

# Subscapularis Fibromatosis as a Cause of Winged Scapula. Case Report and Literature Review

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## ABSTRACT

Winged scapula is usually caused by neurological injuries to both the spinal nerve and the long thoracic nerve. Its presence as a result of a ventral scapular tumor makes initial diagnosis difficult. We present the case of a young woman with limited external shoulder rotation, no known traumatic history, and images consistent with a soft tissue lesion dependent on subscapular muscle aponeurosis, which was confirmed by biopsy as a desmoid tumor. Although it is a benign, self-limiting tumor, it has an alarmingly high rate of relapse after resection, so many treatments are available, and many teams choose to closely monitor the patient's prognostic factors and functional limitations, obtaining satisfactory outcomes and, in some series, superior to those of surgical treatment.

**Keywords:** winged scapula, desmoid tumor, treatment.

**Level of Evidence:** IV

## Fibromatosis subescapular como causa de escápula alada. Presentación de un caso y revisión bibliográfica

## RESUMEN

La escápula alada suele producirse por lesiones neurológicas tanto del nervio espinal como del nervio torácico largo. La aparición a causa de un tumor ventral de la escápula dificulta su diagnóstico inicial. Presentamos el caso de una mujer joven, con limitación para la rotación externa del hombro, sin antecedentes traumáticos conocidos, evolución progresiva e imagen compatible con una lesión de partes blandas dependiente de la aponeurosis del músculo subescapular confirmada por biopsia como un tumor desmoide. Aunque se trata de un tumor benigno y autolimitado, tiene una inquietante alta tasa de recidivas después de la resección, por lo que se dispone de numerosos tratamientos y muchos grupos optan por hacer un seguimiento médico estrecho de los factores pronósticos y las limitaciones funcionales del paciente, con lo que obtienen resultados satisfactorios y, en algunas series, superiores a los del tratamiento quirúrgico.

**Palabras clave:** Escápula alada; tumor desmoide; tratamiento.

**Nivel de Evidencia:** IV

## INTRODUCTION

Extra-abdominal fibromatosis is a rare aggressive monoclonal fibroblastic proliferation of musculoaponeurotic tissues. Two types are distinguished: superficial ones, such as Dupuytren's contracture, Ledderhose disease or penile fibromatosis, and deep ones, which are called aggressive desmoid tumors because of their local behavior.<sup>1</sup> The term desmoid, derived from the Greek 'desmos' meaning 'band', was initially applied by Müller because of its consistency similar to that of tendons. They have a slight prevalence in the population between the ages of 25 and 35, although, in many series, no differences are observed according to age; it represents about 0.03% of all skin and soft tissue neoplasms, and the risk increases if the patient has a genetic condition, such as Gardner syndrome. The prevalence of extra-abdominal desmoid tumors increases to 15% if associated with familial adenomatous pol-

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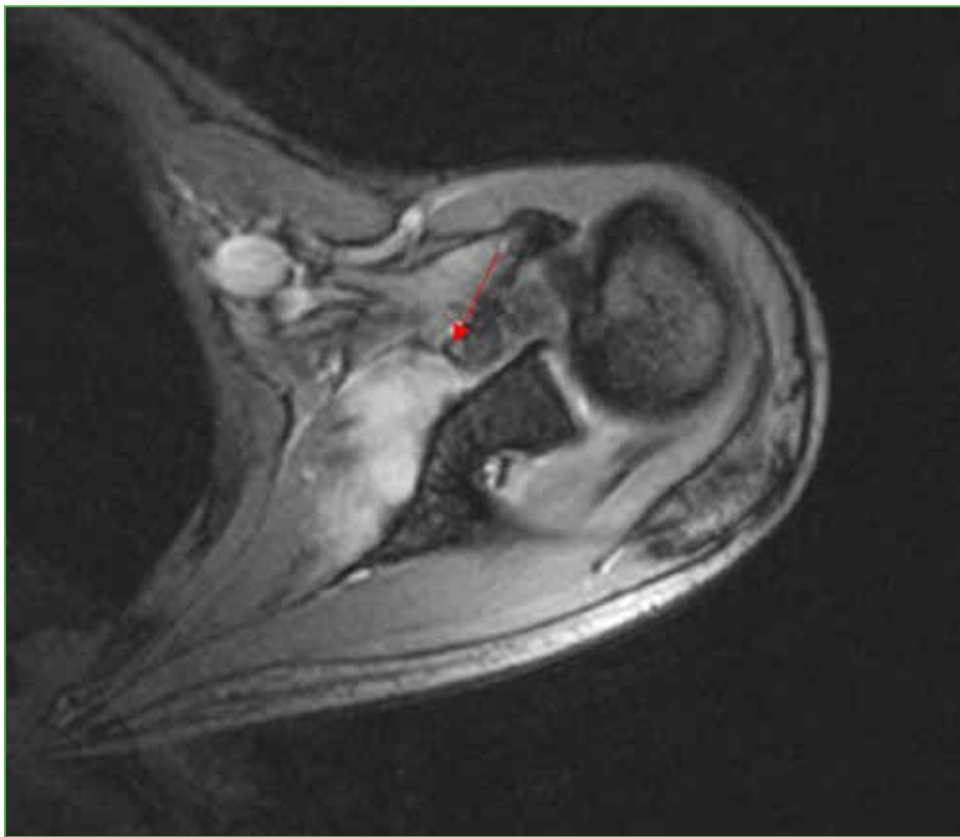
yposis,<sup>2</sup> which, in addition, determines that those with desmoid tumors are more prone to malignancy than those without this condition.<sup>3</sup>

Desmoid tumors do not metastasize, but they have irregular limits because they are not contained in a capsule, this factor is associated with greater aggressiveness and recurrence after surgery.

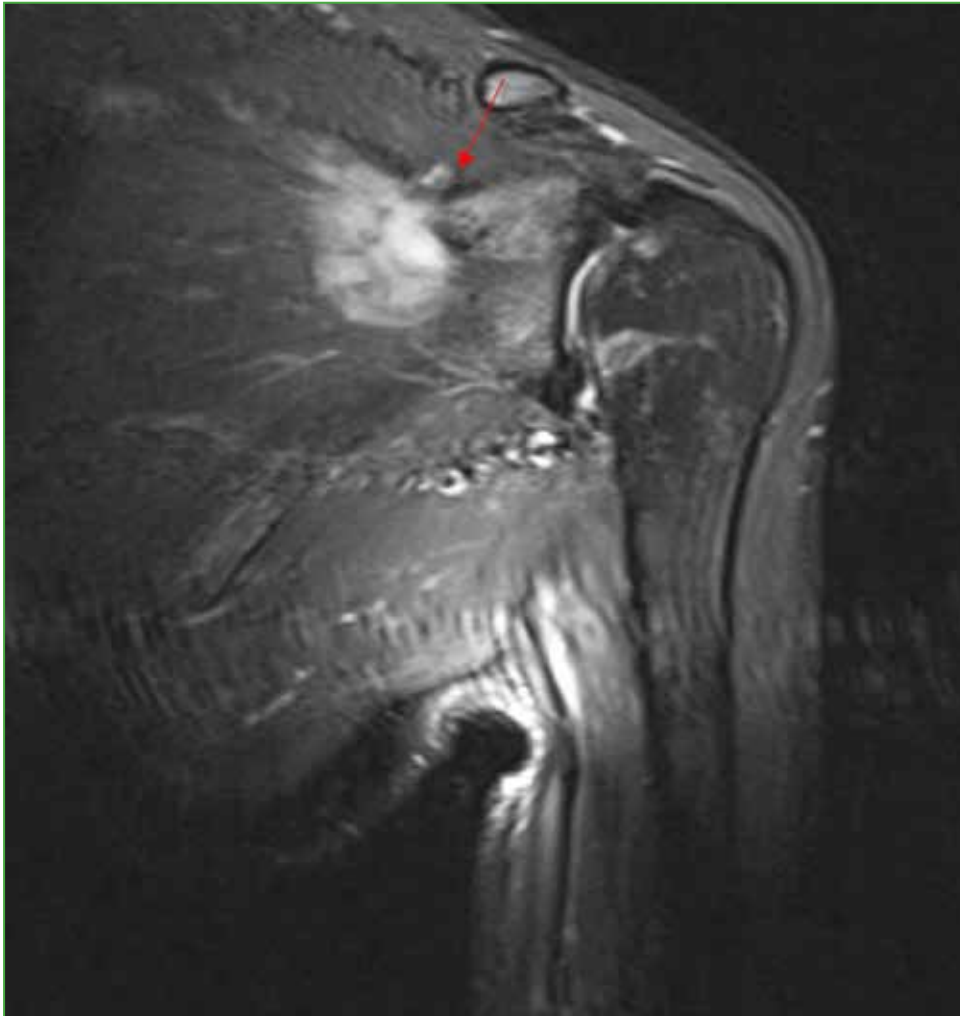
Periscapular tumors and, even more so, desmoid tumors, are a rare cause of scapulothoracic dissociation, making them difficult to diagnose. In this regard, winged scapula is usually due to paralysis of the serratus anterior or trapezius muscles due to nerve injury to the long thoracic or accessory nerves, but these injuries are dynamic and must be differentiated from static ones that do not occur due to neuromuscular causes, which means that this type of winged scapula is present at rest and does not increase with anterior flexion of the arm.<sup>4</sup>

## CLINICAL CASE

A 28-year-old woman with a history of idiopathic adult scoliosis and with an active job that involved carrying material. She was referred to the trauma outpatient clinic for post-traumatic left omalgia of eight months of evolution and limitation for external rotation, contracture and trapezius pain, with no improvement after rehabilitation. On physical examination, relative hypotrophy of the left scapulohumeral musculature compared to the contralateral scapulohumeral musculature was detected. No masses were palpated or inflammatory signs were observed. Active and passive range of motion were preserved, except for an external rotation of 0° for the left shoulder and 75° for the right shoulder. In addition, scapulothoracic dyskinesia compatible with a left winged scapula was found, so different complementary studies were requested. With the CT scan, bone disease was ruled out. On electromyography, a lesion of the long thoracic nerve, with chronic characteristics and of moderate intensity, was detected, which impeded the mobility of the scapula. In addition, magnetic resonance imaging revealed a lesion in the subscapularis muscle of about 5 cm, with poorly differentiated margins, discrete heterogeneity and a slight signal hyperintensity in the T2-weighted sequence, compatible with a fibromatous process (Figures 1 and 2).



**Figure 1.** MRI of the left shoulder, axial section, STIR sequence. The red arrow indicates the subscapular tumor.



**Figure 2.** Magnetic resonance imaging of the left shoulder, coronal section. The red arrow points to the subscapular tumor.

To confirm the diagnosis, a thick needle biopsy was performed under sedation and guided by computed tomography. The sample was kept fresh until it arrived at the pathological anatomy laboratory where it was processed. Histological analysis showed a low-grade mesenchymal spindle cell proliferation, suggestive of desmoid-type fibromatosis that locally infiltrates the subscapularis muscle. Therefore, the case was treated in conjunction with a tumor unit, and it was decided to complete the evaluation with a scan that showed the absence of tumor activity.

In the evaluation of the specialized unit, it was decided to manage the inflammatory disease by controlling the symptoms through radiotherapy and hormonal suppressive therapy. Surgical intervention was ruled out due to the high morbidity of the procedure, the difficulty in performing an extensive resection, and the high rate of relapses after surgery.

The patient received radiotherapy sessions and started hormonal treatment with tamoxifen 40 mg daily. After a year of work leave, she was told that she could return to her activities and she resumed her work with moderate-severe pain, both during active range of motion and at rest. This required monitoring by the chronic pain unit, where analgesia was reinforced at the third level of the World Health Organization pain scale and trigger point injections were indicated, first, with local anesthetics on a diagnostic basis and then with botulinum toxin, in order to improve the reflex contracture associated with the muscles of the shoulder girdle.

The patient underwent biannual monitoring with magnetic resonance imaging, which showed a decrease in tumor size of 10 mm since diagnosis. Cryo-sclerotherapy was contemplated, but the patient decided to continue symptomatic medical treatment because of the risk of a possible reported nerve injury.

Tumor growth and functional impairment were stabilized from diagnosis to 20 months of follow-up. Periodic checks were carried out every six months, using magnetic resonances without contrast medium and with the usual sequences, which showed that the mass had not grown. In addition, clinical examinations were carried out every three months to assess the progression or improvement of symptoms. The doctors in the chronic pain unit administered ultrasound-guided injections of extended-release betamethasone (2 ml) combined with mepivacaine 2% (6 ml) into the underlying muscles, which achieved good pain control. Three years after the diagnosis, the patient was discharged because the injury had not progressed.

## DISCUSSION

Currently, the treatment of extra-abdominal desmoid tumors remains a controversial topic, mainly due to the difficult local control of the disease and the high rate of recurrence in published case series. In a study of 194 patients with extra-abdominal desmoid tumors, most of which were in the extremities, attempts to control the disease locally through surgery with intralesional resection, whether marginal or extensive, had a recurrence rate of up to 76%, 70% within the first two years, and 60% if radiotherapy was used.<sup>5</sup> In another study, a series of 83 cases were evaluated. Most had been treated only with surgery and the local recurrence rate was 45%, with no difference with a small group that could not be properly resected and was prescribed radiotherapy.<sup>6</sup> More recently, a study of a series of 234 operated patients has been published, with a success rate of 83% in primary surgeries; these results include 10 recurrences in a group of 39 patients with radiotherapy and four in one of eight with chemotherapy only. In addition, a higher rate of recurrence of tumors located in the upper limb is reported, which is probably due to the limitation to perform more aggressive surgeries.<sup>7</sup>

The authors of most of the studies on these tumors highlight the importance of controlling tumor margins and their relationship with local relapse. For this reason, radiotherapy is incorporated into the therapeutic approach, both as an adjunct treatment to surgery and as a single treatment when surgery is not possible. In this regard, some studies have obtained good outcomes with adjuvant radiotherapy in patients with affected resection margins and neoadjuvant radiotherapy during a six-year follow-up.<sup>8,9</sup> The case presented is based on these results to justify the choice of radiotherapy at the beginning of the therapeutic regimen.

Systemic treatment is recognized as a line of therapy for extra-abdominal desmoid tumors. It may include nonsteroidal anti-inflammatory drugs, hormonal response-modulating drugs, and chemotherapy drugs. In this regard, in some studies of patients with desmoid tumors, the response to meloxicam has been good, stabilizing the disease and even making it remit.<sup>10</sup> Although up to 80% of sporadic desmoid tumors have mutations in the gene coding  $\beta$ -catenin, a specific study on mutations in this gene with a series of 145 samples found no relationship between the variants and the risk of recurrence.<sup>11</sup>

According to the European Soft Tissue and Bone Sarcoma Group, patients with clearly progressive, unresectable conditions, such as that of our patient, or in whom extensive resection leads to impairment or loss of limb function, are candidates for systemic therapy.<sup>12</sup> Despite correct case selection, there is currently no standardized treatment, which must be adapted to each patient. Specifically, and clearly considering these inclusion criteria, in the study by Mankin et al.,<sup>7</sup> only 4% of patients were treated with chemotherapy from the beginning, unlike other series in which these rates triple.

With regard to the treatment of desmoid tumors through chemotherapy, the use of doxorubicin associated with dacarbazine and vinorelbine stands out, with significant clinical response rates, without adverse reactions due to serious toxic effects.<sup>13,14</sup>

In the last decade, interest has grown in the use of molecules that interact, in a specific way, in the cell cycle, such as the tyrosine kinase inhibitors, imatinib and sunitinib. In several clinical studies, a reduction in radiological size has been detected in symptomatic patients treated exclusively with sorafenib,<sup>15</sup> and even a two-year progression-free survival of 81% of patients with advanced, treatment-resistant disease.<sup>16</sup>

On the other hand, it is believed that desmoid tumors may be related to hormones, since 80% have been found to affect women and their progression related to pregnancy has even been observed.<sup>17</sup> In 2003, a compilation of 34 reports of primary and recurrent cases with an average follow-up of 17 months was published. Three of them had a complete response; 15, a partial response and 12 showed no change in response to tamoxifen during follow-up.<sup>18</sup> In another study, similar outcomes were obtained with toremifene.<sup>19</sup> As for testolactone, an enzyme that controls the conversion of testosterone to estrogen, the largest published series is of 17 patients. The disease control rate was 40% and reached 70% if it was combined with a non-steroidal anti-inflammatory drug, such as sulindac or indomethacin.<sup>20</sup>

### Conservative treatment — *wait-and-see* policy

Conservative management of the disease through symptomatic medical treatment to avoid the morbidity associated with surgery and radiotherapy is defined as serial magnetic resonance imaging monitoring combined with symptomatic treatment with or without nonsteroidal anti-inflammatory drugs. This therapeutic option is receiving increasing support even as a first-line treatment for primary tumors.<sup>19</sup> Specifically, in a retrospective series of 87 patients analyzed based on the management of their primary desmoid tumor, either with surgery or through symptomatic treatment and periodic magnetic resonance, no significant difference was found in disease-free survival between the two groups.<sup>21</sup> Along these lines, in 2017, the French Sarcoma Group published a prospective study with 771 patients that compared event-free survival in an operated subgroup and another treated symptomatically. It was concluded that there are no statistically significant differences between the two options. It was also reported that, after a univariate analysis, the location of the desmoid tumor is the isolated parameter that most influences event-free survival.<sup>22</sup>

Currently, sarcoma research groups recommend conservative symptomatic management (*wait-and-see policy*) for extra-abdominal desmoid tumors with periodic magnetic resonances, especially during the first year, since only a very small amount grows beyond the first 36 months of diagnosis.<sup>23</sup> This avoids overtreatment and the excessive indication of surgery in these patients and helps to reduce the rate of sequelae.

## CONCLUSIONS

Because the etiology of desmoid tumors is multifactorial and their nature is unclear, it is difficult to determine a specific treatment and its possible evolution if no treatment is indicated. Many factors influence the poor outcome, such as the size, location and age of the patient. On the other hand, the factors that favor spontaneous resolution are still unknown.

Conservative treatment as the first line of treatment for extra-abdominal desmoid tumors is a completely valid option, although, in most studies, the abandonment of this therapeutic option remains a problem that may alter the results. A high percentage of patients achieve the spontaneous stabilization of the tumor in an average of two years. Therefore, according to our criteria and following the consensus of the Soft Tissue and Bone Sarcoma Group, we have chosen this line of treatment with the predictability of obtaining good outcomes in tumors that are unresectable—due to the high comorbidity that resection may entail, especially in the limbs—as well as in patients without risk factors for progression with tumors with few symptoms. We choose surgery for patients who do not respond to medical treatment after two years of follow-up, provided that the resection of the tumor does not entail a loss of relevant functionality and in cases where the surgical management of these tumors may not result in greater morbidity due to their location, such as those that settle in the neck or on the walls of the abdomen, and always reserving limb amputation for patients with a significant functional compromise present or expected after resection.

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