

## Case Report

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# Giant desmoid tumor of the abdominal wall in a patient with Gardner Syndrome

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**ABSTRACT:** Gardner syndrome (GS) is a rare entity characterized by a triad of familial colonic polyposis, multiple osteomas and soft tissue tumors, including desmoid tumor (DT). This is a case report of a 30 year-old patient with GS who developed giant DT in the abdominal wall after undergoing several laparotomies. The patient has taken a long time to search for medical care, and at first he saw another team that refused to operate him by judging the lesion unresectable. The surgery in our department was performed in three steps. Initially, we resected the lesion with macroscopic margins, and as there were small bowel adhesions in the tumor, we performed enterectomy and closed using the “Bogotá” technique, with skin closure on the bag. On the fourth postoperative day (POD), we reoperated the abdomen without identifying any signs of fistula. On the seventh POD there was another surgical intervention, this time to insert a double-sided mesh. The patient recovered well, and had no debilitating motor deficit, despite the extensive resection of the abdominal muscles. Curative treatment of DT is based on surgical resection and only sequential surveillance allows us an early diagnosis, when the lesion is still resectable.

**Keywords:** intestinal polyposis; colorectal surgery; Gardner syndrome.

**RESUMO:** Tumor desmoide gigante de parede abdominal em paciente portador da Síndrome de Gardner. A Síndrome de Gardner (SG) é uma entidade rara caracterizada pela tríade polipose colônica familiar, múltiplos osteomas e tumores de tecidos moles, dentre eles o tumor desmoide (TD). Tratou-se de um relato de caso de um paciente de 30 anos, com SG que evoluiu com TD gigante em parede abdominal, após ser submetido a diversas laparotomias prévias. O paciente levou longo tempo para procurar o serviço de cirurgia, passando por outra equipe que se negou a abordá-lo por julgar a lesão irressecável. A cirurgia no nosso serviço se deu em três tempos. Inicialmente, foi feita a ressecção da lesão com margens macroscópicas e, por haver aderências de alças no tumor, realizamos enterectomia e fechamos a Bogotá com síntese da pele sobre a bolsa. No quarto dia pós-operatório (DPO), reabordamos o abdômen sem identificar sinal de fistula. No sétimo DPO houve nova abordagem, agora para colocar tela dupla face. O paciente evoluiu bem, sem déficit motor debilitante, apesar da extensa área de ressecção muscular abdominal. O tratamento curativo dos TD é baseado na sua ressecção cirúrgica e somente a vigilância sequencial nos permite seu diagnóstico precoce e a abordagem enquanto a lesão é ressecável.

**Palavras-chave:** polipose intestinal; cirurgia colorretal; síndrome de Gardner.

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## INTRODUCTION

Gardner Syndrome (GS) is a variant of familial adenomatous polyposis (FAP), characterized by the triad: gastrointestinal polyposis, multiple osteomas and soft tissue tumors, including the desmoid tumor (DT), which occurs in approximately 13% of the cases. DT is a myofibroblastic benign neoplasm originated from the muscle aponeurotic tissue, belonging to the group of fibromatosis. Its most common sites are intestinal mesentery, aponeuroses of the rectus abdominis muscle and extremities, related to areas with scars or previous trauma. They are more frequent among females, especially between the second and fourth decades of life. Most cases are histologically well differentiated, with no metastases. However, it can develop local invasive behavior, with aggressive growth and high rates of recurrence, especially in sites of difficult surgical resection with free margins, which many times leads to death<sup>1-3</sup>.

## CASE REPORT

A 30 year-old male patient diagnosed with GS at the age of 27. The patient has six siblings, out of which 4 were also diagnosed with FAP. The first case was identified in one of the siblings, presenting with acute abdominal obstruction due to colonic neoplasm. The other relatives were investigated, diagnosing a total of five cases of FAP among the seven siblings. The patient in this case report was submitted to prophylactic elective total proctocolectomy by videolaparoscopy with creation of an ileal pouch.

He underwent exploratory laparotomy in the first 48 postoperative hours due to abdominal bleeding. Afterwards, a diverted ileostomy was closed and the patient developed a fistula in the ileal pouch later, so more surgical interventions were necessary, completing a total of four laparotomies. At the occasion, one of the siblings diagnosed with GS presented with advanced DT and died after the surgical approach of the recurrent lesion. The patient abandoned medical follow-up and developed a mass in the abdominal wall; however, afraid of having the same problem as the late brother, he hesitated to look for the medical service, and the lesion became a giant DT in the abdominal wall. He lost his follow-up and the Brazilian Unified Health System

(SUS) referred him to another medical service, where the lesion was considered unresectable. Then, he came back to the first service where he was reassessed two years later, and physical examination showed a giant tumor on the anterior abdominal wall, extending from the right breast region to the left iliac fossa, involving practically the whole right abdominal wall and part of the left side (Figure 1). Staging did not show distant lesions and computed tomography (CT) of the abdomen revealed a local advanced lesion confined to the abdominal wall, with 30x15x13 cm, with no signs of other structure invasion. At first, we proposed neoadjuvant therapy with tamoxifen for 2 months; however, the response was unsatisfactory, and the lesion progressed. Then, the surgical approach was defined. Surgical indication was challenging, since the area of the abdominal wall affected with the tumor was large, and the resection would cause a great ventral defect. So, the reconstruction of the abdominal wall using a prosthetic mesh was programmed.



Figure 1. Preoperative.

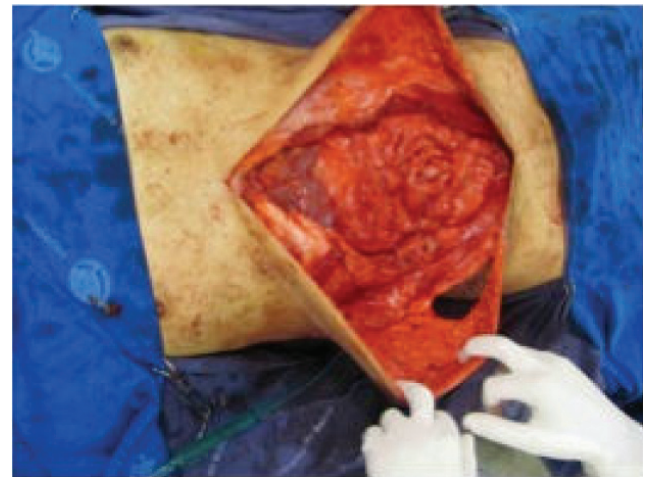
The surgery was performed by a multidisciplinary team involving the areas of coloproctology, oncologic and plastic surgery. It was divided into three parts. Initially, a broad resection was performed, removing almost the whole right anterolateral muscles and parte of the left rectus abdominis muscle with free macroscopic margins (Figure 2). There was invasion of the small intestine by the tumor, which demanded segmental enterectomy and mechanical intestinal anastomosis. The choice was to not immediately reconstruct the abdominal wall with prosthesis due to the condition of potentially contaminated surgery. The patient was kept in laparostomy with skin closure over a plastic bag to avoid adherences. On the fourth postoperative day (POD), we reviewed the cavity,

and there was no evidence of anastomotic fistula. On the seventh POD, a new approach was performed to reconstruct the abdominal wall using a double sided prosthesis (polytetrafluorethylen – PTFE – and polypropylene) fixated to the left residual aponeurosis, right lateral muscle and right rib cage, fulfilling the great ventral defect. Anatomopathological examination showed a 3,375 g lesion measuring 33x17x15 cm, with small intestine segment and microscopy of desmoid fibromatosis.

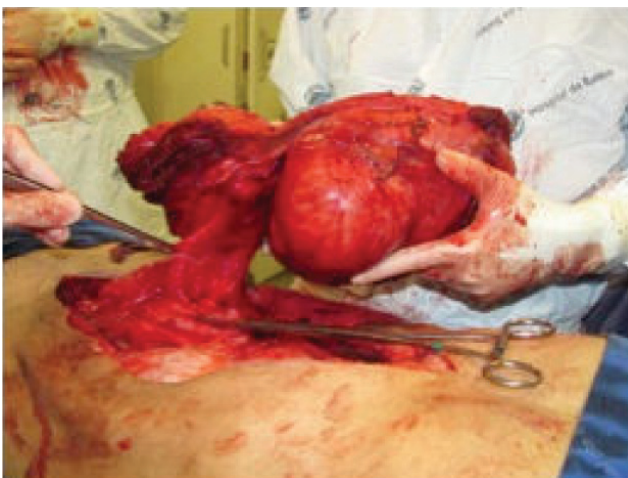
The patient improved without postoperative complications and no debilitating motor deficit, despite the extensive muscle resection. He has now been on outpatient follow-up for two years, with no complaints, and imaging tests show no signs of recurrence (Figure 3).



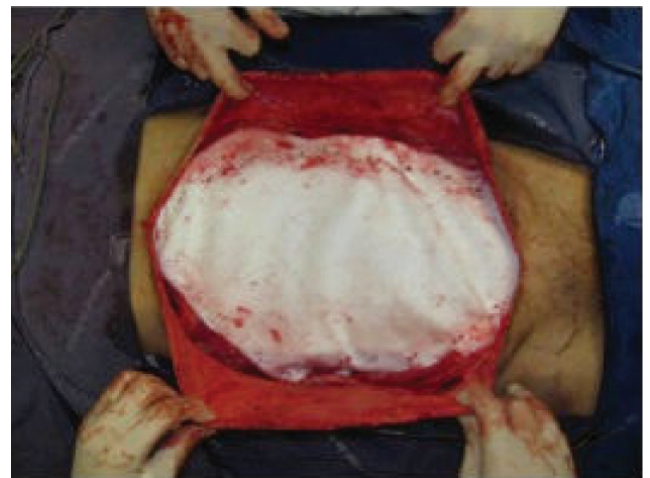
(A) Resection



(C) Area to be reconstructed



(B) small bowel adherence



(D) double-sided prosthesis

**Figure 2.** Perioperative and reconstruction of the seventh postoperative day.



Figure 3. Third postoperative month.

## DISCUSSION

DT is rare, affecting approximately 3.7 people out of 1 million subjects a year. Patients with FAP have one thousand times more chances to have this type of tumor than the general population, and it is one of the main causes of mortality in this group.

DT diagnosis is performed by imaging tests, and CT and nuclear magnetic resonance are useful to assess the characteristics of the lesion, such as size, extension and relation with adjacent structures, thus identifying possible invasion<sup>4-6</sup>. In the reported case, abdominal CT showed a giant lesion on the abdominal wall, with 30x15x13 cm, confined to the abdominal wall, with no signs of invasion of intra-abdominal structures. However, the involvement of intestinal loops was identified intraoperatively. The correlation of radiological findings with clinical and epidemiological data, such as age, previous surgery and FAP, turns DT into an important diagnostic hypothesis. However, the definitive diagnosis is based on the histopathological analysis of the lesion by incisional biopsy or from surgical specimen. The patient in this case report was directly referred to surgical resection due to the great probability of DT.

The definitive treatment of DT is based on broad surgical resection with free margins. Radical approach may lead to anatomical defects, especially when located in the abdominal wall, which requires correction with myocutaneous flaps. In cases of major defects, the use of prosthetic mesh is common<sup>7-9</sup>. For the patient operated in our service, we chose to reconstruct

with a double-sided prosthesis made of synthetic material (PTFE and polypropylene), once the extensive ventral defect left no alternative to repair the abdominal wall.

Three major reference cancer centers have performed an extensive review of their DT cases and retrospectively analyzed the possible prognostic factors related to local recurrence, coming to controversial conclusions. The MD Anderson Cancer Center (Texas, USA) analyzed 189 cases of extra-abdominal DT and found the mean size of tumors was 7 cm. Most of them were treated with isolated surgery, and recurrence rates ranged from 20 to 30%. The authors did not find statistical association between macroscopically compromised surgical margins and the increased local recurrence. The same result was found in a series by the Sloan-Kettering Cancer Center (New York, USA) and Istituto Nazionale Tumori (Milan, Italy), with 105 and 203 cases, respectively. However, the series from Milan showed that compromised margins were an important prognostic factor in patients submitted to a new resection of recurrent lesions. The standard treatment for DT is still surgical resection with free margins. Radiotherapy has a questionable role to treat DT, but it is indicated in cases of compromised margins. It can be used as neoadjuvant therapy in cases of lesions that are considered unresectable, or those with difficult resection. Due to the poorly consistent data on its efficacy, there are no standard strategies of treatment. Some authors still defend hormonal therapies for unresectable DT or those with no response to radiotherapy. The patient in this report used tamoxifen as a neoadjuvant therapy; however, there was no satisfactory response. The histopathological analysis of this patient's lesion revealed a mass of major proportions, weighing 3,375 g and measuring 33x17x15 cm, which is much higher than the average of reports found in literature. Microscopy confirmed desmoid fibromatosis. Regarding adjuvant treatment, it was chosen not to indicate radiotherapy due to the great ventral area occupied by the mass, which would cause irradiation of practically the whole abdominal cavity.

## CONCLUSION

DTs are challenging, therefore, it is important to screen patients who have GS, because the early diagnosis and aggressive surgical treatment with complete resection and free margins are the main curative factors, with low recurrence rates.

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