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DESMOID TUMOR IN A PATIENT WITH FAMILIAR ADENOMATOSIS POLYPOSISIS

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Abstract: Desmoid tumors (TD), also known as aggressive fibromatosis or musculo-aponeuritic fibromatosis, are rare, benign, non-metastatic, locally infiltrative neoplasms that originate from soft tissues or muscular-aponeurotic structures. **Goal:** To report a rare case of a patient with desmoid tumor with a history of familial adenomatosis polyposis (PAF). **Case report:** Male patient, 32 years old, with a family history of FAP, underwent protocolectomy with ileo-rectal reconstruction in 2021. In 2023, the patient noticed the presence of a solid, painless mass on the right flank. Computed tomography of the total abdomen was performed. with contrast, a large heterogeneous mass was revealed, measuring 11.5 x 9.5 x 12.0 cm (W x AP x T), suggestive of Desmoid Tumor. Submitted to tumor resection with rotation of a muscle flap and enterectomy with burial-anastomosis. **Discussion/Final Considerations:** These tumors often occur in the abdominal wall. Imaging exams assist in diagnosis and can determine the approach modality. Surgical resection with a histologically negative margin has been indicated as an approach for this disease in most cases.
Keywords: Case Report; Demoid tumor; Aggressive fibromatosis.

INTRODUCTION

Desmoid tumors (TD), also known as aggressive fibromatosis or musculo-aponeuritic fibromatosis, are rare, benign, non-metastatic, locally infiltrative neoplasms that originate from soft tissues or muscular-aponeurotic structures [4,6].

Although considered benign as it does not have the potential to cause metastases, TD is aggressive, presenting rapid growth locally in most cases. [2,6].

Its occurrence is often associated with congenital alterations of the familial adenomatous polyposis (FAP) type, reaching 1000

times higher than in the general population[2].

The tumor is rarely found, accounting for only 3% of all connective tissue tumors and being seen in only 2 to 4 women per million of the population in the United States[2,4,6].

Desmoid tumors, depending on their location, are classified into three types. The first type is abdominal, which affects the anterior abdominal wall; the second type is intra-abdominal, which affects the mesentery and pelvis in intra- or retroperitoneal situations; the third is extra-abdominal, which affects the chest, extremities, head and neck [4,5].

Ultrasonography, computed axial tomography and magnetic resonance imaging are the main diagnostic tools. But MRI is the most suitable method to define the extent of the lesion and its best approach[1,3,6].

Biopsy is necessary for definitive diagnosis, mainly with immunohistochemical analysis[1,6].

To date, surgery is the gold standard for treating desmoid tumors. Despite being the most used therapy, surgery alone has a high rate of recurrence, which is why extensive resection is indicated to prevent recurrence of the disease[1,2,4,5,6].

The objective of this study is to report a case of Desmoid Tumor in a patient with a history of FAP, successfully treated by surgical resection with flap rotation.

CASE REPORT

Male patient, 32 years old, mixed race, single, professional driver, born in Rio de Janeiro, residing in Campo Grande, with a family history of FAP, sought medical evaluation in May 2021 due to hematochezia accompanied by asthenia. A colonoscopy was performed, revealing the presence of numerous polyps (+ 100) with an adenomatous appearance, distributed throughout all segments of the colon and rectum. Polypectomy was performed on 1 polyp in the lower rectum, in the upper rectum, a vegetating lesion, with an irregular surface, soft consistency, measuring 20mm in diameter. In August 2021, the patient underwent total proctocolectomy with terminal ileostomy. Material sent to the Anatomopathology service, whose histological result revealed that it was a well-differentiated, ulcerated ADC, located in a large Tubulovillous adenoma with High Grade epithelial dysplasia. January 2022, transit reconstruction was performed with L-T ileo-rectal anastomosis.

In August 2023, the patient notices the presence of a mass on the right flank. With this situation, on 11/30/23 he went to the HCP emergency, where he was evaluated by the surgery team. Enhancement physical examination; abdomen: Presence of a solid mass on the right flank, below the ileostomy scar, painless on palpation, irregular surface, not very mobile.

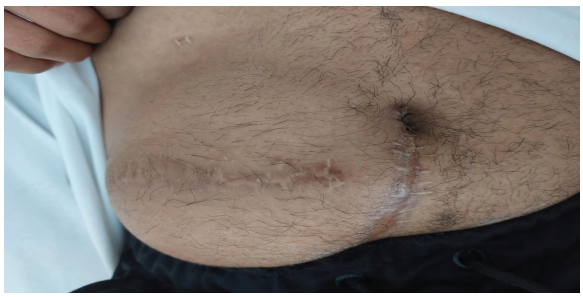

Figure 1: Initial location of the Desmoid Tumor on the anterolateral aspect of the abdomen.

Computed tomography of the total abdomen with contrast was performed, revealing a large heterogeneous mass, with areas of necrosis, with contrast enhancement, located in the right rectus abdominis, with intra-abdominal extension and into the subcutaneous tissue, located in close contact with the surgical threads colonized the right iliac fossa, measuring 11.5 x 9.5 x 12.0 cm (W x AP x T), and the Desmoid Tumor could not be ruled out.

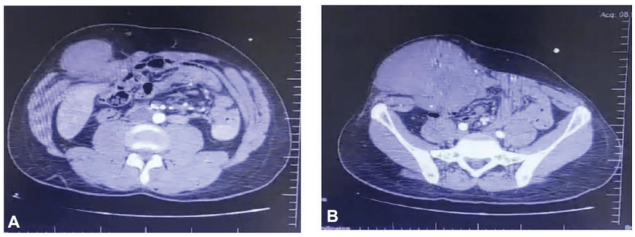

Figure 2. A. Computed tomography of the abdomen showing a desmoid tumor located in the wall. B. Image showing involvement of intra-abdominal organs.

On 12/09/2023, the patient underwent tumor resection with rotation of a muscle flap, plus enterectomy with burial-anastomosis, with placement of a Marlex mesh in the abdominal wall and leaving a hemovac drain.

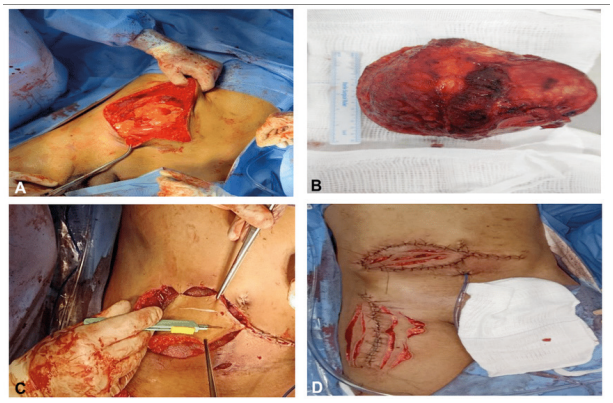

Figure 3. Surgery to resect the desmoid tumor and reconstruct the abdominal wall. A. Open abdominal region after tumor removal. B. Surgical specimen after resection. C. Flap rotation after closure of the abdominal cavity. D. Image after wall closure with flap rotation of the right thigh.

On the 1st postoperative day (POD), the patient presented with abdominal pain and nausea. On general physical examination, the patient was in good general condition, lucid, oriented in time and space, mucous membranes colored and hydrated, apyretic. Flat abdomen, painful on palpation, without signs of peritoneal irritation, no flatus, FO clean, dry, hemovac working, as a conduct he remained on a zero diet.

On the 2nd POD there was an improvement in his previous condition, and he was advised on a liquid diet and ambulation. 4th POD, patient evolves satisfactorily, without complaints, tolerating diet, emitting gases and feces, walking, FO clean, dry, without signs of infection, drain with a small amount of liquid less than 50 ml, serohematic.

As a procedure, the hemovac drain was removed and the patient was discharged from hospital on the 6th postoperative day. Guided outpatient follow-up.

Currently, seven (7) months later, the patient is in good general condition, with no complaints, no signs of tumor recurrence or limitations in his work activities.

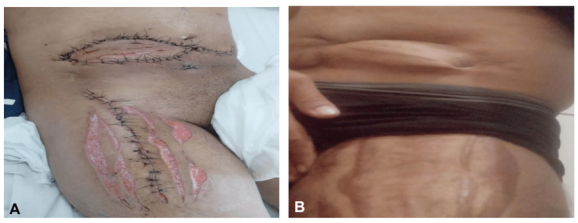

Figure 4. A. Image after the 4th day of the surgical approach. B. Seven (7) months later after surgical resection with muscle flap rotation.

DISCUSSION

Desmoid tumor, also called aggressive fibromatosis or musculoaponeurotic fibromatosis, is a connective tissue neoplasm found mainly in the abdominal region of women who have suffered trauma or who are of reproductive age [4].

Its occurrence is often associated with congenital alterations such as familial adenomatous polyposis. (PAF) [2.] In this report, the patient had a pathological history of FAP.

Depending on their location, they are classified into three types, the first type is abdominal; the second type is intra-abdominal and the third is extra-abdominal [4,5]. In our case, the patient presented abdominal type with intra-abdominal infiltration.

Imaging exams are essential for diagnosis and surgical planning. MRI is the most suitable method to define the extent of the lesion and is the best approach due to its ability to evaluate the structures [1,6].

The treatment of TD must be guided according to its extension, location, infiltration to adjacent tissues and organs [6].

Complete surgical excision is considered the only effective treatment. However, there is a certain probability of recurrence [2,5,6].

The use of chemotherapy, with tamoxifen, antiestrogenic therapy, with progesterone or medroxyprogesterone and also non-steroidal anti-inflammatory drugs, radiotherapy and cytotoxic drugs are described in the literature with varying success rates [6].

FINAL CONSIDERATIONS

Therefore, Desmoid Tumors are rare, benign, aggressive, locally infiltrative neoplasms, with high local recurrence even after adequate treatment, however, without metastatic behavior. These tumors often occur in the abdominal wall. Imaging exams assist in diagnosis and can determine the approach modality. Although there are several therapeutic alternatives, most still do not have scientific proof of their benefits and harms. Surgical resection with a histologically negative margin has been indicated as an approach for this disease in most cases. In this case report, surgical resection with free margins was the basic treatment for this disease. The patient will be monitored on an outpatient basis in order to diagnose signs suggestive of recurrence.

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