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Surgical approach and resolution without prosthesis of desmoid fibromatosis of the shoulder girdle. Report of a case

Abordaje quirúrgico y resolución sin prótesis de fibromatosis desmoide en cintura escapular. Reporte de un caso

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Keywords: desmoid fibromatosis, surgery, Tikhoff-Linberg.

Palabras clave: *fibromatosis desmoide, cirugía, Tikhoff-Linberg.*

ABSTRACT

In a 46-year-old woman with an intermuscular lipoma who started suffering from pain in the right shoulder, with the presence of a tumor of progressive growth in the ipsilateral supraclavicular region, a biopsy was performed. As there was no improvement, computerized axial tomography was taken, showing a right subscapularis tumor that eroded the scapular spine and replaced the subscapularis muscle. Resection of the right shoulder girdle type 4, Tikhoff-Linberg, without the use of scapula or humerus prosthesis, with a histopathological and immunohistochemical report of fibromatosis, was done. Currently, the patient is with pharmacologically controlled pain and limited movement. Desmoid fibromatosis has an incidence of two to five cases per million population. Surgical resection is the beginning of treatment, using the Tikhoff-Linberg technique with the use of a prosthesis. In this case, an alternative surgical variable was performed without the use of a prosthesis.

RESUMEN

Mujer de 46 años que inicia padecimiento con dolor en hombro derecho, al que se agrega la presencia de tumoración de crecimiento progresivo en la región supraclavicular ipsilateral, se realizó biopsia que reporta lipoma intermuscular. Al no tener mejoría, se lleva a cabo tomografía axial computarizada donde se observa tumor subescapular derecho que erosiona espina escapular y reemplaza al músculo subescapular. Se hace resección de cintura escapular derecha tipo 4, Tikhoff-Linberg, sin uso de prótesis de escápula ni de húmero, con reporte histopatológico e inmunohistoquímico de fibromatosis. Paciente actualmente con dolor controlado farmacológicamente y con movimientos limitados. La fibromatosis desmoide tiene una incidencia de dos a cinco casos por millón de habitantes, la resección quirúrgica es el inicio del tratamiento, al emplear la técnica Tikhoff-Linberg con el uso de prótesis, en este caso se realizó una variable quirúrgica sin el uso de prótesis.

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INTRODUCTION

Desmoid fibromatosis, also known as a desmoid tumor or aggressive fibromatosis, is a rare condition with a frequency of two to five cases per million inhabitants per year;[1-3] it is characterized by the proliferation of fibroblasts, which, despite not having histological characteristics of malignancy, are locally aggressive and with an unpredictable clinical behavior.[2,3] The World Health Organization (WHO) describes it as a monoclonal proliferation of fibroblasts affecting soft tissues with infiltrative growth and a tendency to local recurrence, but without distant metastasis.[4]

This type of tumor occurs in patients between 20 and 70 years of age, with a peak in frequency between 30 and 40 years of age.[2,4]

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According to their location, they are divided into the extra-abdominal, the abdominal wall, and the intra-abdominal. Of the extra-abdominal tumors, 17% are in the shoulder girdle.[3] The treatment of choice is surgical resection, which is a surgical challenge due to the difficulty to distinguish the capsule from the tumor and the high risk of injury to the brachial plexus and axillary vessels.[1-3] In addition, these factors may require multiple surgical procedures and cause severe aesthetic and functional morbidity, so the decision-making is complex in this type of case.[5] The complexity of the surgery is related to the tumor size and location, for which a local control rate of 80% at five years is reported.[4] Due to all these factors that make resection with negative margins complex, adjuvant radiotherapy is the method of choice to avoid recurrences.[2] The main treatment is surgical resection, whose priority is to preserve function, but in cases where morbidity is high, observation can be considered while the patient is asymptomatic; when surveillance fails, surgery is the next option.[6]

CASE REPORT

This is a 46-year-old woman with a history of bilateral tubal occlusion performed 10 years earlier. She denies any familiar oncologic history. Her current condition began some seven years before with right shoulder pain, non-radiating, continuous, and moderate to severe in intensity, with no response to non-steroidal anti-inflammatory drugs, but with partial response to buprenorphine, which was discontinued due to adverse effects.

About 10 months later the patient self-detected a slow-growing right supraclavicular mass, with mild pain elicited only on palpation. Magnetic resonance imaging (MRI) was performed that showed an infrascapular lesion. Four months later a new MRI was performed showing almost complete loss of the subscapularis tendon insertion compatible with a tear. Surgical exploration of the right rotator cuff with a biopsy of the ipsilateral supraclavicular tumor was performed. In February 2015 the tumor was resected through the right trapezius muscle, which was approached through a previous scar on the right shoulder. This first histopathological report (HPR) indicated an intermuscular lipoma, with no other alteration of the muscle or fatty tissue cytoplasm, nor changes in the nuclei.

Due to postoperative pain, the patient was sent to radiotherapy, after an MRI that showed a subscapular lesion with extension towards the supraclavicular fossa, involving the subscapularis muscles. It could not be established with certainty if it was a fatty tumor, nor if it had an intrathoracic extension; therefore, a CT scan was performed showing a heterogeneous right subscapular tumor measuring 11.6 × 11.5 × 9.4 cm, with increased vascularity, that was compressing and displacing the internal jugular vein. It was not eroding the ribs, but it was eroding the scapular spine. No adenopathy was seen. The tumor was shown to involve the subscapularis muscle and replacing it in its entirety *(Figures 1 and 2)*. The axillary and supraclavicular neurovascular bundles were identified. In May 2055 a percutaneous biopsy was performed with a histology report of myofibromatosis.

On July 2015, a Tikhoff-Linberg type 4 right shoulder girdle resection surgery was performed without the use of a scapula or humerus prosthesis. Cephalothin was administered as a preoperative antibiotic without any additional drug. The surgical approach was through an incision and lifting of a supraclavicular skin flap, with dissection of the deltoid and suprascapular muscles. Once the area was exposed *(Figure 3)* the dissection of the mass was performed with resection of the clavicle two distal thirds, as well as the elimination of the proximal humerus up to its surgical neck. The humerus was anchored to the clavicle with a polyester suture #3. The aponeurotic fasciae were closed in planes with a 2-0 vicryl anchored continuous suture. In the postoperative period, there was no dehiscence, seroma, or hematoma formation, nor infection.

The histology report described a neoplasia with the anterior surgical edge in contact with the tumor. The rest of the margins were tumor-free and respected the subclavian vein and artery. The definitive histology report histologically described a non-atypical cellular proliferation, without evidence of mitosis, with spindle-shaped proliferating elements with

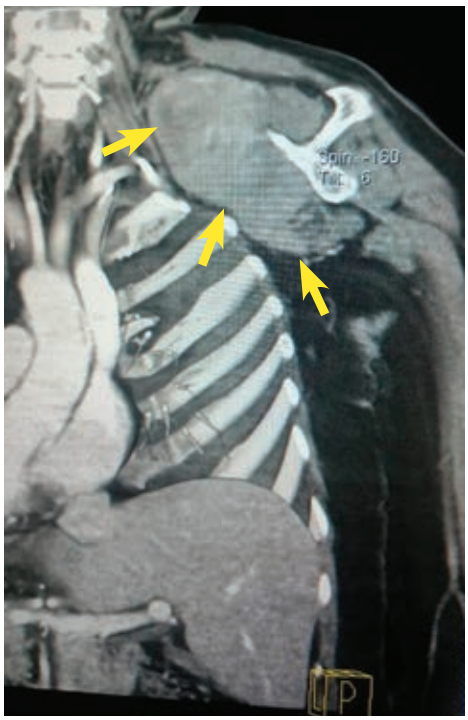

Figure 1: *CT scan showing a heterogeneous right desmoid subscapularis tumor with hypervascularity, replacing the entire subscapularis muscle (shown between arrows).*

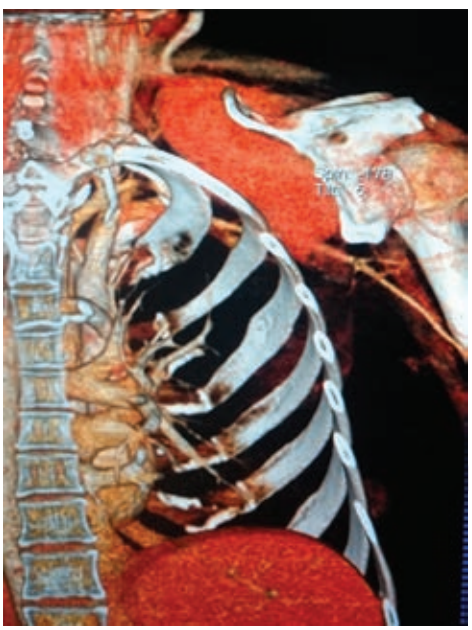

Figure 2: *A computed axial tomography reconstruction showing the right subscapularis tumor.*

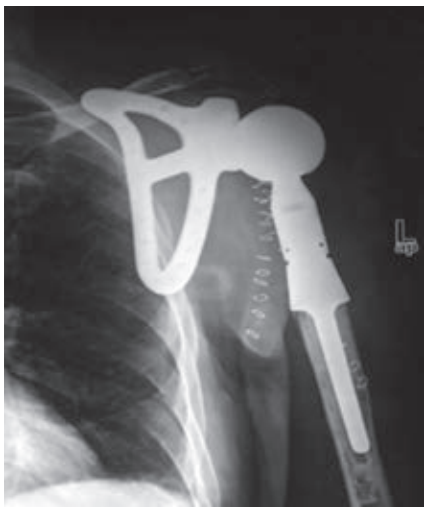

Figure 3: *Prosthesis of the scapula and proximal humerus after resection of these structures.*

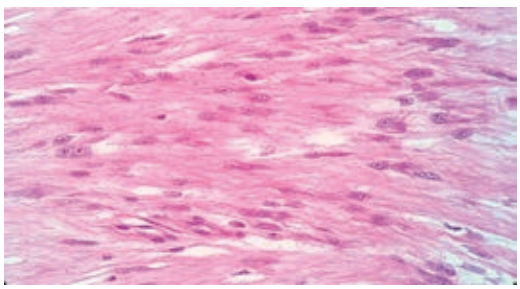

Figure 4: *Histopathologic section (40×), showing non-atypical cell proliferation, without evidence of mitosis.*

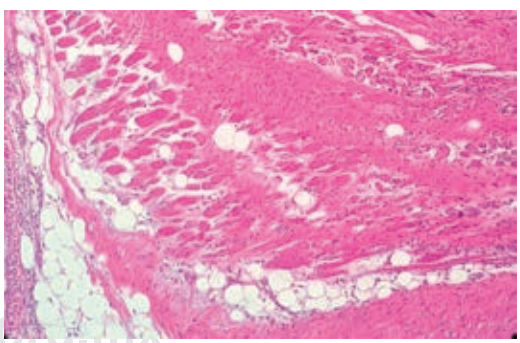

Figure 5: *Histopathologic section (10×) showing the fibromuscular bundle.*

intercellular collagenization and infiltrating adipose and muscular tissue in the periphery *(Figures 4 to 6)*.

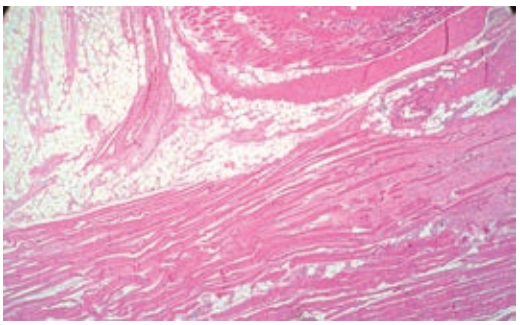

Figure 6: *Histopathologic section (40×) showing the fibromuscular bundle.*

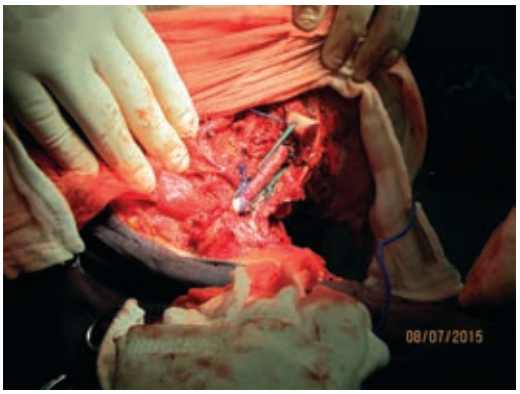

Figure 7: *"Anchoring" of the humerus to the residual clavicle with polyester suture after tumor resection.*

After the third week following surgery, the patient was found to have mild pain on passive movement of the shoulder, with adequate strength in the deltoid muscle, but without abduction, flexion, or extension of the shoulder due to lack of support sites in the joint. A rehabilitation program was started. She was sent for adjuvant radiotherapy but was not considered a candidate. Instead, she was prescribed thalidomide which she had continued until her last evaluation.[6]

A comparative computed tomography (CT) scan taken in 2017 showed a heterogeneous lesion at the site where the humeral head was, with calcifications inside, extending intramedullary, which had decreased in size from 3.5 to 2.7 cm. Another CT scan taken in September 2018 showed no lesion. In 2019, the patient mentioned she was stable, and her pain was well controlled with paracetamol and gabapentin, which is the treatment suggested in the current National Comprehensive Cancer Network (NCCN) guidelines.[6] She kept full hand and elbow functions, and no shoulder functions. Due to the COVID-19 pandemic, the patient was lost to follow-up.

DISCUSSION

Currently, 95% of patients with sarcomas of the shoulder girdle are treated with surgery, which preserves the limb and function of the elbow, wrist, and hand.[7] The Tikhoff-Linberg surgery was first described in 1928[8] but underwent subsequent modifications, including the Malawer classification that is used today.[9]

Depending on the patient, and the symptomatology and morbidity expected from resection, the surgical approach versus observation may be considered. The choice of surgery depends on the size of the tumor, the speed of growth, and the symptomatology it causes.[4] As it was shown in the case presented, the growth evolved for two years and the pain was the main symptom, which coincides with the reported literature.[10] The local control rate with surgery ranges from 68 to 75%. These rates do not improve with adjuvant radiotherapy and even recurrences increase with this type of treatment.[5,10]

The approximate incidence of this case is 2-5:1'000,000, so it is a very rare entity. In studies where the use of the Tikhoff-Linberg type 4 technique is reported,[10,11] scapula and/

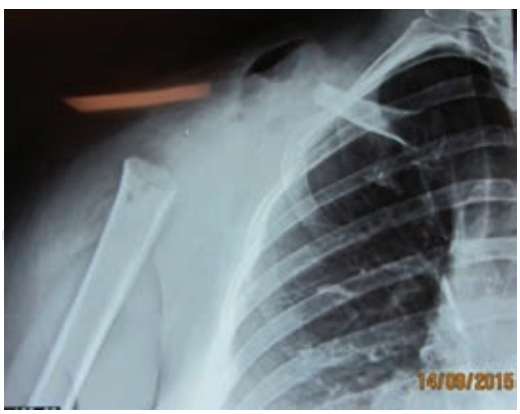

Figure 8: *X-ray after the surgical procedure.*

or humerus prosthesis are used *(Figure 7)*. In this case, an alternative surgical procedure was performed without the use of a prosthesis. This way, the humerus of the residual clavicle was "anchored" using polyester suture *(Figure 3)*, a technique which, despite being described by Linberg,[8,11] has not been used in any of the published cases of its use. We show the image of post-procedure radiography *(Figure 8)*.

An expected medium-term outcome is a symptomatic improvement. Regarding the lineage, the variable found in this case is even rarer. At five years it has a prognosis of 80% of local control after surgery if the surgical margins are negative margins.[4,12] CAs far as we know, the patient is stable, and pain is controlled with drugs.

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