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Case Report

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Desmoid Tumor Mimicking a Strangulated Hernia: About A Case

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Abstract

Desmoid tumors of the abdominal wall are very rare. Nonspecific clinical presentation may lead to misdiagnosis. Radiological explorations are very contributive to help guide the diagnosis. Histological and immunohistochemical examination are mandatory to confirm the diagnosis. In our case, a rare desmoid tumor mimicked a strangulated hernia on a Pfannenstiel scar. The patient was operated. Preoperative findings revealed a solid tumor developing from the muscles of the anterolateral abdominal wall. The tumor was resected taking away a macroscopically healthy tissue. The peritoneum was not invaded and a polypropylene mesh has been set. The postoperative follow-up was simple. Anatomopathological examination associated with immunohistochemical determination concluded to a desmoid tumor.

Keywords: Desmoid Tumor, Strangulated Hernia, Surgery.

Introduction

Desmoid tumor (DT) is a very rare entity representing less than 0,003% all tumors [1]. It is defined histologically by a monomorphic fibroblastic proliferation of soft tissues, classified as intermediate tumors in the 2020 WHO classification, with local malignancy (infiltrating, invasive tumor) without metastatic potential [2]. Representing 3,5 % of fibrotic tumors, they have a high recurrence potential [1]. Diagnosis is difficult, due to rarity of this tumor and the lack of specific clinical expression. Patients usually consult late. Symptomatology is dominated by the appearance of a palpable tumor sometimes associated with pain, or by signs of compression. Certain radiological elements, such as computed tomography, and magnetic resonance imaging, can provide great diagnostic help. Especially since the clinical presentation can be misleading.
Only histological examination provides a definitive diagnosis.

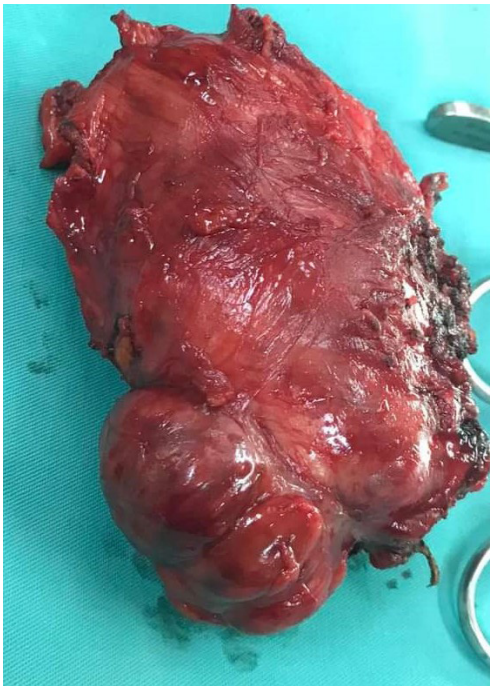

Figure 1: Surgical specimen with macroscopically healthy muscle tissue.

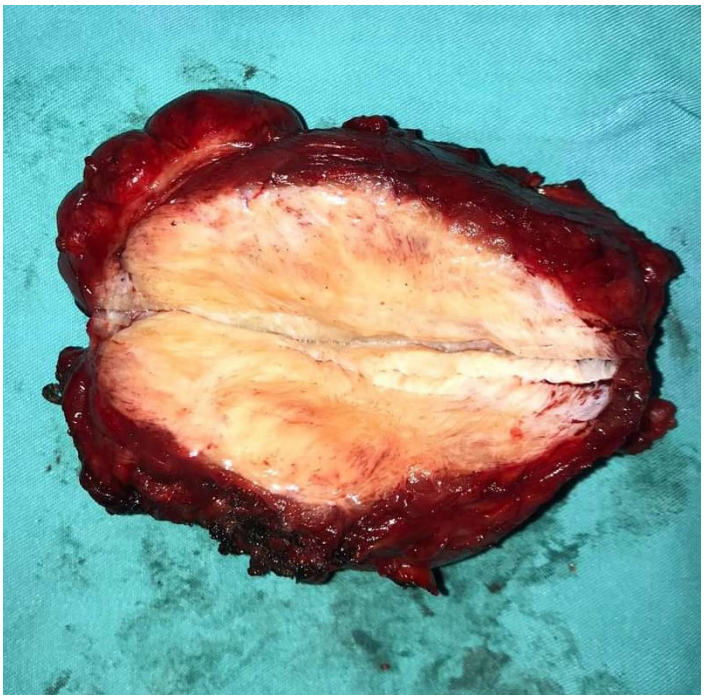

Figure 2: Sectional surgical piece.

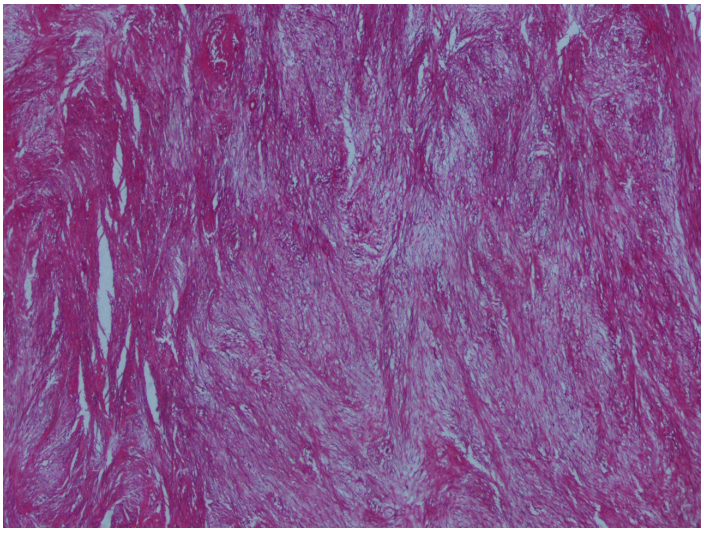

Figure 3A: Microphotograph showing a homogeneous myofibroblastic proliferation made of long bundles

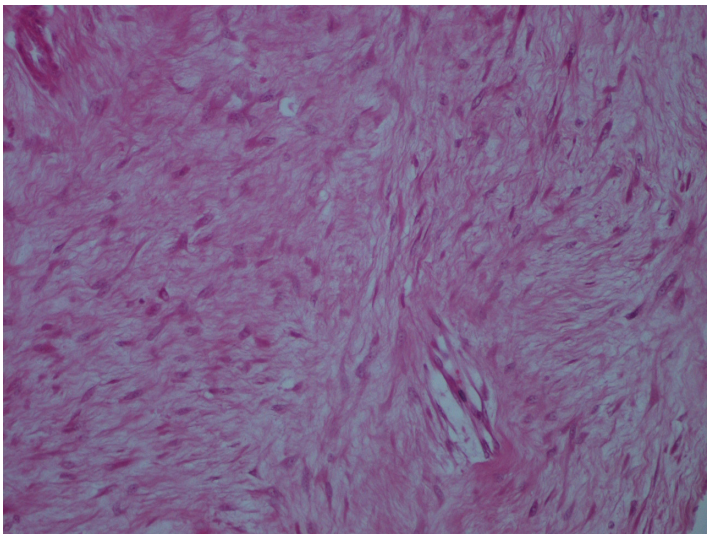

Figure 3B: the tumor cells are not very atypical

Case report

A 47-year-old patient with a history of a Pfannenstiel caesarean presented to the emergency room with abdominal pain that had been evolving for 48 hours. The history of her family is unremarkable. Anamnestic data revealed the notion of nausea and gas stoppage. The patient noticed the appearance of a mass next to the Pfannenstiel scar for which she did not consult. Until recently this mass was completely painless, but non-impulsive to cough.

Clinical examination found an abdominal mass 5 cm long axis next to the Pfannenstiel scar. This mass was slightly painful, mobile relative to deep planes. The abdomen was bloated. Rectal examination was normal.

An abdominal x-ray showed a single air-fluid level.
The diagnosis evoked was a strangulated eventration on Pfannenstiel scar. The decision was to operate the patient after short resuscitation with no further radiologic exploration.

We took over the old scar, dissection revealed a 6cm mass developing from the muscles of the antero-lateral wall of the abdomen. This mass did not infringe the peritoneum. We proceeded to the excision of the tumor through macroscopically healthy muscle tissue.

Reconstruction of the abdominal wall was made using a polypropylene mesh which was attached to the anterosuperior iliac spine, Cooper's ligament and pubic spine. The postoperative course was simple.

Histological examination revealed mesenchymal proliferation made up of myofibroblast-like spindle cell bundles. Further immunohistochemical study showed highly positive Beta catenin expression. CD117, CD34, PS100 and Desmine were negative. It concluded to a histological and immunohistochemical profile of a desmoid tumor.

A total colonoscopy was conducted and was normal. Patient is now

five months post operative. No clinical signs of local recurrence. CT scan and a total colonoscopy were conducted 3 months after surgery and showed no abnormality.

Discussion

Desmoid tumors are rare mesenchymal tumors [1]. Desmoid tumors are localized in the abdominal wall in 30% of cases. In 15% to 20% they are associated with familial adenomatous polyposis [2]. In the majority of cases, they are sporadic (85% to 90%) and are associated with a mutation of the beta cadenin CTNNB1. these tumors have a female preponderance (2/3 of cases) with a median age of diagnosis of 35 to 44 years old [2]. The history of abdominal trauma or surgery is considered to be a risk factor for developing desmoid tumors [1, 2].

Clinical presentation may vary due to its localization and its aggressivity. they can be locally aggressive and have increased growth causing a mass effect or local invasion without metastatic potential [3]. In our case the clinical presentation mimicked a strangulated eventration and surgery was undergone urgently. Therefore, in doubt further explorations should be undertaken when there is atypical clinical picture and an evocative context.

Radiologic examinations are of paramount importance in the diagnosis of desmoid tumor. With abdominal wall localization, as in our case, MRI is the mainstay [1, 3, 4]. It is often an ovoid or infiltrating mass, with generally lobulated boundaries, presenting a homogeneous iso or hypointensity in the T1-weighted sequence and often an hyperintensity in the T2 sequence. The contrast gain is intense and heterogeneous after injection of gadolinium.

CT scan is preferred in the intraabdominal localization. It appears as an heterogenous enhanced lesion, more or less well-defined tissue lesions with spiculated contours. It permits to identify intraabdominal complications [2].
The diagnosis of certainty is based on histological examination completed with an immunohistochemical study. Molecular analysis in search of pathogenic variants of the ß-catenin gene (CTNNB1) or APC is to be carried out systematically [4-6].

The specimen can be obtained, after appropriate imaging assessment, with multiple core needle biopsies.

Surgery was les first line therapy (4) till the year 2000. Retrospective series have shown progression-free survival rates of 50% at 5 years for asymptomatic patients managed with a frontline conservative watch and see approach [7-9]. One of the most recent and largest series comparing initial surgery to initial observation was reported by Penel et al. The results did not show any difference in event-free survival [10]. In this French, nationwide prospective study, including 771 patients, anatomic location seemed to influence the course of the disease [10]. The authors distinguished favorable location which were abdominal wall, intra-abdominal, breast, digestive viscera and lower limb. Patients with unfavorable locations, defined as chest wall, head & neck and upper limb, had a significantly better 2 years out come with a conservative strategy. In the first group the 2 years outcome (event free survival) was the same for patients treated with a conservative strategy compared to those who underwent surgery [11].

Furthermore, surgery is not the only treatment option. Several papers in the literature defined active treatment for desmoid tumors as being surgery, radiotherapy and systemic treatment. The Milan guidelines of 2020 recommend the use of active treatment should be considered in case of persistent progression [11]. However, when the disease is located close to a critical structure that may pose significant problems to the patient's life, or evidently in case of a complication, active therapy may be established first intension.

For abdominal wall localization, surgery is still the first option in case of progression [11]. The is no consensus regarding abdominal wall reconstruction after resection [12]. Most authors use mesh of synthetic material that could be in contact with the bowel [12].

In our case peritoneum was intact and we could fix a polypropylene mesh in a prefacial plane. Systemic treatment options comprise antihormonal therapies (tamoxifen), non-steroidal anti-inflammatory drugs, tyrosine kinase inhibitors (imatinib), and conventional chemotherapeutic regimens [1, 11].

Conclusion

Desmoid tumors of the abdominal wall are rare. They are locally aggressive tumors with a high risk of local recurrence.
The clinical manifestations of these tumors are atypical and may lead to diagnostic errors. In doubt radiologic exploration may provide critical information. MRI is showing to be the most contributive especially in abdominal wall localization. When the diagnosis is suspected, biopsy and histological examination must be done to confirm diagnosis. CTNNB1 and APC mutation must be systematically sought.

Conservative treatment should now be proposed in first intension providing close monitoring by MRI as recommended in the literature.

In case of persistent progression, complication or particular localization active therapy should be considered.

In our case, unusual clinical presentation misled us in our diagnosis. In doubt, surgeon must keep in mind this rare diagnosis and further exploration should be underwent to avoid unnecessary surgery.

Acknowledgments

None.

Conflicts of interest

None.

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