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Radiology

Atypical Desmoid Tumor, A Case Report

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Abstract **Case Report**

Desmoid tumors are rare and benign tumors of soft tissues, characterized by local invasion and high tendency to recur despite surgical resection, we study the case of a 19-year-old patient admitted for cervico-facial and parieto-costal swellings, without any significant family history, and we study the CT and magnetic resonance imaging aspects, with an unpredictable and recurrent evolution despite surgical excision, with many studies that affirm the hormonal factor with the progression of the swelling, as well as its decrease in size after menopause for the treatment, there is no consent using the different components, surgery, chemo-radiotherapy with different and uncodified effectiveness.

Keywords: Desmoid tumor, Cervico-facial swelling, Parieto-costal swelling, Hormonal factor, Recurrence after surgical excision.

INTRODUCTION

Desmoid tumors are benign soft tissue tumors characterized by local invasion and a high tendency to recur despite surgical resection. Through the experience of our department, we will analyze the results of the radiological appearance on CT and MRI.

OBSERVATION

This is a 19-year-old patient, admitted for cervical-facial and parieto-costal swellings at the dorsal level, of soft consistency, not painful (Figure I) with a feeling of heaviness and weight loss, evolving for 4 years, the patient comes from a consanguineous marriage of the 1st degree without any similar case in the family.

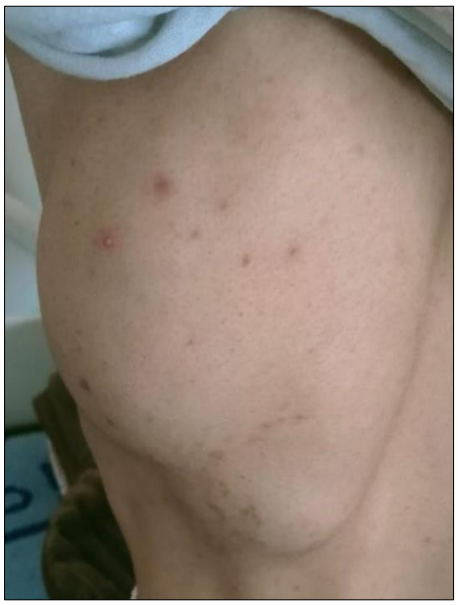

Figure I: Image A: Left parieto-costal swelling

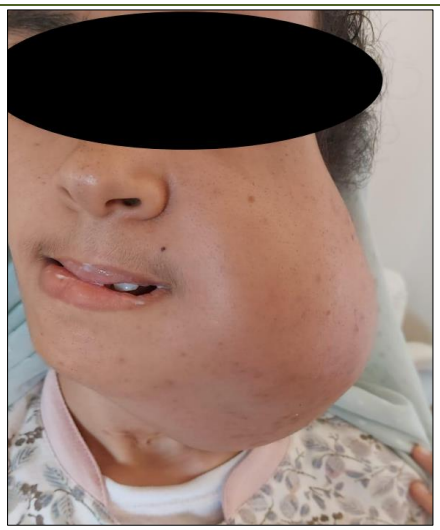

Figure I: Image B-1: Left cervicofacial swelling

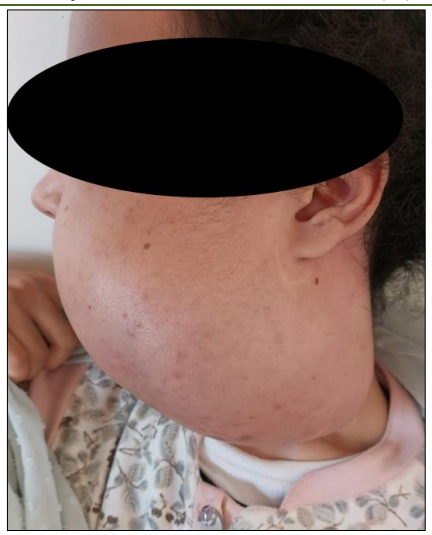

Figure I: Image B-2: Left cervicofacial swelling

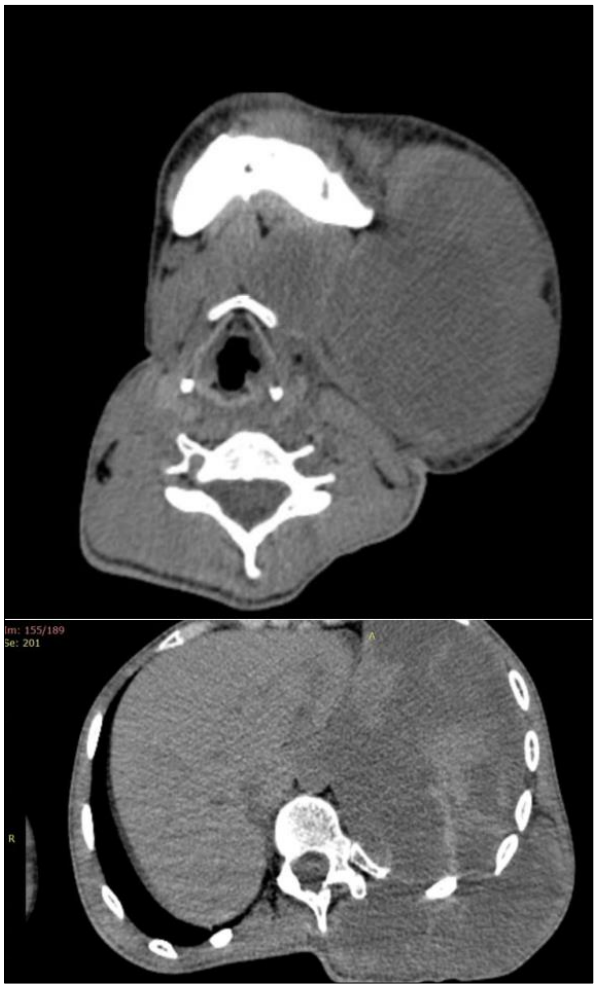

Figure II: CT image of a left cervico-facial and parieto-dorsal lesion formation of tissue density, well-defined and compartmentalized, heterogeneous site of areas of necrosis

DISCUSSION

Desmoid tumors (DT) are deep fibromatoses; benign but locally aggressive tumors; consisting of a proliferation of differentiated myofibroblasts within a loose collagen matrix. They develop from the connective tissue of fascia, aponeuroses and intermuscular septa of striated muscles. These tumors represent less than 3% of soft tissue tumors. Their exact pathogenesis is not known, but traumatic, endocrine and genetic factors have been incriminated [1].

CT scan and MRI are indicated for diagnosis and follow-up. Imaging allows to determine the extent of the tumor, the infiltrated organs and the plan of a possible surgical resection, a cervico-facial and thoracic CT scan was performed without and after injection of PDC objectifying locally infiltrating left thoracic and left latero-cervical masses, centered on the ascending branch of the mandible which is lysed in places with macro lobulated contours, well limited, heterogeneous hypodense seat of calcifications and fine septa discreetly enhanced after injection of PDC, (Figure II) this mass is related to:

- **Inside:** With the deep spaces of the face.
- **Outside:** It pushes back and lifts the skin plane.
- **Up:** it infiltrates the homolateral infratemporal fossa.
- **Down:** It pushes back the oral floor.
- **Back:** It pushes back the left IJV and common carotid It also extends to the prevertebral space and left paravertebral muscles
- **Ahead:** it comes into contact with the posterolateral wall of the left maxillary sinus.

For histology, the patient underwent a total resection of the first left latero-cervical mass, revealing an appearance of cells with an elongated and fusiform nucleus associated with rare mitoses and absence of tumor necrosis. These are elements in favor of a desmoid tumor and help to distinguish it from a sarcoma. In addition, immunohistochemical analysis showed that the tumor cells reacted to anti-smooth muscle actin and did not react to S-100 protein and MIB-1 (Ki67), which are excellent markers of the proliferative activity of sarcomas, which is in favor of a desmoid tumor.

On MRI On T1-weighted images, desmoid tumors are hypointense or isointense to muscles, while on T2-weighted images, they are hyperintense. With gadolinium contrast, the desmoid tumor shows moderate enhancement with hypointense bands reflecting collagen bundles.

The Evolution

Is unpredictable and characterized within two years of diagnosis by spontaneous regression (30-50%) (after menopause), local progression (~20-30%) or stability in the size of the lesions (~30%) [2].

CONCLUSION

- Very rare.
- Location: mandibular, monofocal.
- Slow evolution, and very recurrent for extra-abdominal locations.
- The diagnosis is radio-histological ++
- Treatment: sensitivity to chemo-radiotherapy varies from one patient to another, however surgical treatment has proven to be more effective:
  - If tumor is resectable and asymptomatic: monitoring for small tumors
  - If tumor is resectable and symptomatic: surgery
  - If the tumor is unresectable and recurrent (which is the case for our patient): Surgery and radiotherapy.

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