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Case Report

Absceded abdominal wall desmoid tumor: a case report

Ana Karen Mena Zenteno[1]*, Aldo Olvera[1], Cynthia Sánchez[1], Jorge A. Montero[2]

[1]General and Gastrointestinal Surgery Department, Hospital General Regional No.1, Santiago de Queretaro, Queretaro, Mexico
[2]Department of Diagnostic and Therapeutic Imaging, UMAE 1 BAJIO, Guanajuato, Mexico

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***Correspondence:**
Dr. Ana Karen Mena Zenteno,
E-mail: dr_AKMZ@outlook.com

ABSTRACT

Desmoid tumor is a rare type of tumor dependent on a well-differentiated fibroblastic monoclonal proliferation with a high capacity for non-metastatic local invasion, it constitutes <3% of soft tissue neoplasms, an annual incidence of 5-6 cases per million is reported, its etiology is unknown and its clinical presentation depends on the location of the tumor. We presented a case of a 53-year-old female patient who presented to the emergency department with clinical characteristics of abdominal wall abscess with no significant history of its appearance. Abscess drainage plus biopsy of muscle tissue in the anterior region of the abdominal wall was performed, histopathology report compatible with desmoid tumor, radical surgical treatment was decided in a second intervention. Desmoid tumor is rare, since its diagnosis and treatment require a multidisciplinary approach; Active surveillance is currently the treatment of choice for patients who have DD in noncritical locations, and at least 1 to 2 years of active surveillance is now recommended because of its likelihood of spontaneous regression. The main objective of all existing therapies is to preserve or improve the quality of life of the patient.

Keywords: Desmoid tumor, Abdominal wall, Abscess, Surgery

INTRODUCTION

Desmoid tumor is a rare type of tumor dependent on a well-differentiated fibroblastic monoclonal proliferation with a high capacity for non-metastatic local invasion, it constitutes <3% of soft tissue neoplasms, an annual incidence of 5-6 cases per million is reported, its etiology is unknown.[1-3] Its etiology is unknown, it can be associated with familial adenomatous polyposis syndrome or Gardner syndrome, its clinical presentation is nonspecific and depends on the location of the tumor. Like all tumors, a biopsy is necessary to establish its diagnosis, plus immunohistochemistry. Some reference centers have reported rates of misdiagnosed cases of up to 30%-40% during the initial study.[3-13] The specific type of intervention is controversial, usually guided according to the anatomical site.

CASE REPORT

A 53-year-old woman, with a history of an abdominal tumor of one month of evolution without prior medical attention, presented to the emergency room due to the presence of chronic abdominal pain exacerbated at the tumor site, without signs of an acute abdomen or digestive symptoms, accompanied by feverish spikes. and analysis with presence of leukocytosis with deviation to the left, on physical examination presence of increased volume in the right flank of approximately 20×15 cm, of hard consistency, not mobile, accompanied by peritumoral erythema, heat and pain on palpation without signs of peritoneal irritation.

A contrast-enhanced tomographic study was taken, showing an image suggestive of an abscess limited to the

abdominal wall; based on the clinical and image, surgical intervention is decided; abscess drainage, debridement, surgical lavage, biopsy and culture are performed. With the following trans-surgical findings: abdominal wall abscess, 2000 cc of purulent fluid were drained, integral aponeurosis does not communicate with the peritoneal cavity, indurated muscle tissue in the anterior region secondary to a probable tumor, so an excisional biopsy was taken, abdominal wall was left open. To continue with healing and surgical cleansing, we started an antibiotic therapy scheme with post-surgical improvement after drainage without the presence of data of systemic inflammatory response, report of abscess culture positive for enterococcus faeceum; after complying with guided antibiotic therapy, a negative culture was obtained, with a positive histopathology report for desmoid tumor, the patient was evaluated by a specialist in oncological surgery and was scheduled for elective surgery.

TAC

Tomographic images were observed in axial, coronal and sagittal sections with application of oral contrast, where a single, rounded image with well-defined edges, homogeneous with soft tissue components, with multiple air bubbles in its surface, can be seen at the level of the right lower quadrant. periphery, this image creates a mass effect on adjacent structures and presents striation of peritumoral fat (Figure 1).

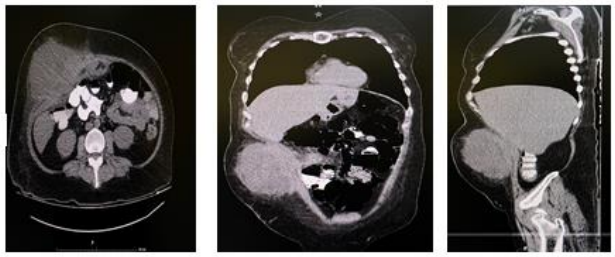

Figure 1: Computed tomography image in which a tumor is observed in the abdominal wall suggestive of an abscess.

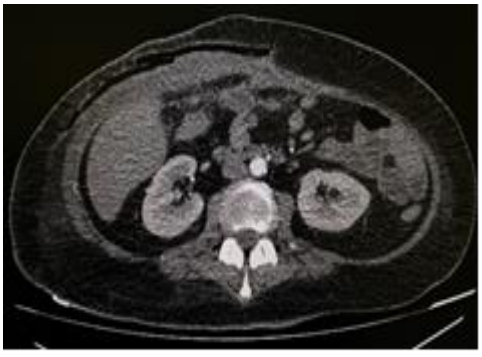

Figure 2: Computed axial tomography taken after the first intervention demonstrating increased size of the muscles in the right hemi-abdomen.

A tomographic image was observed with axial section with application of intravenous contrast where thickening of the abdominal wall muscles can be seen in the right hemiabdomen. Upon application of contrast, discrete enhancement of the adjacent muscles in the venous phase was observed, as well as striation of fat and subcutaneous emphysema secondary to a previous surgical event (Figure 2).

Histological study

Macroscopic description: A 9×9×4 cm specimen was sent to histopathology and reported as an abdominal wall tumor.

Microscopic description: Mature adipose tissue and muscle wall with extensive acute and chronic xanthomatous inflammation, delimited by proliferation of fibroblasts and hyalinized connective tissue.

Immunohistochemical study: It was negative for calretinin, smooth muscle actin and CD34 and positive for B-catenin.

Final pathological diagnosis: Desmoid-type fibromatosis, negative for malignancy.

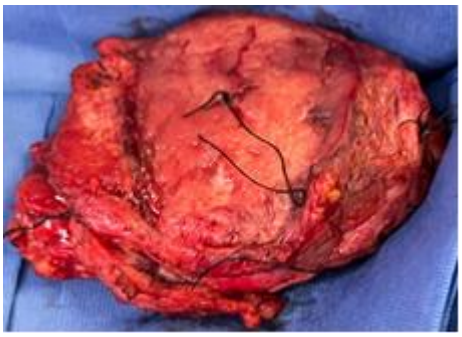

Figure 3: Surgical specimen after its resection referred with silks for marking.

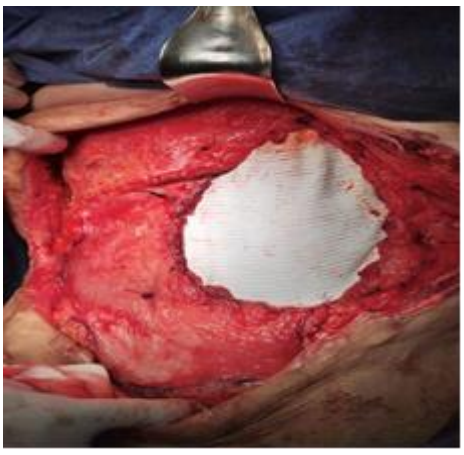

Figure 4: Aponeurotic defect requiring placement of a 15×15 cm non-absorbable expanded polytetrafluoroethylene synthetic mesh.

Therapeutic intervention

With the presence of an abdominal wall without evidence of an active infectious process and with a histopathological and immunohistochemical report of a desmoid tumor, due to the association of chronic abdominal pain plus a history of abscess, it was decided to approach surgery by performing *en bloc* resection where multiple tumor adhesions were found, loop-wall resecting a tumor of approximately 15×15 cm. The abdominal wall is reconstructed with placement of dual mesh in the aponeurotic defect and is concluded with primary closure of the subcutaneous cellular tissue and skin

Monitoring and results

The patient was discharged 96 hours after surgery, with an appointment to the surgical oncology clinic in one month, with adequate progress without local or general complications. Without receiving additional treatments, due to the report of negative pathological margins, R0 surgery. Surveillance, appointment and imaging control are suggested in 6 months to assess recurrence.

DISCUSSION

Desmoid tumors (DT) are rare lesions, representing 0.03% of all neoplasms and <3% of soft tissue neoplasms.[1,2] The majority of these tumors are associated with mutations in the CTNNB1 gene that encodes the b-catenin protein.

One of its main characteristics is infiltrative growth without generating metastasis and a high percentage of recurrence after surgery. There is a characteristic age peak for this disease (approximately 30-40 years), trauma, being a woman, familial adenomatous polyposis (PAF) and previous surgery are known risk factors.[2] A female predominance is recognized with around 70% and is associated with pregnancy, with development or growth being possible during or after it.[1]

TD can be classified in two ways: as sporadic and another associated with familial adenomatous polyposis. Most cases (80%) occur sporadically, but 2-15% of patients with desmoid tumor have associated familial adenomatous polyposis or Gardner syndrome.[4] 50% are located in the medial abdominal wall (rectus abdominis), but 40% are extra-abdominal (mainly in the extremities and mesentery).[5]

The clinical presentation varies from asymptomatic to disabling and is related to location, tumor size, and growth rate may be misdiagnosed in up to 30% to 40% of cases and may be associated with an unpredictable course of the disease, including spontaneous regressions in 20% to 30% of patients who are followed for 2 to 3 years.[2,6] There may be sites where regression is more common (i.e., abdominal wall) however regression has been observed at all sites.[3] Patients with TD often have a palpable mass at diagnosis.[6] In the clinical case described, it presents as an abdominal wall abscess without any other history or clinical association for its development other than the underlying tumor. This finding was beyond the clinical manifestations reported in the literature, which led us to make the timely diagnosis.

As an office study to assess its extension, size, invasion to adjacent structures, therefore its resectability as well as recurrences, we have CT and MRI, the first mainly for when the tumor is intra-abdominal and the second especially in the extremities since it can show tumor infiltration into the muscle and distinguish the limits between vital structures and fascial planes.[2] The definitive diagnosis is by histopathology and immunohistochemistry. In accordance with the rarity of TD and the multiple potential histological mimics, some reference centers have reported rates of misdiagnosed cases of up to 30%-40% during the initial study. In a review of 320 surgical specimens diagnosed as desmoid-type fibromatosis, 94 lesions were misclassified. A wide range of reactive and neoplastic soft tissue tumors were identified in this series, including 4 sarcomas. Many of these lesions can be distinguished from desmoid tumor by correlating the histologic findings with pertinent clinical data. It is strongly recommended that the diagnosis of TD be confirmed by an expert soft tissue pathologist and ideally, mutational analysis should be seriously considered in cases with equivocal or uncertain diagnosis.[3-13] The treatment is controversial, in recent years multiple therapies have been studied, without there being one of choice and having to be adjusted to each case.

The treatment is controversial, in recent years multiple therapies have been studied, without there being one of choice and having to be adjusted to each case. There has been a paradigm shift towards more conservative management of TD. Immediate surgery is no longer the standard treatment and available systemic therapies have increasingly become the focus of interest.[11]

The specific type of intervention is usually guided according to the anatomical site, and the decision should be made with the patient in a stepwise approach according to the latest treatment algorithm published by The Desmoid Tumor Working Group.[7] Among the possible treatments are surgery, clinical follow-up, targeted therapies, medical management, radiotherapy, chemotherapy and a combination of these.

The management of asymptomatic patients with initial observation can be considered, regardless of the site and size of the tumor, but only under the supervision of a team experienced in connective tissue tumors from a reference center to minimize the risk of active surveillance and avoid unnecessary surgeries. debilitating or mutilating when possible.[8]

Currently, the indication for surgery was based on the presence of progressive symptoms and increase in size.[7] In the case of our patient, due to her atypical presentation of an abscessed tumor associated with abdominal pain and with the approval of the patient, surgical management was decided. They are benign tumors, they do not have the capacity to metastasize, but they do have local aggressiveness, so a resection as wide as possible is indicated to guarantee tumor-free margins R0 (negative microscopic margins) and R1 (positive microscopic margins) to reduce the possible risk of local recurrence. 35% relapse five years after surgery. Relapses seem to be more frequent in women over 30 years of age.[9]

The local recurrence could be larger than the primary tumor. Some clinical parameters are associated with a high risk of relapse after surgery in different studies; These parameters include young age, large tumor size, positive margins, Ki67 expression.[10-12]

Although the risk of local recurrence appears to be lower after a combined modality with radiotherapy, surgery alone and surgery plus perioperative radiotherapy do not present statistically significant differences, while the morbidity of the 2 combined modalities is higher, due to this and R0 resection.[7] After surgery, our patient will remain under active surveillance without any other treatment modality. The postoperative recurrence rate can be reduced by maintaining the negative surgical margin during the operation, plus radiotherapy or chemotherapy after the operation. The prognosis of patients with TD is notoriously variable.

CONCLUSION

DT is rare, since its diagnosis and treatment requires a multidisciplinary approach. Due to the atypical presentation of wall abscess, the timely diagnosis was made in this case. In Mexico there is no report on randomized controlled studies to assess the best approach, but at a global level they propose that active surveillance is currently the preferred treatment for patients who have TD in non-critical locations, now at least 1 to 2 years of surveillance is recommended. active, this due to its probability of spontaneous regression. There are specific indications for other types of therapies (surgery, medical management, radiotherapy, chemotherapy and combinations of these). But the main objective of all of them is to preserve or improve the patient's quality of life.

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