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A Progressive Desmoid Tumor of the Shoulder Girdle: Case Report and Literature Review

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Abstract

Background: Desmoid tumor (DT) is a rare locally aggressive tumor of unpredictable evolution that develops in deep soft tissues with an infiltrative growth pattern and with a high tendency to local recurrence. Only 17% of extra-abdominal DT develops in the shoulder girdle. **Aim:** Describe the behavior and unpredictable evolution of DT as well as the different treatment options currently recommended, along with their advantages and disadvantages. **Case Presentation:** A 29-year-old female with a DT in the shoulder girdle of 6 years of evolution with progressive growth, pain, and limitation in the joint movements treated with chemotherapy presenting stable disease and switching to Tyrosine Kinase Inhibitors (TKI) with progressive disease for which a R0 resection was performed. Currently, under surveillance with a 3-year disease-free survival and a control computed tomography (CT) scan with no data of tumor activity. **Conclusion:** DT treatment should be multidisciplinary and individualized based on the behavior, location, and size of the tumor, the functional status of the limb, secondary symptoms, the risk of complications if the disease continues to progress, and the patient's decision.

Keywords

Desmoid Tumor, Desmoid-Type Fibromatosis, Aggressive Fibromatosis, Deep Fibromatosis, Musculoaponeurotic Fibromatosis, Shoulder Girdle

1. Introduction

A desmoid tumor (DT) (desmoid-type fibromatosis, aggressive fibromatosis, deep fibromatosis, musculoaponeurotic fibromatosis) is a rare locally aggressive tumor of unpredictable evolution that is histologically characterized by a monoclonal proliferation of fibroblasts and develops in deep soft tissues with an infiltrative growth pattern and with a high tendency to local recurrence (25% - 77%) [1]-[6]; although it does not have the capacity to metastasize [5], it can be multifocal in the same extremity or in the affected body site [2] [3]. It currently constitutes < 3% of soft tissue neoplasms [3] [7], has an incidence of 5 to 6 cases per million people per year [1] [5], predominates between 30 and 40 years of age, and is more frequent in women (up to 70% of cases) [1]-[4] [7]-[9].

Although it can occur in any part of the body, DT predominates in the extremities (especially when it is sporadic), abdominal wall, and mesentery [3] [4], and based on its location, it is classified as extra-abdominal (60%), abdominal wall (25%) and intra-abdominal (8% - 15%) [6]. Only 17% of extra-abdominal DTs develop in the shoulder girdle [10]-[13], and unfortunately, it is common for these patients to present for medical evaluation after a large tumor growth, with persistent pain and/or limited joint mobility [13]. Through this case, in which our patient's disease evolved through different treatment modalities, we will describe the current recommendations for the management of DT, including active surveillance, pharmacological therapies, surgery, and radiotherapy, along with their advantages and disadvantages.

2. Case Presentation

A 29-year-old female with a Body Mass Index (BMI) of 34.9 with no relevant pathologic history and a soft tissue tumor in the left scapular region of progressive growth of 6 years of evolution. In the initial consultation, she reported not having previously sought medical attention because the tumor growth was slow and asymptomatic; however, in the last 8 months, the growth rate had increased and began with intermittent pain at the tumor site, which radiated to the arm, and with limitation in the movements of the shoulder joint; physical examination revealed an indurated tumor in soft tissues of the left scapular region partially mobile with extension to the posterior region of the deltoid muscle up to the level of the posterior axillary line and to the posterior third of the arm limiting joint movements.

CT scan (**Figure 1**) revealed a tumor at the left shoulder girdle extending to the posterior scapular region, with irregular morphology and borders, solid and hypodense, and infiltrating the teres minor, infraspinatus and deltoid muscles towards their posterior border, without metastatic lesions. The core biopsy reported a tumor formed by long fascicles of spindle cells between a collagenous stroma without pleomorphism or the presence of mitosis or atypia, and by immunohistochemistry, the expression of actin, vimentin, and beta-catenin was detected (**Figures 2(A)-(D)**), findings compatible with a DT.

Due to the time of evolution, the growth rate, and the associated symptomatol-

ogy, such as joint limitation, in a multidisciplinary session, it was decided to initiate active treatment because although the lesion was resectable, achieving an R0 resection because it did not involve the axillary vessels or the brachial plexus, the morbidity and sequelae of the procedure were considerable. The patient started receiving 6 cycles of liposomal doxorubicin monodrug scheme, presenting symptomatic improvement with pain reduction; however, she did not present improvement in joint limitation, and in control CT scan, she presented stable disease.

Given the stable behavior of the disease, we decided to progress the treatment line with sorafenib at a dose of 400 mg every 12 hours with partial tolerance requiring dose adjustment to 600 mg per day at the 4˚ month due to gastrointestinal toxicity presenting clinically stable disease as well as by CT scan in her two subsequent evaluations at 6 and 9 months respectively after starting the TKI. Subsequently, after completing 12 months of the second treatment, the patient reported an increase in tumor size and symptoms, corroborated by a CT scan of an increasing tumor extension to the axillary and deltoid region, infiltrating the subscapular muscle, the coracobrachialis muscle, the triceps brachii muscle, and the subcutaneous cellular tissue with adjacent skin in the posterior region, and an intimate contact with the neurovascular bundle in the axillar region as well as data of bone erosion at the level of the subscapular fossa (**Figures 3(A)-(C)**).

In this scenario of progressive disease, in multidisciplinary sessions and in consultation with the patient, based on the persistence of pain and functional limitation of the limb, the tumor growth, the proximity of the tumor to the neurovascular structures that were still respected, the risk of further progression, and by the patient's wish (due to the general affection in her quality of life and lifestyle) it was decided to continue with surgical treatment because although the functional result would be limited, R0 resection was still feasible. We do not consider radiotherapy as the next therapeutic option because of the possibilities of an uncertain and/or slow response and secondary tissue damage that would make it difficult to close the defect secondary to wide resection and coverage of the neurovascular structures of the extremity in case of a low tumor response.

The patient presented a tumor of approximately 25 cm in diameter, indurated, over the entire scapular region with an extension behind the arm up to the anterior axillary line (surrounding almost 180˚ the diameter of the arm) involving the proximal third of the triceps, the entire deltoid and subscapular region (**Figure 4**). Clinically, there was no evidence of major vascular or brachial plexus involvement; however, the patient reported isolated episodes of paresthesia in the elbow with irradiation to the forearm and a slight decrease in muscle strength in the forearm and hand.

Surgery began with the identification and dissection of the axillary vessels and the structures of the brachial plexus and later the complete resection of the tumor that involved the entire deltoid muscle, a portion of the proximal third of the triceps, and most of the supraspinatus, infraspinatus, subscapularis and teres minor muscles (**Figure 5(A)**, **Figure 5(B)**); although it did not present bone invasion, resection of the lower third of the scapula was necessary since the tumor surrounded it in more than 270˚ due to its subscapular extension. The secondary defect

was repaired by the closure of muscle planes and fasciocutaneous flaps, placing a mesh in the periphery of the humeral head. The histopathological report concluded a DT of 22 × 16 × 14 cm with negative surgical margins, and the postoperative evolution was favorable with elbow and hand functions without motor or sensory deficits.

After a period of rehabilitation sessions, the patient was able to maintain shoulder posture, and the limited joint mobility was compensated with intact forearm and hand functions, in addition to a great improvement in her quality of life for no longer presenting pain secondary to the tumor or the surgical procedure, improving their ability to carry out their daily physical activities. She is currently under surveillance with a 3-year disease-free survival and a control CT scan with no data of tumor activity (**Figures 6(A)-(C)**).

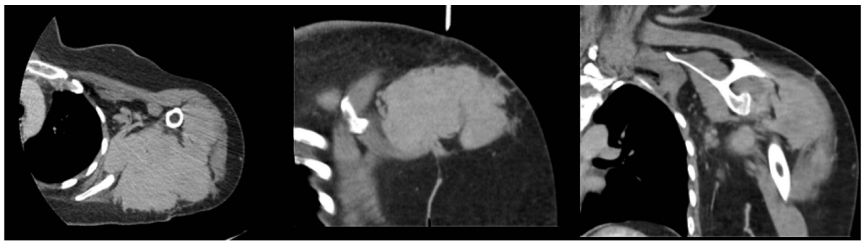

Figure 1. Tumor at the left shoulder girdle, which is of irregular morphology and borders, solid, hypodense, which infiltrates the teres minor, infraspinatus, and deltoid muscles towards its posterior border.

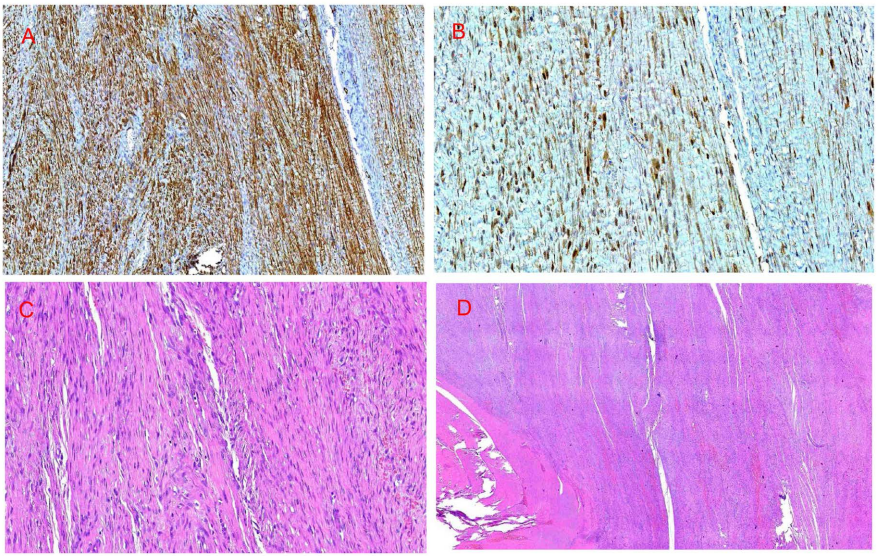

Figure 2. **A.** Immunoreactivity for smooth muscle actin is identified in the cytoplasm of spindle cells, highlighting irregular bundle formations. **B.** Immunoreactivity for β-catenin, nuclear positivity is seen mainly in spindle cells (10%) with positive control in endothelial cells. **C.** Photomicrography at medium magnification of soft tissue lesion showing irregular bundles of spindle cells with tapered nuclei, eosinophilic cytoplasm, and poorly limited borders arranged in divergent directions. **D.** Panoramic photomicrograph of soft tissue lesion showing densely cellular areas arranged in poorly limited lobules between hyalinized areas.

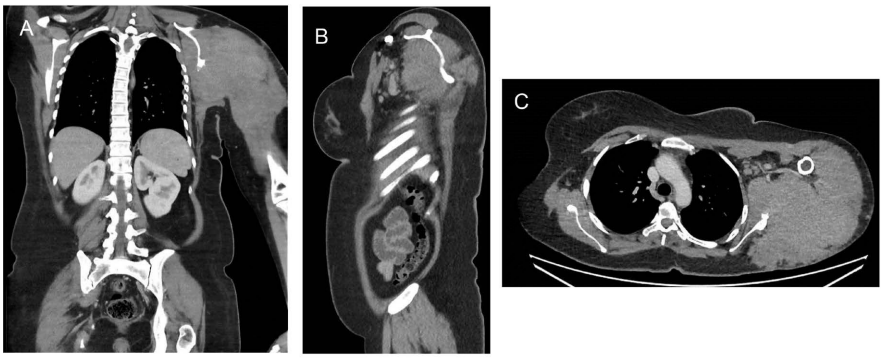

Figure 3. **A-B.** Increased tumor size as well as inferior and medial extension infiltrating the subscapularis, coracobrachialis, and triceps brachii muscles. **C.** Disease progression with greater extension and infiltration of the muscle groups and infiltration of the subcutaneous cellular tissue and skin of the posterior region with intimate contact with the neurovascular axillar bundle.

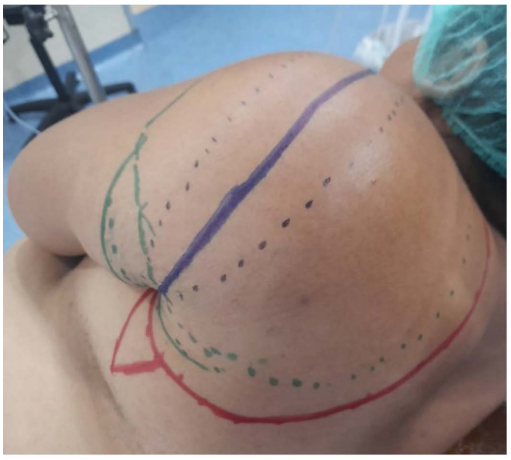

Figure 4. DT of 25 cm in diameter over the entire scapular region with an extension behind the arm up to the anterior axillary line (surrounding almost 180˚ the diameter of the arm) involving the proximal third of the triceps, the entire deltoid and subscapular region.

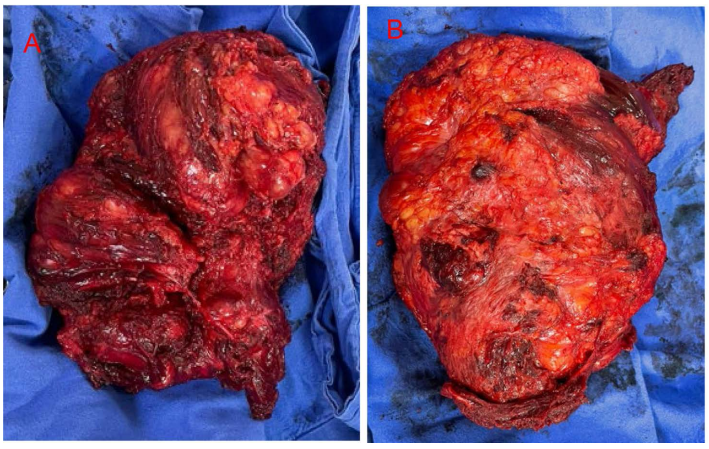

Figure 5. DT of 25 cm in diameter over the entire scapular region with an extension behind the arm up to the anterior axillary line (surrounding almost 180˚ the diameter of the arm) involving the proximal third of the triceps, the entire deltoid and subscapular region.

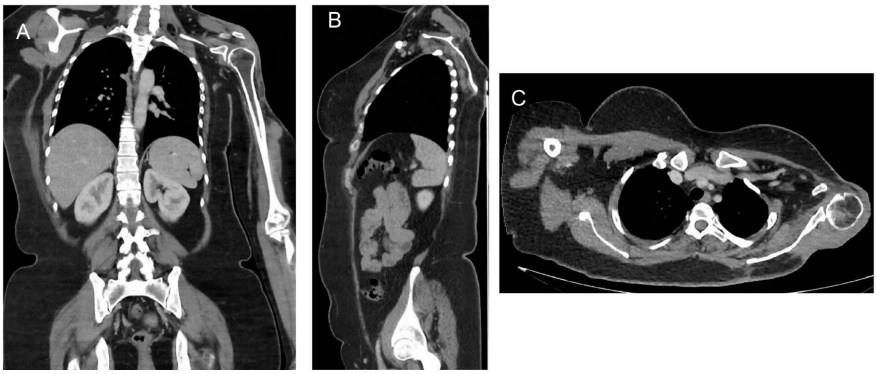

Figure 6. A-C. CT scan control with a disease-free survival of 3 years.

3. Discussion

Although DT is not a tumor that usually compromises life, its growth can progressively limit the quality of life due to chronic pain, psychological alterations, and functional deficits [1]; it can also infiltrate adjacent organs and muscles, compress neurovascular structures, erode bones and cause intestinal occlusions [3]. Our patient presented a progressive decrease in her ability to perform daily physical activities due to both pain and joint limitation, resulting in weight gain and a mixed anxiety-depressive disorder (MADD) exacerbated by the physical appearance secondary to the tumor, lack of response to medical treatment, and tumor progression.

5% to 10% of all DTs are associated with familial adenomatous polyposis (FAP) and Gardner's syndrome [1] [8] [12], and 10 to 15% of patients with FAP develop DT, with intra-abdominal localization being the most frequent in them [1] [3] [8]; these patients present an increased risk of development 1000 times higher than that of the general population [2]-[4] [7], and in them DT tend to present a larger size, multifocality, a more aggressive behavior, and develop at younger ages, up to 10 years earlier than the average age compared to sporadic [4] [7].

A history of trauma (including surgical interventions), estrogens, and pregnancy are considered risk factors for the development of DT [12] [14] [15]. A history of previous trauma has been reported in up to 25% of soft tissue DT cases, and 68% - 86% of intra-abdominal and abdominal wall DT cases are detected after surgery [14]. Because DT is more frequent in women, estrogens could influence tumor growth and spontaneous regression [14]. This fact could be supported by the following data: 11% to 32% of DT in women occur during pregnancy [9], the rate of development of DT increases during pregnancy, and in women on oral contraceptive treatment, the rate of tumor growth is higher in women of childbearing age than in men and in postmenopausal women, and the risk of recurrence is higher in women than in men [14]. Finally, there could be an association between hormonal changes developed during gestation and minor trauma to the muscles and fasciae of the abdominal wall secondary to the elongation of their fibers during uterine growth [9]. Although our patient had no history of trauma or recent pregnancies, her overweight and increased weight gain secondary to physical in-

activity and anxious-depressive disorder could have contributed to the development and progression of DT by a secondary hyperestrogenic activity.

85% - 90% of sporadic DT present somatic mutations in the CTNNB1 gene coding for the β-catenin protein [2] [3], whereas in patients with FAP, DT develops due to germline mutations in the APC gene coding for a protein regulating β-catenin levels. Both mutations affect the Wnt/β-catenin pathway, causing excessive nuclear accumulation of β-catenin that stimulates the activation of genes such as cyclin-D and c-myc, thus triggering a cascade of events that culminates in excessive cell proliferation and differentiation developing DT [1] [2] [4] [7]. These 2 mutations are mutually exclusive [3]-[5]; the finding of the CTNNB1 mutation rules out the APC mutation and vice versa; therefore, the wild-type status of CTNNB1 should lead to the suspicion of FAP [1] [2] [5].

The presenting symptoms depend on the location of the DT [3]; in extremities and abdominal wall, it generally presents as a palpable tumor that can be accompanied by pain and limitation of mobility, especially if it develops in sites adjacent to neurovascular structures [3] [4]; pain tends to be more frequent and progressive in neck and shoulder DTs like in our patient, it is less in abdominal wall DTs, and its presence at the time of diagnosis can be associated with a poorer prognosis and lower response rates to treatment [14]. The pain had been present in our patient since her first consultation, and although she had periods of remission, these were only partial since the pain continued to progress in intensity simultaneously with tumor growth; this symptom was one of the main reasons why the patient opted for surgical treatment. Intra-abdominal DTs can cause weight loss and compressive symptoms such as constipation, early satiety, hydronephrosis, and, in rare cases, intestinal perforations [3] [4]. 50% to 60% of DT show no further growth after diagnosis [1] [2], 20% to 30% may decrease in volume and even disappear even after initial progression [2] [4] [8], and 10% may show accelerated progression [1] [14].

Magnetic Resonance Imaging (MRI) is considered the imaging study of choice for the approach to DT [3], especially for extremities [14], although the signal intensities demonstrate the proportions of its diverse components (collagen fibers, spindle cells, extracellular matrix), DT does not present a characteristic image [3]. On CT, the imaging study of choice for intra-abdominal DT [14], most of them present irregular borders and moderate enhancement to contrast medium [3] [6], and on ultrasonography, they may present a linear extension along the fascial plane known as the "tail sign" [3]. Likewise, CT and MRI are the imaging studies of choice for monitoring and evaluation of response to non-surgical treatments [1] [4]; although they can be evaluated using the RECIST 1.1 criteria based on the decrease in tumor size, this can underestimate the response to treatment, and therefore it has been proposed to additionally take into account the changes in tumor attenuation on CT and in signal intensity on MRI, since these changes may reflect a response due to the secondary increase in the fibrous component and the decrease in the inflammatory component of the DT [4]. PET CT does not have a definite role as DT generally exhibits mild 18-FDG avidity [1] [4].

Initial histologic diagnosis should be made by core biopsy; incisional or exci-

sional biopsy is not recommended, and due to the rarity of DT and possible histologic similarities with other entities, misdiagnosis rates of up to 40% have been reported with the most difficult diagnoses to differentiate being nodular fasciitis and low-grade fibromyxoid sarcoma [2] [3] [14]. Morphologic findings include low to moderate cellularity, a proliferation of uniform spindle cells resembling myofibroblasts during the proliferative stage of wound healing, a dense collagen stroma, and the absence of malignant features such as hyperchromasia and atypia [3] [4] [7]. Likewise, in more than 80% of cases, an aberrant nuclear localization of β-catenin is demonstrated by immunohistochemistry [7].

Immunohistochemistry is positive for β-catenin, smooth muscle actin, vimentin, COX2, androgen receptors, and estrogen receptors and negative for desmin, S 100, CD 34, h-caldesmon and c-kit [3] [4] [7]. β-catenin mutational analysis has been proposed as a specific diagnostic tool for DT, and even some of them have been related to prognosis, evolution, and prediction of response to treatment [2] [4]; specifically, the presence of the S45F mutation has reported an increased risk of recurrence and progression [2] [4] [8], and an effective response to meloxicam treatment. Finally, genetic testing can identify the presence of mutations in CTNNB1 or APC [14].

Currently, the initial treatment for almost all patients, especially if they are asymptomatic or with mild symptoms, without progression, and without associated morbidity [14], should be active surveillance with initial MRI or CT and subsequently with serial studies at intervals of 3 to 6 months (lack of radiation exposure makes MRI the ideal choice) [1] [2]. This surveillance begins once the histopathologic diagnosis and tumor extension have been established and continues for at least 2 to 3 more years [1]-[3]; subsequently, it can be continued with studies every 6 to 12 months with shorter time intervals if the tumor is located in critical areas such as the head and neck or mesentery [3]. Due to the evolution time greater than 5 years that the patient presented and the symptoms secondary to the tumor and its progression, we decided to initiate active treatment without an active surveillance period.

Although no factors have been identified that differentiate at the time of diagnosis between patients who will not require active treatment and those who may require it [1] [2], the persistence and/or increase of pain and a location in the shoulder girdle could be associated with a higher probability of progression [10] [16]; our patient presented both factors.

Radiological progression alone is not an indication for immediate treatment, especially if there is no symptomatology and the DT has an anatomical location that does not involve critical structures [1]. Likewise, the presence of symptoms alone does not constitute an indication to initiate active treatment because although pain control is a priority to achieve active surveillance, its presence is not an indication for active treatment because its pathogenesis may be multifactorial; in addition, it should be kept in mind that treatment for DT should not cause further secondary symptomatology [1].

Pregnancy should also not be considered as a direct indication for intervention

[2]. Although the risk of progression during pregnancy is up to 50% [3], DT can be kept under active surveillance since it does not increase obstetric risks and should not be considered as a contraindication for future pregnancies [2]; however, it is recommended to wait at least 1 to 2 years of active surveillance after the initial diagnosis prior to pregnancy to assess the behavior of DT [2]. In our literature review, we found only 23 reported cases of DT in pregnant patients (most of them developed in the abdominal wall), of which only 5 required surgical resection during pregnancy due to progressive growth; of the remaining patients, most were operated on after delivery and in 5 of them the surgery was very invasive due to tumor sizes of up to 20 cm [9]; regarding tumor size, we did not find cases similar to our patient that reported tumors larger than 20 cm.

Active treatment should be considered in the presence of tumor growth, progressive and persistent symptomatology (pain), progressive functional limitation of the limb, or an increased risk of associated morbidity [1] [14]; generally, the decision to initiate treatment should be made at least 1 to 2 years after diagnosis and/or in the presence of tumor size progression in consecutive imaging studies (at least 3) [1] [2], thus avoiding overtreatment in a patient who could present spontaneous regression or stable disease [1].

In another scenario, if the DT presents a critical anatomic location, such as head and neck or close to the mesenteric vessels, an early therapeutic decision could avoid potential morbidity secondary to tumor progression and/or wide resection (1 year after the start of active surveillance) [1] [3]. In addition to the time of evolution, persistent pain, and progression of tumor size, we decided to initiate active treatment because of the proximity of the tumor to the neurovascular structures of the thoracic limb.

After an initial period of active surveillance, if therapeutic intervention is required, the choice is generally guided by the anatomical site involved and the evolution of the disease [1]. For the treatment of our patient, we relied on the algorithm proposed by the Desmoid Tumor Working Group, which recommends active treatment based on the tumor site (in this case, the shoulder girdle) in the presence of clinical, symptomatic, and radiological progression after a surveillance period of at least 2 years [5]. We started with medical treatment, and due to the lack of response, we opted for surgical treatment because although the procedure entailed morbidity, it was still feasible to achieve an R0 resection as recommended in the NCCN guidelines, and there was also the risk of neurovascular involvement of the extremity due to the proximity of the tumor to these structures. Due to the size and unpredictable tumor behavior, we did not consider radiotherapy as the next therapeutic option because of the possibility of an uncertain and/or slow response and the secondary tissue damage that would hinder the closure after the wide resection and the coverage of the neurovascular structures of the extremity in case of a low tumor response. Furthermore, fortunately, the wide resection could be closed using musculocutaneous flaps without the need for complex flap rotations; this leaves us with other closure and reconstruction options in the event

of tumor recurrence.

Until the year 2000, surgical resection was considered the standard treatment [8], and today it continues to be an adequate option especially if it does not entail great morbidity [1]; although local control rates reported at 5 years are up to 80% [2], the incidence of local recurrence is 15% to 77% and the functional results can be unfavorable [4] [6] [7] [10] [13]. Surgery should currently be considered as a therapeutic option when morbidity is limited, in the presence of persistent symptoms, when there is a risk of further loss of function of the affected site, and if it is anticipated that an R0 resection is achievable [7] [8]; we opted for surgical treatment because although the procedure entailed morbidity, secondary symptoms and decreased quality of life persisted, there was a risk of neurovascular involvement of the limb and/or further tumor growth that would further infiltrate the arm muscles causing greater functional limitation of the limb, it was still feasible to achieve an R0 resection, and closure of the defect after resection could still be achieved by advancement of fasciocutaneous flaps without requiring advanced reconstruction techniques. We consider this last point relevant since the patient presents several risk factors for recurrence (tumor size, location, and age), and therefore, it is important to have other alternatives for closure and reconstruction in case a new resection is required.

The primary objective should be to achieve a microscopically wide resection margin (R0) while preserving as much functionality as possible (especially in extremities) [1] [2], however, this is not always possible due to the infiltrative growth pattern that can make the intraoperative distinction of the edges difficult [6] [8], and therefore if the likelihood of an R1 margin is anticipated other therapeutic options may be chosen [1] [3]. On the other hand, if the R0 margin is not possible for functional or cosmetic reasons, a positive microscopic margin (R1) may be acceptable if the R0 margin leads to excessive morbidity [1] since, as stipulated in the NCCN guidelines 2024, recent studies suggest that there is no difference in outcome between R0 and R1 margins [3]; Likewise, there is still no consensus on whether a positive margin correlates with an increased risk of recurrence and vice versa [6] [9], nor have improvements in recurrence rates been reported with reinterventions for margin extensions that may even result in increased morbidity due to the double procedure [1] [3]. Likewise, an aggressive resection that may significantly sacrifice function does not convincingly ensure a better oncologic outcome [6].

Surgery is also the initial approach in abdominal DT with secondary complications such as bowel perforation and/or obstruction, either limited to control of the complication or with tumor resection, taking into account morbidity and functional outcome [1]. Secondary gastrointestinal tract bleeding can be treated conservatively by interventional radiology and medical treatment against DT initially [1]. Likewise, patients with DT with skin invasion and/or secondary ulceration or with large cosmetic defects can be treated with initial surgery as long as it carries acceptable morbidity [1] [2], and finally, surgery can also be considered as a sec-

ond-line treatment (after surveillance) for abdominal wall DT (AW abdominal wall) as morbidity and recurrence risks are limited, elsewhere other therapeutic modalities should be chosen [5].

Radiotherapy may be an alternative to surgical treatment when the size or location of the tumor predicts a high risk of functional sequelae in the face of unresectability and/or as an alternative to radical surgery (amputation) [7]. Reported 5-year local control rates are 65% - 83% [6] [7], and as a single therapeutic modality, it may offer adequate control in patients with progressive disease in whom surgery is not a reasonable option and cannot be controlled with medical therapy [1] [3] [6]. Its main disadvantages include post-irradiation fibrosis, joint contracture, neuropathy, radium-induced neoplasms, and the time required for a complete response, which may take several months [6]. We do not consider radiotherapy as a therapeutic option because of the possibilities of an uncertain and/or slow response and secondary tissue damage that would make it difficult to close the defect secondary to wide resection and coverage of the neurovascular structures of the extremity in case of a low tumor response.

As adjuvant treatment after R1 resection, radiotherapy is not considered necessary because although reductions in the risk of recurrence have been reported with double therapy (36% versus 25%) [5] [7], these have not been statistically significant [5], and there is also a risk of development of secondary malignancies, especially in young patients [1] [7]. NCCN guidelines 2024 support the use of adjuvant radiotherapy in patients with positive margins only if it is anticipated that a new recurrence would be difficult to treat and/or lead to increased morbidity.

In locally advanced DT of extremities where resection would entail significant functional sacrifice, isolated limb perfusion (ILP) with tumor necrosis factor-alpha and melphalan may be useful, especially for multifocal, unresectable DT and/or those located in the hands and feet [2]. Other therapeutic modalities that have reported decreases in tumor volume and symptomatic improvement with low morbidity rates include high-intensity focused ultrasound (HIFU), percutaneous cryoablation, radiofrequency ablation, and selective delivery of cytotoxic chemotherapy through intra-arterial doxorubicin drug-eluting embolization [2] [3] [7].

Tumor location is one of the main risk factors for recurrence, with abdominal wall and lower extremity DT having the best prognosis; those of the chest wall, head and neck, mesentery, and upper extremity are associated with a higher risk of recurrence [2] [4] [5]. Other factors associated with an increased risk of recurrence include a history of previous recurrence, β-catenin mutational status, age (less than 37 years), tumor size (greater than 7 cm), and FAP [2] [3] [13] [14]; surgical margins do not appear to be associated with recurrence [2] [4] [8] [16]. Our patient presents as risk factors for recurrence, the size and location of the tumor, and her age, so she continues to be under close surveillance.

Systemic treatments include antihormonal therapies, non-steroidal anti-inflammatory drugs (NSAID), tyrosine kinase inhibitors (TKI), and "low-dose" or conventional chemotherapeutic regimens [2] [5]; if the tumor is not aggressive, fast-

growing, very symptomatic and/or unresectable, it is recommended to start with the least toxic drugs (antihormonal therapies and NSAID) [1] [2] [5] [8].

The use of antihormonal therapies is supported by the higher incidence of sporadic DT in women, the association of DT with pregnancy, and estrogen receptor tumor expression [7]; the most widely used is tamoxifen, which has reported clinical benefits in approximately 30% of cases, mainly in symptom control without significant radiological changes [7] [8]. As for NSAIDs, their mechanism of action could be based on the overexpression of COX-2 reported in many tumors that causes an increase in the expression of platelet-derived growth factors (PDGF), stimulating angiogenesis and resistance to apoptosis [7] [8]. Both drugs can be used alone or in combination as a first line because of their limited toxicity, low cost, and low rate of adverse events [2]; however, the reported response rates have been low, and there is no clear information on symptom improvement, nor changes in tumor size and/or changes in MRI signals, nor prospective comparative studies with active surveillance. Therefore, it cannot be assumed as a general recommendation [2] [7].

In DT with critical anatomic locations, with the progression of size and/or symptomatology despite anti-hormonal treatment, and/or when a more rapid response is required, the next therapeutic option is chemotherapy [2] [8]; at conventional doses, doxorubicin has reported response rates of up to 50% [8]. Due to the life expectancy of most patients with DT and the cumulative toxicity of anthracyclines, another alternative is low-dose chemotherapy with methotrexate and vinblastine/vinorelbine, which has reported clinical and radiological benefits in up to 80% of patients [8].

Due to the lack of comparative studies, the Desmoid Tumor Working Group has not yet established a sequence for the use of systemic treatments; there are only randomized phase 3 studies for sorafenib and phase 2 for the combination of low-dose methotrexate and vinblastine and for imatinib and pazopanib [1]. Reported response rates are 2.6% - 10%, 26.3% - 77%, 33%, and 37% for imatinib, sunitinib, sorafenib, and pazopanib, respectively, and 1-year PFS rates are 66% for imatinib, 89% for sorafenib, and 86% for pazopanib [14].

4. Conclusion

The treatment of DT should be multidisciplinary and individualized and should be based on its behavior, the location, and size of the tumor, its relationship with critical structures (sites where progression would lead to greater morbidity), the functional status of the limb if it is the site of development, secondary symptoms and whether or not their control is possible, the risk of complications secondary to tumor growth, age, and the patient's decision.

Consent

Written informed consent was obtained from the patient for publication of this case report.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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Abbreviation

Desmoid Tumor (DT); Tyrosine Kinase Inhibitors (TKI); Computed Tomography (CT); Body Mass Index (BMI); Mixed Anxiety-Depressive Disorder (MADD); Familial Adenomatous Polyposis (FAP); Magnetic Resonance Imaging (MRI); Non-Steroidal Anti-inflammatory Drugs (NSAID).