

A RARE CLAVICULAR DESMOPLASTIC FIBROMA ACCOMMODATION: CASE REPORT

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Article Info

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Abstract

Desmoplastic fibroma is a benign, rare, locally aggressive and non-metastatic myofibroblastic tumor. It affects more young patients between 20 and 30 years, in whom it usually presents high morbidity, causing pain and local edema. The objective was to report a rare case of involvement in an elderly man, with a complaint of bone prominence in the right clavicle, associated with pain and pruritus. The diagnosis was made from the anatomopathological exam with immunohistochemical study and the treatment was the excision of the affected clavicle. Even in view of the current arsenal, it is still a diagnostic challenge and the evidence regarding the most efficient surgical treatment is controversial, requiring further studies about this subject.

Keywords: Desmoplastic Fibroma. Bone Neoplasms. Neoplasms. Clavicle. Thoracic surgery.

Introduction:

With few reports in the literature, desmoplastic fibroma is a benign, rare and locally aggressive myofibroblastic tumor, and presents itself as a differential diagnosis in both benign and malignant cases¹. It can occur in any age group; however, it is more frequently observed in the second or third decades of life².

Tumors affecting the clavicle are rare, especially desmoplastic fibroma, with only two cases described in the literature before 1985. Its incidence among primary bone tumors is 0.1 to 0.3%, affecting mainly the mandible (22%), bones pelvis (13%) and long bones. Only 0.45 to 1.01% of bone tumors are present in the clavicle^{3,4}. Morbidity is higher when it comes to younger patients⁵ and, despite conflicting data, there seems to be no predilection for sex⁴.

The clinic is usually indolent, with nonspecific symptoms and the pathology has a locally aggressive pattern, but not metastatic^{3,6}. Pain complaints and local edema are the most frequent symptoms, and are related to the locoregional growth of the tumor⁸. The radiological pattern of the lesion assists in propaedeutics, with well-defined radiographic signs, non-sclerotic, geographically marginalized, with internal pseudotrabeulation and bone expansion. In magnetic resonance imaging, a fibrous tissue pattern in T2 is expected and a hypodense or isodense signal in T1, compared to muscle structure. The

diagnosis, however, can only be confirmed through histopathology. We report here a rare case of clavicular involvement by desmoplastic fibroma in an elderly patient.

Clinical Case:

Male patient, 65 years old, type 2 diabetic and hypertensive under drug treatment, social drinker. Previous history of left colectomy due to colon cancer, without the need for adjuvant treatment. He presented with a history of bone prominence in the right clavicular manubrium joint for about three months, associated with moderate pain and occasional itching. He denied any other symptoms. Preserved upper limb sensitivity and mobility. Computed tomography scan of the chest (Figure 1) showed a focus with the presence of air density at the sternal end of the right clavicle.

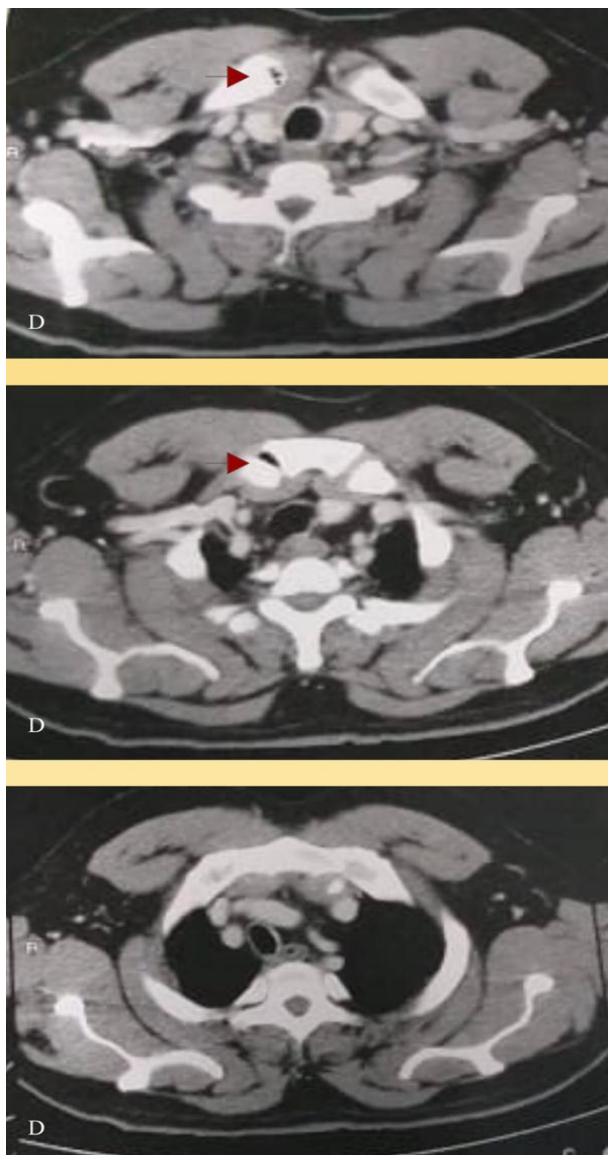
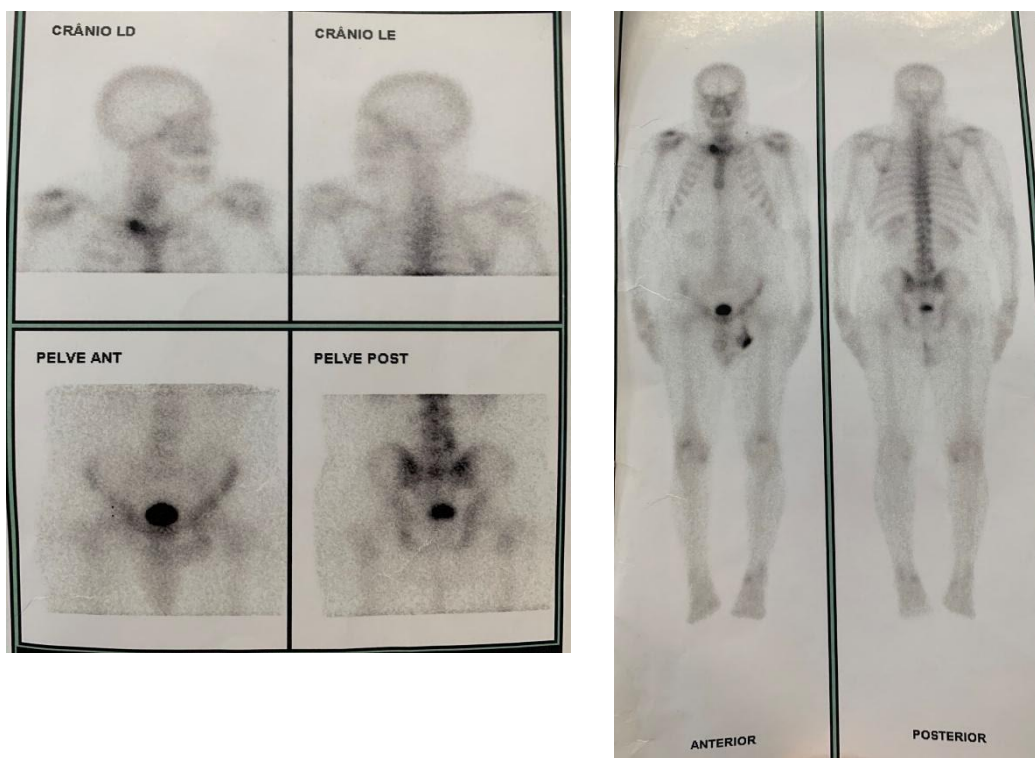


Figure 1. Chest tomography images.

Bone scintigraphy (Figures 2 and 3) with an area of increased osteogenesis to the right of the sternal manubrium, suggesting an osteoarticular process.



Figures 2 and 3. Bone scintigraphy images.

He underwent a biopsy of a fragment of the proximal part of the clavicle, with a pathological examination showing low-grade osteo-cartilaginous mesenchymal neoplasia. Immunohistochemical study indicated primary bone lesion, including desmoplastic fibroma (Figure 4).

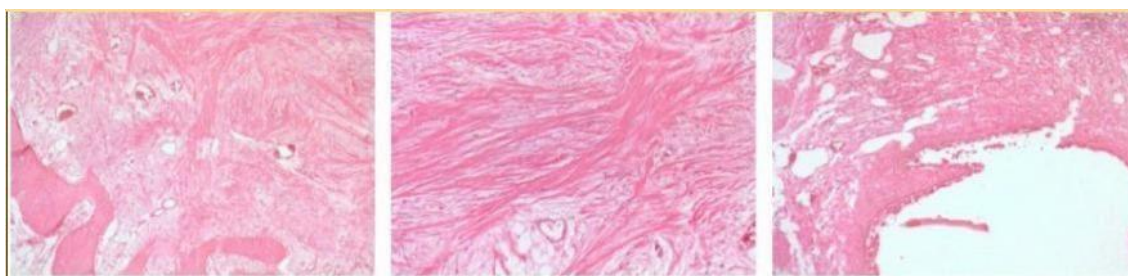


Figure 4. Optical Microscopy - HE staining.

We then chose to total the removal of the right clavicle. The procedure was uneventful, with the patient being discharged on the 5th postoperative day and maintaining asymptomatic outpatient follow-up and with no signs of locoregional recurrence.

Discussion:

Desmoplastic fibroma should be considered in the differential diagnosis of a locally aggressive bone lesion, also including reactive and neoplastic fibroblastic lesions. As a differential diagnosis, rhabdomyosarcoma, fibrosarcoma, giant cell tumor, among others should be considered^{4,9}. Despite the radiographic signs, the lesions are not easily differentiated from other benign or malignant tumors. However, T2-weighted MRI with a predominant pattern of osteolytic lesions makes the diagnosis plausible³.

Zlotecki divided the tumors according to the type of manifestation: marginal or central. The marginal is the most frequently seen as modification of the cortex, compression and absorption, with soft tissue

mass. In relation to the central manifestation, a local osteolytic aspect is observed, which may be associated with a periosteal reaction⁵.

The diagnosis is made only with biopsy and histopathology, although imaging methods assist in the delimitation. It is characterized by spindle cells without atypia and pleomorphism, in addition to areas with fibrous acellular connective tissue, with increased cellularity correlated with recurrence⁶. The immunohistochemical study is negative for s100 and MAS, positive for vimentin, with Ki-67 less than 5%³.

According to Lorenzo et al¹¹, the choice of the type of surgical approach in this type of injury is a source of controversy. Many surgeons prefer curettage of the lesion, while others opt for excision of the lesion with free margins, sometimes including the use of bone graft. In the case in question, surgical treatment was chosen, with complete excision of the lesion, which is for diagnostic and therapeutic purposes. In these cases, oncological excision is performed, and recurrence is common if total resection is not possible due to the locally invasive pattern⁴. The recurrence rate is approximately 17%. Some factors such as genetic predisposition, estrogenic stimulus and trauma can be associated with tumor genesis, but more studies are needed^{5,6}.

Conclusion:

Desmoplastic fibroma represents a rare pathology that makes a differential diagnosis with both benign and malignant lesions, thus representing a diagnostic challenge. The role of the imaging study is to act as a guide in clinical / surgical management, bearing in mind that benign pathologies can also behave extremely aggressive behavior simulating malignant conditions, therefore it is essential to open the range of diagnostic possibilities including them in the differential diagnosis. The present study highlights the rarity of clavicular involvement of this pathology and reinforces the need for further studies on the disease in question.

References:

- [1.] Shekhar MG, Reddy RS, Ravikanth M, Lavanya R, Kumar M. Desmoplastic fibroma of the mandible: case report and review of literature. *Prim Dent Care*. 2011;18(3):115-118.
- [2.] COELHO, Henriques et al . Fibroblastoma desmoplásico envolvendo a artéria braquial: um caso clínico único. *Angiol Cir Vasc*, Lisboa , v. 13, n. 4, p. 46-49, dic. 2017 .
- [3.] KANG, DM, Juhng SK, Sohn YJ, Kim HS. Imaging findings of desmoplastic fibroma rarely involving the clavicle: case report. *Korean J Radiol*. 2014;15(1):130-133. doi:10.3348/kjr.2014.15.1.130
- [4.] RIBEIRO, Bruno Niemeyer de Freitas et al . Fibroma desmoplásico com disseminação perineural: achados nas sequências convencionais de ressonância magnética e na difusão. *Radiol Bras*, São Paulo , v. 48, n. 4, p. 266-267, Aug. 2015 . <http://dx.doi.org/10.1590/0100-3984.2014.0135>.
- [5.] Xu, Yongming et al. "Desmoplastic fibroma of the femur with atypical image findings: A case report." *Medicine* vol. 97,52 (0): e13787. doi:10.1097/MD.00000000000013787
- [6.] LICEAGA ESCALERA, C. et al . Desmoplastic fibroma, report of a case and a literature review. *Rev Esp Cirug Oral y Maxilofac*, Barcelona , v. 32, n. 1, p. 21-24, Mar. 2010 . DOI: 10.4321/S1130-05582010000100005
- [7.] CAZAL, Claudia et al . Collagenous fibroma (desmoplastic fibroblastoma) of alveolar bone: a case report. *J. Bras. Patol. Med. Lab.*, Rio de Janeiro , v. 41, n. 3, p. 185-188, June 2005 .
- [8.] <http://dx.doi.org/10.1590/S1676-24442005000300008>.
- [9.] GREWAL, Reetu et al. "Desmoplastic fibroblastoma of the left upper arm." *BMJ case reports* vol. 2018 bcr2017221738. 26 Jan. 2018, doi:10.1136/bcr-2017-221738
- [10.] BRASILEIRO JUNIOR, Vilson Lacerda et al . Pediatric fibromatosis involving mandible: case report and a five-year post-operative follow-up. *J. Bras. Patol. Med. Lab.*, Rio de Janeiro , v. 49, n. 3, p. 208-211, June 2013 .
- [11.]