaging diagnosis can be made by Ultrasound, Computed Axial Tomography, preferably with contrast^{2,4,9}, although some authors prefer Nuclear Magnetic Resonance^{3,9}. However, its definitive diagnosis is made by biopsy and immunohistochemistry, where the Vimentin result is usually positive^{2,3,9,10}, as in the reference case. However, our diagnosis was entirely incidental during emergency surgery during acute appendicitis.

The treatment is surgical^{2-5,8,9,11}, which was the one performed in our case, although chemotherapy and radiotherapy can also be used^{1-3,5,7,9,10}, as well as anti-inflammatories, hormonal agents^{1,4,9-11}, cytotoxic agents or molecular therapies^{2,3,11}. Patient follow-up must be guaranteed at the end of treatment due to the high rate of local recurrence that accompanies these tumors², which was not possible with the patient in our study due to his definitive departure from the country.

This research allowed us to conclude that fibromatosis or desmoid tumor appearance is not frequent, the intra-abdominal location is the rarest, and the mesentery of the small intestine is its most common site. Our study coincided with this. Most publications talk about the presentation as a single tumor; however, in this case, there were multiple. Surgery continues to be considered the treatment of choice, and patients should be followed up postoperatively due to the possibility of recurrence.

PERSPECTIVE OF THE PATIENT

The patient was grateful for the work of the doctors who diagnosed and resolved the health problem.

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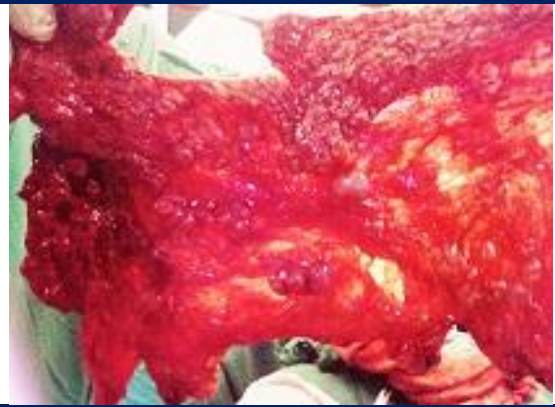
1. Mesoileon tumor



2. Mesoileon tumor.



3. T mesoileon and «seeds» in Epiplon mayor.



4. Mayor Epiplon with multiple «seeds»

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CONFLICTS OF INTEREST

No conflicts of interest are declared.

Authors' contributions

Conceptualization: Iván Baigorria Ortega, Maitte Puente Peña, José Luis Manso Ramírez

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All authors approve the final version of the manuscript.



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