Case Report

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# Two Cases of Initially Unresectable Desmoid Tumors

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## Abstract

Desmoid tumor, also called aggressive fibromatosis, is a rare fibroblastic proliferation of connective tissue and skeletal muscle aponeurosis. The aetiology of desmoid tumours is poorly understood, but they have been related with oestrogen stimuli, previous trauma, surgical interventions and pregnancy. Although it is characterized by not having potential for metastasize or differentiate, it is a non-encapsulated lesion, with an infiltrative growth and unpredictable behaviour. In this way, it could have either an aggressive pattern, when compared with other low degree malignancy sarcomas, or a very indolent one, even with spontaneous regression. Therapeutic options in desmoid have been changing over these last years, as surgery upfront in asymptomatic patients has been superseded by active surveillance. However, treatment has to be individualized in some specific scenarios such as intra-abdominal lesions. We present two cases of patients diagnosed with an intra-abdominal desmoid tumour with threatening behaviour, the treatment approach and final resolution.

**Keywords:** Desmoid, Surgery, Oncology, Tumour

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## Introduction

Desmoid tumor, also called aggressive fibromatosis, is a rare fibroblastic proliferation of connective tissue and skeletal muscle aponeurosis. This entity represents less than 3% of all soft tissue tumors in the general population, with a higher incidence in patients diagnosed with familial adenomatous polyposis (FAP). The etiology of desmoid tumors is poorly understood, but they have been related with estrogen stimuli, previous trauma, surgical interventions and pregnancy (1). Even though, this neoplasm can appear at any age, the highest incidence peak is in the third decade, with a clear preponderance in females (5:1); the abdomen and extremities are the most frequent sites of involvement. Magnetic resonance imaging (MRI) is the gold standard image test to characterize this lesion, and it can be used for diagnosis, local staging and follow-up. Nevertheless, histopathologic confirmation is mandatory before initiating treatment.

Although it is characterized by not having the potential for metastasis or differentiation, it is a

non-encapsulated lesion with infiltrative growth and unpredictable behavior. In this way, it could have either an aggressive pattern, when compared with other low degree malignancy sarcomas, or a very indolent one, even with spontaneous regression. This unforeseeable natural history could lead to a high symptom burden and severe physical impairment.

Therapeutic options in desmoid tumors have been changing over these last years, as upfront surgery in asymptomatic patients has been superseded by active surveillance. However, treatment has to be individualized in some specific scenarios such as intra-abdominal lesions, where surgery remains the standard of treatment, if feasible (2). Systemic treatment, like anti-hormonal therapy, non-steroidal anti-inflammatory drugs (NSAIDs), tyrosine kinase inhibitors, and chemotherapy, is an option in symptomatic patients (3, 4). Considering the variable clinical presentation and biologic behavior, it is highly recommended to personalize the management of patients within the context of an expert multidisciplinary team (5).

We present two cases of patients diagnosed with an intra-abdominal desmoid tumor with threatening behavior. The treatment approach and outcome for each case is also discussed.

## Objective

To report the diagnosis and management of two patients diagnosed with intra-abdominal desmoid tumors.

### *Case 1*

A 53-year-old female presented with a four-week history of abdominal pain related with symptoms of high intestinal sub-occlusion. After several emergency room visits, an abdominal CT-scan was performed, reporting a mass of 194×111×85 mm originating in the third duodenal portion. The lesion infiltrated the proximal jejunum, conditioned duodenal dilatation and showed a radiologic aggressive behavior (Figure 1). Initial biopsy was rejected given the risk of intestinal injury.

An exploratory laparotomy was performed on August 2017, evidencing a huge mass located in the angle of Treitz, invading the first jejunal loop and the superior mesenteric vessels. Due to the vascular involvement, it was initially considered as an unresectable lesion, and biopsies were obtained.

In order to guarantee the patient's nutrition, a gastrojejunostomy was attempted, but the retraction of the meso conditioned a jejunal blockage not allowing the bowel ascension to ensure a safe and functional anastomosis. Finally, a descompressive gastrostomy and a jejunostomy were performed.

The pathology reported a desmoid tumor and, due to the impossibility of surgery, evaluation in a multidisciplinary team was required. Considering its destructive local behavior, fast growth and life-threatening nature, neo-adjuvant chemotherapy was decided. A fibrocolonoscopy and fibrogastroscopy were also requested in order to exclude a FAP-associated tumor.

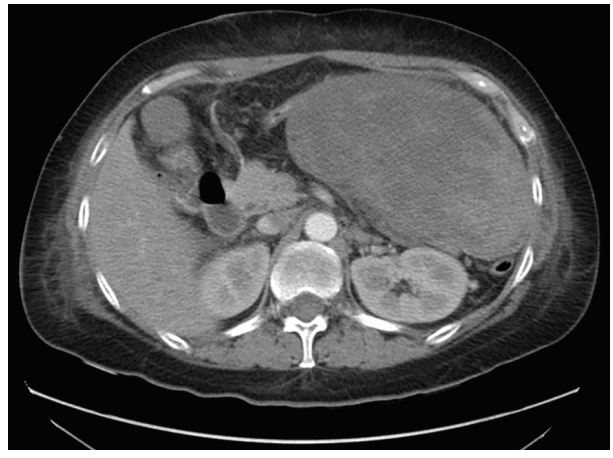

**Figure 1:** Abdominal CT scan reporting a mass with radiologic aggressive behavior

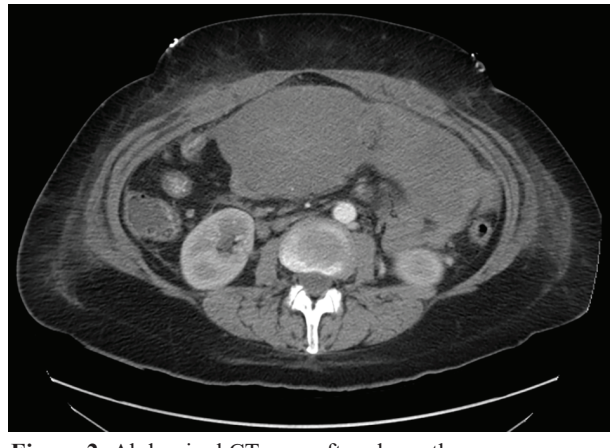

**Figure 2:** Abdominal CT scan after chemotherapy

Chemotherapy treatment with Adriamycin 75 mg/m2 was started on September 2017.

After 3 cycles, another abdominal CT-scan was performed, bringing to light a reduction in the volume of the intra-abdominal mass, with a bi-lobed shape featuring axes of 60×108 mm and 70×110 mm (Figure 2). The patient also presented an improvement in her clinical status, restarting oral diet without nausea or vomiting. Chemotherapy was continued for up to 6 cycles.

Another abdominal CT-scan was carried out after chemotherapy, confirming the shrinkage of the lesion volume, with a tumor size of 76×44×59 mm (LL×AP×CC) and a lateral portion of 57×63×67 mm. A cleavage plane was also noticed between the superior mesenteric artery and vein, as well as its principal branches and the tumor.

Due to the partial response to neo-adjuvant chemotherapy and, more importantly, due to the improvement in the vascular involvement, a second surgery was attempted. Through an exploratory laparotomy, the mesenterium mass was noticed, involving the first jejunal loop. The resection of the tumor was performed and, with the aim of reconstructing the intestinal transit, an isoperistaltic

manual latero–lateral duodenojejunostomy was completed.

After an adequate postoperative course and tolerance of oral nutrition without any issues, the patient was discharged on postoperative day (POD) 14 after a correct intestinal transit test.

She is now on follow- up, without signs of recurrence and good quality of life.

### *Case 2*

A 52-year-old man presented with a 2-week history of abdominal pain and temperature. The very first test was an abdominal ultrasonography, performed on October 2018; a large, poorly delimitated solid mass of 153×110 mm was reported anterior to the iliac vessels in the mesogastrium.

The study was completed with an abdominal CT-scan, reporting an intra-abdominal mass fistulized to the ascending colon, causing a right pielic dilatation (Figure 3). The suggested diagnostic options were GIST, sarcoma or, less likely, lymphoma.

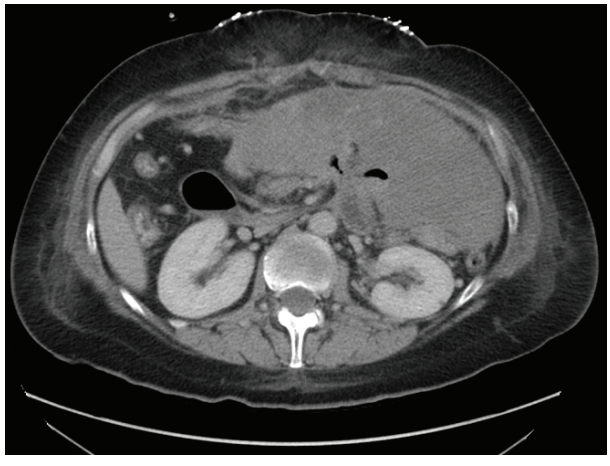

**Figure 3:** Abdominal CT scan showing a fistulized mass

After an accurate assessment in a multidisciplinary board, due to the fistula and the patient instability, biopsy and subsequent surgery were indicated. The pathology report was negative for malignancy, so a laparotomy was performed, showing a large abscessed mass fistulized to the right colon. A right hemicolectomy was accomplished.

As an immediate postoperative complication, the patient presented with paralytic ileus, requiring a nasogastric tube and total parenteral nutrition. He also suffered acute obstructive renal failure with oliguria and elevated creatinine levels, solved by conservative treatment with intensive fluid therapy.

The final pathological report was consistent with intra-abdominal desmoid fibromatosis (desmoid tumor), with a tumor size of 23 cm and clear margins.

After the resolution of those postoperative complications and tolerance to oral nutrition without hassle, the patient was discharged on POD 14.

## Discussion

Desmoid tumor, also called aggressive fibromatosis, is a rare condition, representing less than 3% of all soft tissues tumors (4). Its origin begins in the aponeurotic muscle, and it can be sporadic or developed within the context of FAP, an inherited disease due to mutations in the APC gene (6). Despite the fact that the etiology remains uncertain, the growth of these tumors has been linked to surgery, previous trauma, and estrogen stimuli (related to pregnancy or estrogenic therapy).

Even though these lesions are benign with no ability to metastasize, they tend to invade and recur locally.

Clinical manifestations at the time of diagnosis depend mainly on the location of the tumor. In the abdominal area, pain and increasing abdominal perimeter are frequent. Toxic syndrome, intestinal occlusion and hydronephrosis can also occur. Other sites where they can develop are the extremities and trunk (7).

Imaging techniques, such as ultrasonography, CT-scan and MRI, provide information about the size, extension and relationship of the tumor with nearby anatomical structures. A biopsy is required to obtain the definitive diagnosis through a histological confirmation and concordant immunohistochemistry. The principal differential diagnosis should be done with other soft tissue tumors such as sarcomas.

As previously said, due to the heterogeneity of the biological behavior of desmoid tumors, including long periods of stable disease or even spontaneous regression, treatment needs to be individualized to optimize local tumor control and preserve patient quality of life. Hence, the application of a multimodality treatment, decided in a multidisciplinary assessment, sets the basis of care for these patients.

For decades, surgery has been the standard of care when treating desmoid tumors. Nonetheless, according to the experts, immediate surgery is no longer the right front-line approach. Retrospective series have shown progression-free survival rates of 50% at 5 years for asymptomatic patients managed with a "watch and wait" approach (8, 9). Nowadays, the conservative watch and wait strategy should be the first approach for newly diagnosed patients, with the aim of understanding the behavior of the disease and conceiving further treatment steps. The patients have to be closely followed, with the first clinical and/or radiological re-evaluation within 8-12 weeks, preferably with an enhanced MRI.

In case of tumor growth or symptoms, surgery is still a correct option (IV, A). When carried out, the aim of surgical resection should be to obtain microscopic negative margins, but always considering functional preservation, especially for tumors located in the limbs.

In the abdominal area, including abdominal wall, mesenterium and retroperitoneum, surgery can be performed when the initial "watch and wait" approach has not succeeded. In this scenario, neo-adjuvant systemic treatment could be appropriate and beneficial.

In some specific clinical situations, like occlusion or perforation, urgent surgery is mandatory, taking into account the patient's morbimortality (10).

Both cases presented above required an early surgical approach given the symptoms caused by the tumor and the patient instability. However, they are also a good example of the value of a diversified opinion through the consensus of surgeons, oncologists, radiologists and radiation oncologists.

Given the rarity of this disease, it is important to lead prospective trials and correlative laboratory tests within the context of a multicenter cooperation. Such efforts would attempt to identify clinical or molecular criteria by which patients could be better selected for different approaches. In this way, systemic therapy, long-term observation, or intense multimodality treatment could be selected with the vision of keeping patients with desmoid tumors alive and preserving their quality of life.

In summary, it is clear that in such an unusual tumor, a multidisciplinary discussion in a soft tissue tumor board is crucial to suit the best therapeutic option for individual management, with an interview with the patient also being fundamental. Defining some molecular characteristics would help to determine its present unforeseen behavior.

**Conflict of Interests:** None declared.

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