



## Chondromyxoid fibroma of the clavicle. Case contribution and classification proposal for clavicular resections

*Fibroma condromixóide de la clavícula. Aporte de caso y propuesta de clasificación para resecciones claviculares*

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### Abstract

The clavicle is an uncommon site for bone tumors; if it is a chondromyxoid fibroma, it is even more so. Social circumstances and alterations in the dynamics of health systems during the COVID-19 pandemic generated a delay in the diagnosis and treatment of many patients. We present the case of a 13-year-old female patient with a large neoplasm located in the left clavicle. After a complete imaging study, two biopsies were performed, diagnosing chondromyxoid fibroma. Radical claviclectomy without reconstruction is planned and performed. The clinical and functional results were excellent. The importance of the case lies in the fact that it corresponds to the tenth clavicular chondromyxoid fibroma reported in literature, and possibly the largest resected.

**Keywords:** clavicle, chondromyxoid fibroma, claviclectomy, classification.

### Resumen

La clavícula es un sitio infrecuente para tumores óseos, si se trata de un fibroma condromixóide, lo es aún más. Circunstancias sociales y alteraciones en las dinámicas de los sistemas de salud durante la pandemia COVID-19 generaron retraso en el diagnóstico y tratamiento de muchos pacientes. Presentamos el caso de paciente femenino de 13 años portadora de una neoplasia de grandes dimensiones localizada en la clavícula izquierda. Previo estudio imagenológico, se realizan dos biopsias que diagnostican fibroma condromixóide; se planea y realiza una claviclectomía radical sin reconstrucción. Los resultados clínicos y funcionales fueron excelentes. La importancia del caso radica en que corresponde al décimo fibroma condromixóide clavicular reportado en la literatura y posiblemente el de mayores dimensiones resecado.

**Palabras clave:** clavícula, fibroma condromixóide, claviclectomía, clasificación.

## Introduction

The clavicle is a very rare site for bone tumors<sup>1,2</sup> and the majority of them are malignant lesions<sup>1,3,4</sup> including metastatic bone disease.<sup>5</sup> The incidence of tumors of the clavicle is reported of less than 1%,<sup>2,4-7</sup>

therefore, surgical experience for the treatment of clavicular tumors is usually limited.

The clavicle is an S-shaped, slender and subcutaneous bone that is connected to the sternal shaft and the acromion, which play an important part in constituting the sternoclavicular and acromioclavicular

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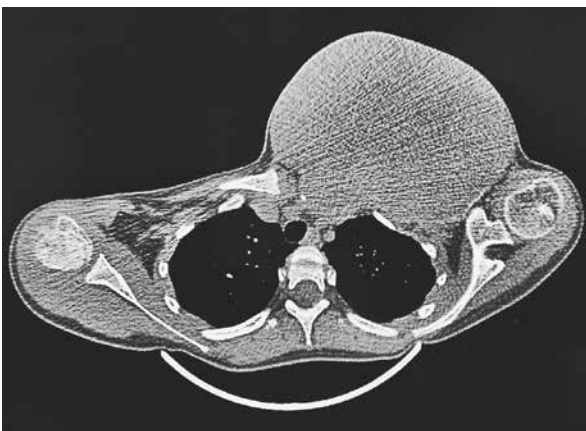
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joints, respectively.<sup>8</sup> The clavicle represents an important element in the anatomical and functional conjunction between the shoulder girdle and the thorax, and consequently with the axial skeleton.

The clavicle is an unusual bone from the perspectives of its embryology, morphology, localization, and function. It is the first ossified bone in the embryo; probably at five months<sup>2,9</sup> and ossifies by membranous ossification.<sup>2,5</sup> Clavicle contains two primary ossifications centers and only one secondary ossification center;<sup>2</sup> it has no significant medullary cavity.<sup>2,5,6</sup> The clavicle has sparse vascular supply with minimal red marrow surrounded by thick cortices of compact bone;<sup>5,6,9</sup> it is the only bone of the long bones whose anatomical position is on the horizontal axis.<sup>2,5,6</sup> Clavicle acts as a link between the upper limbs and the thorax, connecting the axial to the appendicular skeleton<sup>5</sup> and shares its oncological characteristics with flat bones and not with other long bones.<sup>4</sup>

The importance of clavicular tumors and their treatment derive from their particular anatomical location relating to structures of vital importance. Neoplasms of the periclavicular area and the neck, are considered with extracompartmental location;<sup>10</sup> there are no anatomical barriers to tumor extension. Fortunately, the clavicle is a bone that can be resected without causing significant disability.<sup>1</sup>

The most common tumors located at clavicle are: eosinophilic granuloma, bone metastasis and plasma cell myeloma.<sup>6</sup> Other common neoplastic lesions include aneurysmal bone cyst, osteochondroma, Ewing sarcoma, chondrosarcoma and osteosarcoma. Some tumors such as chondrosarcoma, usually occur in sternum



**Figure 1:** This axial tomography projection allows us to observe the size of the neoplasm in relation to the dimensions of the patient's chest.



**Figure 2:** In the coronal tomographic projection it is possible to observe the compression and displacement of the left structures of the neck due to tumor size.

and scapula;<sup>11-14</sup> when they extend extra-articularly, at least one clavicular segment should be resected *en bloc* with the originally affected area. Systematically, chondromyxoid fibroma is not considered as presumptive diagnosis for tumors that involve the clavicle.

Chondromyxoid fibroma has been defined by World Health Organization in 2020 as a benign lobulated cartilaginous neoplasm with a zonal architecture composed of chondroid, myxoid and myofibroblastic areas.<sup>15</sup> This lesion represents less than 1% of all primary bone tumors<sup>16-18</sup> and less than 5% of benign chondrogenic bone tumors.<sup>16</sup>

Chondromyxoid fibroma of the clavicle is exceedingly rare. After an exhaustive targeted search, Arora et al<sup>19</sup> were able to find six cases reported in the literature, and the mention of two more in reviews focused on the diagnosis. As far as we have found, the case contributed by Arora et al<sup>19</sup> is the most recently reported. Under this context, the objective of the present paper is to present the tenth case of chondromyxoid fibroma located in the clavicle, and possibly the largest resected one.<sup>19-27</sup>

### **Case presentation**

We present the case of a 13-year-old female patient referred to the Pediatric Orthopedics Service

of the High Specialty Medical Unit in Traumatology and Orthopedics of the Northeast National Medical Center, belonging to the Mexican Social Security Institute, in Monterrey, Mexico. Upon admission, the patient had a history of pathological fracture of the left clavicle, since this event, progressive growth of a mass with neoplastic characteristics has occurred at the expense of the clavicle; a biopsy diagnosed chondromyxoid fibroma. The late reference to our hospital derived from the alterations generated in the health system by the COVID-19 pandemic situation. Once in the Pediatric Orthopedics Service, her general blood tests and imaging studies are updated.

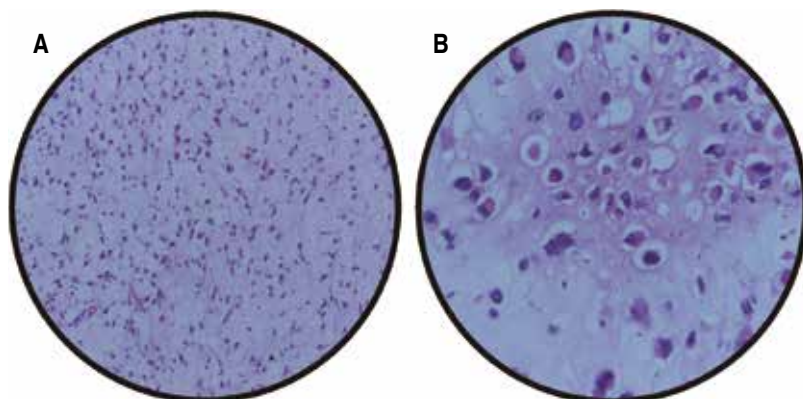
Clinically, a firm mass is observed in the left clavicular area whose dimensions are 16 × 12 × 13 centimeters. An important collateral venous network is evident, and palpation does not generate pain. The scar from the previous open biopsy is located longitudinal to the clavicular axis.

By conventional radiology, the absence of the clavicle was observed with its replacement by a soft tissue mass with neoplastic characteristics. Computer tomography (*Figures 1 and 2*) and magnetic resonance imaging (*Figure 3*) show a neoplastic lesion which replaces the left clavicle and generates compression and vascular displacement. No evidence of sternal and/or acromial involvement is seen. The lesion rests on the first and second left ribs, however tumor involvement of it is not proven due to continuity. Both studies showed tracheal and thyroid shift to the right and the neurovascular structures of the neck (internal and external jugular veins, subclavian vein, common carotid artery and brachial plexus) are shown compressed and displaced. By magnetic resonance imaging, T1-weighted sequences showed isointensity to muscle, and T2-weighted sequences showed heterogeneous hyperintensity (*Figure 3*).

After the complete review of the case, and considering the rarity of the diagnosis and the dimensions of the tumor mass, a second open biopsy is performed continuing the axis of the first. Chondromyxoid fibroma was diagnosed again. Total claviclectomy (radical resection of the clavicle) without reconstruction is then planned, and prior signing of the corresponding informed consent, was performed. The patient's transoperative and postsurgical hemodynamic status was stable. There were no neurovascular sequelae in the immediate postoperative period. The healing process of the surgical wound was carried out without major



**Figure 3:** This sagittal MRI projection reveals the tumor's proximity to vital vascular structures.



**Figure 4:**

**A)** 10×. Mesenchymatous neoplasm in a lax myxoid background, with moderate cellularity and absence of atypia. **B)** 40×. Mononuclear mature chondrocytes with reactive characteristics in a myxoid background.



**Figure 5:** Radiological control after one year of radical resection of the left clavicle. Based on the classification proposed in the presentation of this case, this is a resection I + II + III.

complications. The histological study of the surgical specimen was consistent with that of the two biopsies previously performed (*Figure 4*).

With one year of follow-up, the patient is asymptomatic, without clinical and imaging data of recurrence, and without functional limitation. Being a neoplastic clinical situation, the methodology to evaluate the functional postoperative result was through the Functional Score of the Musculoskeletal Tumor Society (MSTS score)<sup>28</sup> that evaluates six domains scored on a 0 to 5 scale and transformed into an overall score ranging from 0 to 100% with higher score for better results: pain, functional activity, hand positioning, dexterity, lifting ability and emotional acceptance. Personal satisfaction and functional result, were completely satisfactory: 30/30 (*Figure 5*).

## Discussion

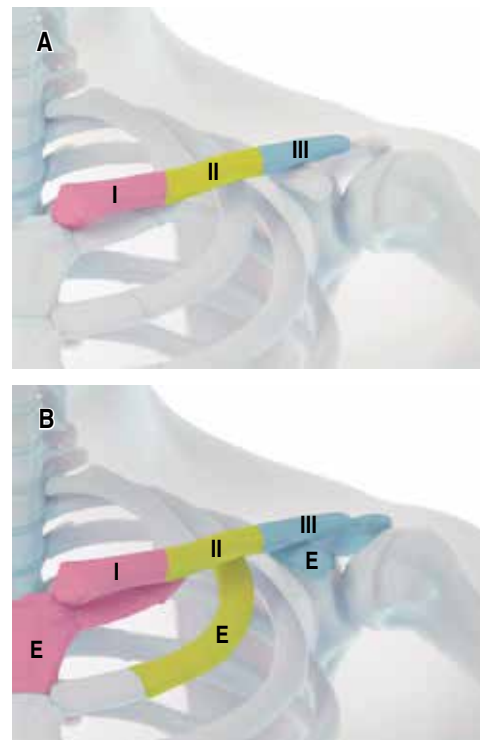
Clavicular tumors may be associated with nonspecific symptoms such as local pain, swelling, or shoulder stiffness. Owing to the rarity of such tumors and lack of specific symptoms, diagnosis can be delayed.<sup>7</sup> Specifically, in the case reported in this paper, social pandemic disruptions played an important role in the dimensions that the neoplasm reached.

Chondromyxoid fibroma can occur at almost any skeletal site. It is most frequent in the long bones, most often the proximal tibia and distal femur; flat

bones are affected in approximately 25% of cases.<sup>15</sup> The differential diagnosis of chondromyxoid fibroma is mainly with chondroblastoma, enchondroma and low-grade chondrosarcoma. Immunohistochemistry and diagnostic molecular pathology are generally not needed for diagnosis. The cartilaginous nature of this tumor is reflected by immunopositivity for S100. In selected cases, identification of the exceedingly specific upregulation of GRM1 expression in chondromyxoid fibroma could be a strong diagnostic adjunct in distinguishing this entity from its mimics.<sup>15</sup>

Clavicular chondromyxoid fibromas are excessively uncommon and practically have been only described as case reports in the literature.

The reported case differs from those found in the literature mainly due to its dimensions and the resulting surgical complexity for its resection. Generally, experience in total claviclectomy is scarce, even in highly specialized centers.



**Figure 6:** The classification used and proposed divides the clavicle into 3 thirds. **A)** The medial third corresponds to segment I, the middle third to segment II, and the lateral third to segment III. **B)** Given the need to extend a resection in any of the segments due to tumor involvement to sternum, rib and/or scapula, the segment or segments must be accompanied by the letter E (Extended).

The clavicle can be divided into three segments: sternal end, middle segment and acromial end. Existing classifications for shoulder girdle resections<sup>29-34</sup> do not satisfactorily consider the clavicle as a leading bone in the shoulder girdle, potentially carrying neoplastic pathology. The clavicle, like the rest of the skeletal structures, can be treated by intralesional or marginal resections, however under certain clinical circumstances including the case that we are contributing to the literature, wide or radical resections are required. Given the low incidence of clavicular tumors and resections due to oncological etiology, as well as considering the articular and non-articular relationships of the clavicle, in the present paper we also propose, for classification purposes, dividing the clavicle into 3 thirds with the possibility of extending the resection extra-articular towards the sternum in the medial third, to rib in the middle third, and / or to scapula in the lateral third. Based in this proposal, in this case the resection performed corresponds to a type I + II + III resection (likewise corresponding to a radical resection of the clavicle under the parameters described by Enneking et al).<sup>10,31</sup> In some cases, given the need to extend towards the sternum, rib and/or scapula, the corresponding segment would be accompanied by the letter E (Extended) (*Figure 6*).

Tumors located in the clavicle can involve the entire clavicle.<sup>2</sup> Total or subtotal excision of the clavicle is rarely associated with a clinically significant loss of function,<sup>1</sup> however many sequels such as shoulder dropping, postoperative pain, limited motion of the shoulder joint, reduction of muscular strength, neurologic function deficit, and restriction of daily activities have been described.<sup>8,35</sup> Fortunately, these complications and sequelae did not occur in the reported case.

Different methods as vascularized fibular graft, bone cement prosthesis and customized 3-D printed titanium prosthesis have been described to reconstruct the clavicle and its anatomical-functional environment, however, and agree with Chen et al (2018)<sup>36</sup> and Atalay et al (2020),<sup>35</sup> we consider that reconstruction after claviculectomy is unnecessary and can be associated with the increase of the incidence of different complications.

## Conclusions

Clavicular tumors are uncommon and chondromyxoid fibroma in the clavicle is exceedingly rare. A late diagnosis makes surgical treatment difficult,

however, clavicular resections are not associated with important functional sequelae. Surgery is still the main treatment modality for primary clavicular tumors and tumorous conditions. In most orthopedic centers, experience in the treatment of clavicular tumors is usually minimal. A classification system for clavicular resections facilitates understanding of the characteristics of a neoplasm, as well as the treatment to be performed. The close anatomic relationship between the clavicle and certain important vascular structures, makes neoplastic resections difficult; fortunately, the clavicle is considered as a dispensable bone. In general terms, the experience in the treatment of clavicular tumors and partial, total and extended claviculectomy, is very scarce.

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## Conflicts of interests

The authors declare that they have no conflicts of interest.