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Desmoid Tumor of the Arm: A Case Report

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Abstract **Case Report**

Desmoid tumor or aggressive fibromatosis is a benign soft tissue tumor that develops from fibrous tissue and consists of elongated cells resembling fibroblasts. Treatment can range from simple monitoring or medical treatment to surgical treatment with or without radiotherapy. We report here the case of a 45-year-old female patient who presented with a desmoid tumor in the right arm, initially confirmed by pathological examination of a biopsy and then treated by surgery (excision) without radiotherapy. At 5 months postoperative, the patient is asymptomatic with near-normal shoulder joint range of motion and normal local examination.

Keywords: Desmoid Tumor, Surgery, Aggressive Fibromatosis, Case Report.

INTRODUCTION

Desmoid tumor or aggressive fibromatosis is a benign soft tissue tumor that develops from fibrous tissue, consisting of elongated cells resembling fibroblasts [1]. It is a benign tumor that does not metastasize but can be locally aggressive and invasive [2]. It is a rare condition, more common in young adults (25-35 years) and more frequently in females (3/1). Its incidence is approximately 300 new cases per year in France and 900 new cases in the United States.

The exact causes are still unknown, but desmoid tumors may be related to traumatic, hormonal (more common during pregnancy), or genetic factors (Familial Adenomatous Polyposis). Most cases are non-genetic (not transmitted within families), mainly linked to a mutation in a gene encoding the protein β-catenin [3], which is involved in the regulation of cell growth genes. Familial cases (5-10%) are associated with Familial Adenomatous Polyposis and Gardner's syndrome [4].

Extra-abdominal desmoid tumors represent more than 50% of all desmoid tumors. The shoulder region is the main extra-abdominal location, followed by the chest and thigh [5].

Below, we describe the case of a 45-year-old female patient with a desmoid tumor involving the long head of the triceps brachii and the deltoid muscle.

The patient was informed that the case data would be submitted for publication, and she gave her consent.

CASE REPORT

The patient is a 45-year-old female with no notable medical history, presenting with progressively worsening right shoulder pain over 3 months, accompanied by limited mobility, prompting her to consult an orthopedic surgeon in private practice. An MRI was performed, indicating an intramuscular tumor process. The patient was referred to us at the Ibn Sina University Hospital in Rabat for further management.

The patient reported persistent, progressively worsening shoulder pain. Clinical examination revealed no local inflammatory signs, no sensory-motor deficit, with palpable pulses. However, there was an inability to abduct the shoulder and limited extension. External rotation was slightly reduced compared to the contralateral side, with no signs of rotator cuff involvement. The palm-up test, lift-off test, and Jobe's test were negative.

A standard radiograph revealed a soft tissue mass at the level of the upper arm without lytic bone lesions (Figure 1).

MRI revealed a tissue lesion process in the junction of the upper and middle thirds of the external aspect of the right arm, intra-compartmental, sub-aponeurotic, roughly oval, with irregular contours,

poorly delineated, involving the lateral head and long head of the triceps and the lateral portion of the deltoid muscle, showing isosignal on T1, heterogeneous hypersignal on T2 and STIR, and intensely heterogeneous enhancement after contrast, measuring 4.8 cm × 3.1 cm × 6.7 cm. It had intimate contact with the outer cortex of the right humerus without periosteal thickening or cortical rupture (Figure 2).

A biopsy was performed, showing a tumor proliferation of spindle cells forming divergent bundles of elongated cells with nuclei devoid of atypia, consistent with a desmoid tumor.

The patient underwent wide tumor resection without adjuvant chemotherapy or radiotherapy.

The patient was immobilized with a sling for one month, with analgesic treatment. Rehabilitation began on the 15th day after surgery. The final diagnosis was desmoid-type fibromatosis.

The patient was re-examined at day 15, one month, and three months postoperatively, and was asymptomatic with no pain or mobility limitation. Her flexion/extension range was 180°/50°, abduction/adduction 180°/50°, and external/internal rotation 70°/80°.

A follow-up MRI for recurrence detection is scheduled at 2 years, 5 years, and 10 years, if the patient remains asymptomatic.

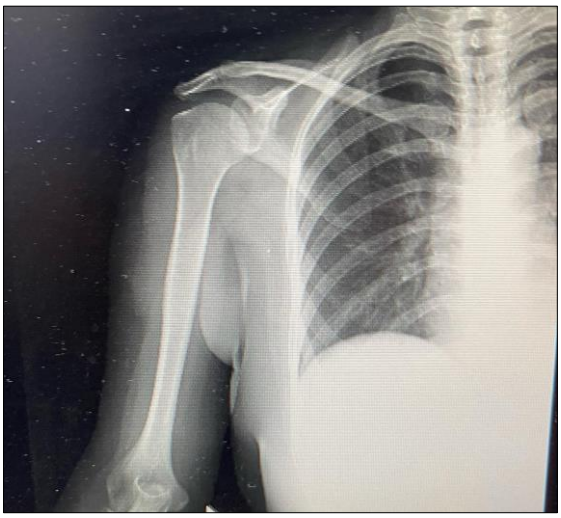

Figure 1: X-ray of the right shoulder, AP view

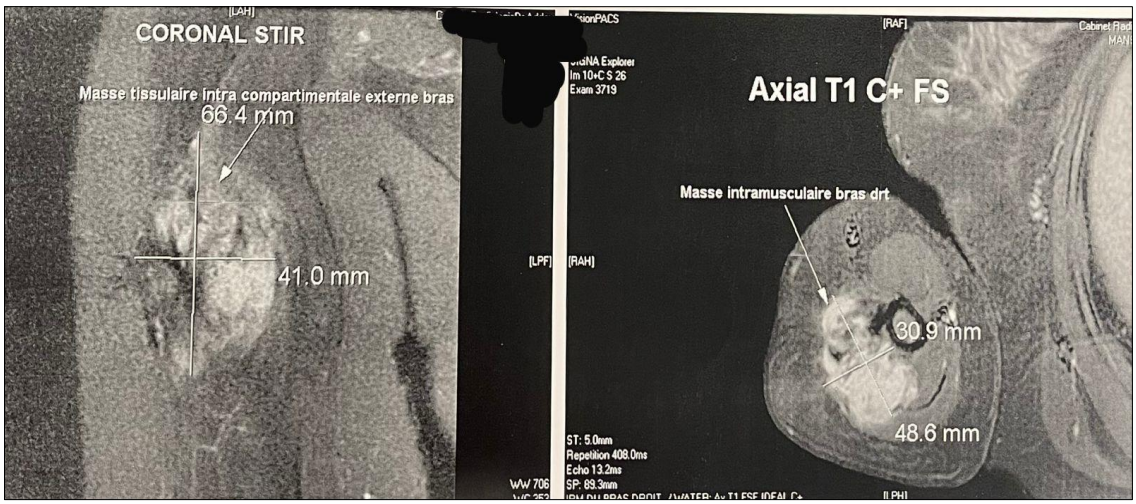

Figure 2: MRI of the right arm, axial and coronal slices

DISCUSSION

Desmoid tumors are rare fibroblastic tumors characterized by the presence of normal-appearing fibroblastic cells proliferating in an abundant collagenous stroma.

Desmoid fibromatosis can occur in the abdominal, intra-abdominal, or extra-abdominal regions. The most common extra-abdominal locations are the limbs, followed by the head and neck [6]. Although these tumors are benign, their clinical behavior is unpredictable. Rock *et al.*, in a series of 194 patients with

extra-abdominal desmoid tumors, reported a recurrence rate of 68% at an average of 1.4 years after the initial treatment [7]. This aggressive nature combined with the presence of vital elements nearby poses a major therapeutic challenge.

Currently, the management of desmoid tumors involves the use of surgery, radiotherapy (RT), as well as cytotoxic (methotrexate, vinblastine, and doxorubicin) and non-cytotoxic chemotherapy (tamoxifen, testolactone). Given the benign nature of these tumors, treatment strategies aim to achieve local control while preserving proper function and providing adequate aesthetic outcomes [2]. When technically feasible, complete tumor resection is the preferred treatment [8].

Primary radiotherapy is an appropriate alternative for patients who are not good candidates for surgery [9], but its role remains debatable, with no definitive scientific evidence regarding the best candidates for this therapy. The use of postoperative radiotherapy, as described in Nuyttens' series of 22 patients [9], demonstrated improved local disease control.

Recently, there has been a trend toward a conservative approach as the initial treatment for desmoid tumors. Rudiger *et al.*, recently reported similar recurrence rates in patients treated with radiotherapy alone compared to those treated with surgery and radiotherapy [10].

Some initial responses to chemotherapeutic agents were described by Weiss and Lackman in 1989, who used a combination of vincristine and methotrexate according to a specific protocol with fairly satisfactory results. Vincristine was later replaced with vinorelbine to reduce the incidence of neurological complications [11].

More recently, chemotherapy regimens based on doxorubicin have been used in the treatment of desmoid tumors. This regimen has often been used in combination with dacarbazine, with positive results [8], but with severe complications, particularly documented cardiotoxicity and myelosuppression. Gega *et al.*, used continuous infusion of these two drugs for 96 hours, followed by meloxicam, achieving a complete response in three out of seven patients and a partial response in the other four. Most studies using such combinations have long-term follow-ups of over 5 years, confirming a long-term positive effect.

Since surgery results in damage to nearby unaffected soft tissues, cryoablation has been proposed, particularly for the treatment of small extra-abdominal desmoid lesions. Kujak *et al*., reported positive results in their series of 5 patients [12].

A desmoid tumor is a benign and rarely fatal tumor, but it is a locally invasive disease. Surgery has been considered a standard treatment despite high recurrence rates ranging from 30% to 40%; because desmoid tumors can infiltrate muscle fibers, it is difficult to obtain a "clear margin." As an alternative, some authors have reported the use and effectiveness of NSAIDs, as well as cytotoxic and non-cytotoxic chemotherapy [13]. However, studies with long-term follow-ups are necessary to determine recurrence rates and functional outcomes of the operated area.

In summary, desmoid tumors are rare and benign. Treatment strategies must be individualized to achieve local control followed by functional and aesthetic control. These patients must be closely monitored through clinical and radiological examinations at regular intervals.

CONCLUSION

Extra-abdominal desmoid tumors are rare tumors that develop in the body's soft tissues outside the abdominal cavity. Although they are generally benign, these tumors can be locally aggressive and impact patients' quality of life.

The management of extra-abdominal desmoid tumors can vary depending on different factors, such as location, size, and disease progression. Treatment options may include surgery, radiotherapy, chemotherapy, or a combination of these approaches.

It's also worth noting that research on extra-abdominal desmoid tumors is constantly evolving, with new therapies and approaches emerging. Participation in clinical trials may offer interesting opportunities for patients seeking innovative treatment options.

Conflicts of Interest: The authors declare no conflicts of interest.

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