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Fibromatosis desmoid of scapula in children (About a rare and aggressive disease)

Driss Hanine[1*], Zakaria Aboulam[2], Jaouad Bouljrouf[3], Mounir Kisra[4]
[1-4] Visceral Pediatric Surgery Department « A » - Children Hospital of Rabat, Faculty of Medicine of Rabat, Morocco

Abstract
Desmoid tumors are non-metastatic mesenchymal tumors with an aggressive local growth. Depending on the anatomic location, morbidity varies.
The successful treatment of desmoid tumors requires an interdisciplinary approach.
There is a need for prospective randomized studies with larger numbers of patients to allow the development of standardized and evidence based treatment algorithms.
We report of a child with a desmoid tumor of the left shoulder which was treated in our department by surgical excision.

Keywords: desmoid, desmoplastic tumors, fibromatosis, shoulder mass

Introduction
Called Desmoid-type fibromatosis or Desmoplastic Fibroma; It is a tumor with fibroblastic and my fibroblastic cells without malignancy, locally very aggressive but not giving metastases.
It is a rare tumor, having the character of benignity seen the absence of metastases, but with a major local aggressiveness.
It exists in two forms:
- Abdominal and retroperitoneal, and
- Extra-abdominal (The object of our study).

Materials & Methods
We report in our study, made at the pediatric visceral surgery department "A" at the children's hospital of Rabat, the case of a 12-year-old girl with no particular antecedents who consults for a retro axillary mass, discovered during minimal trauma since 2 months before admission, increasing rapidly in volume.
The clinical examination showed a girl in good general condition, not yet menarche, having a left back, axillary mass, hard, 4cm, painful on palpation, mobile on the superficial plane and fixed on the deep plane.
- A standard radiograph was performed (Figure 1) objecting infiltration of the soft parts probably, without visible bone involvement.
- An ultrasound performed afterwards which shows a mass of tumor appearance interesting mainly the left subscapular muscle, lumbar axillary adenopathies and cervical bilateral infracentimétriques.
- A biological assessment made of NFS, VS and CRP income without particularities.

A CT scan of the left shoulder was requested later showing (Figure 2)
A tissue mass of the soft tissues of the anterior aspect of the scapula, interesting the sub-scapular muscle, hypoechoic with discrete contrast enhancement within it and heterogeneously, measuring 74x33 mm.
- No clear limits with the deltoid muscle and the small round.
- Upwards goes up to the humeral head.
- Inwardly pushes the subclavian pedicle with compressed aspect of the subclavian artery.
- Cervical ADP bilateral and axillary ipsilateral.

An MRI of the left axillary region was performed (Figure 3) objectivizing
- A lesion process of the axillary region of the left signal tissue hypo signal T1, hyper signal T2 and diffusion enhancing strongly after injection and having no greasy component.
- This process grows astride the tip of the scapula and invades the subscapularis muscle forward and infraspinatus backwards.
- Measures 77x38x38 mm.
- There is a loss of segmental continuity of the cortical bone of the body of the scapula.
- It encompasses several vascular structures that remain permeable.
- It is flush with the axillary pedicle with intimate contact with the vein.

In front of this clinico-radiological table we evoked the diagnosis of Ewing's sarcoma or Rhabdomyosarcoma.
An initial biopsy was performed, having been inconclusive, reverting to fibro-inflammatory remodeling with signs of muscular dystrophy.
A second biopsy returned in favor of a fusiform cell tumor with an aggressive appearance since it encompasses the muscular and nerve structures at the periphery. An immunohistochemically complement eliminated rhabdomyosarcoma and retained the diagnosis of desmoid fibromatosis.
The patient received chemotherapy without visible improvement with increased tumor mass and increased pain. We opted for a surgical treatment, certainly mutilating but in front of the strong demand of the parents. Surgical exploration revealed an enormous mass composed of 2 parts: an easily

resected retro scapular part (Figure 4) and a prescapular part which is voluminous and sticky to the axillary pedicle inside and to the pedicle under clavicle at the top, this part is arriving up to the clavicle with a good dissection plan, which helped us to resect the entire mass with healthy margins (Figure 5).
A muscle reconstruction is then performed to fill the residual cavity (Figure 6).

The pathological study confirmed the diagnosis.
The immediate postoperative evolution is good. In the short term, we note the disappearance of pain and thus a resumption of physical activity.
With a follow-up of one year, one does not note of recurrence, the patient is always followed in search of a possible recurrence, something which is very frequent in this pathology.

Discussion
Fibromatosis Desmoid Tumor is a rare tumor affecting 2 to 4 / 1,000,000 inhabitants. It affects all ages but is especially seen between puberty and 40 years with a peak between 25 and 35 years. It predominates in women (3/1) [1].
The preferential localizations of extra-abdominal desmoid tumors are [1, 2]:

- Lower limb (31-37%), particularly thigh
- Upper limb (23-28%)
- Chest wall and para-vertebral space (17-22%)
- Head and neck (10-23%)

Its seat is deep, subaponeurotic, in contrast to superficial plantar and palmar fibromatosis.
It can present as multiple lesions (5 to 15% of cases), or limited to a single limb in
75 to 100% of cases [3, 4].
The tumor is often connected to a fascia or other oriented fibrous structure stretched between two fixed points. This structure is perhaps the site of origin of the proliferation.
Pathologically, the lesion is composed of long and divergent bundles of monomorphic fusiform cells, with no cellular atypia or hyperchromatism, with a pale nucleus containing 1-3 small nucleoli, within a collagenous stroma of corrugated fibers. and containing a variable number of vessels, sometimes with perivascular edema [2].
At the cytogenetic level, activating mutations of the β-catenin gene (CTNN B1) are present in 85% of sporadic cases. These lead to the intra-nuclear accumulation of the β-catenin protein and result in immunohistochemistry by nuclear staining of the tumor cells (in addition to cytoplasmic staining). Since this immunohistochemistry is difficult to develop, the detection of the mutation is an aid to the diagnosis of certainty [5].
In terms of imaging, the tumor is usually intermuscular and can therefore be surrounded by a split fat sign as a nerve tumor, when it does not invade the surrounding muscles [1].
It measures between 5 and 10 cm, most often [6].
It is a mass of often multinodular shape with typically starry boundaries.

- In ultrasound, the mass is hypoechoic, well or poorly limited, often with marked shadow cone, without vascularization in 66% of cases [7].
  The "fascial tail sign" and a "stag antler" appearance corresponding to branched stellate tumor extensions in the subcutaneous fat (unrelated to the appearance of branched vessels made of "antler" of the solitary fibrous tumor), may be visible on ultrasound [8].
- In scanner, its density is close to that of the muscle or a little higher [6].
- In MRI, its limits can be sharp (49 to 54%) or fuzzy (46 to 51%), especially in young subjects [1].
  Its MRI signal is of variable intensity, heterogeneous most often.

In T1, the signal is close to that of muscles in 83 to 95% of cases [1, 9, 10].
In T2 without fat saturation, the signal is close to that of fat in 46 to 77% of cases but can also be predominantly hyposignal (24 to 31%) or, conversely, in hypersignal T2 (30%) [1, 10, 11].
In T2 with fat saturation, the tumor is predominantly hypersignal moderate with hyposignal ranges.
Moderate to severe contrast enhancement after gadolinium injection is present in 90% of cases [9].
T1 and T2 hypointense zones in bands, with little or no contrast, are present in 62 to 91% of cases, and would be the most suggestive sign of desmoid tumor [1].

The MRI signal also varies with the age and composition of the tumor

- Moderately moderate T1 hypointense and T2 hypersignal in young, predominantly cellular tumors
- Rather marked hyposignal T1 marked and partial hyposignal T2 partial, more or less extended, in tumors aged, rich in collagen

The MRI signal does not quantify the risk of recurrence [12].
Upon initial discovery, the tumor may have either sharp or invasive boundaries (about 50% of cases) [9].
Infiltration of neighboring muscles with stellate extensions is characteristic of this tumor.
Fascial tail extensions (83% of cases) are a very characteristic sign that is shared only by certain fibrous tumors [6, 9, 10].
Erosion of the adjacent bone is present in 6 to 37% of cases [6].
The histologically differential diagnosis may involve scar tissue, nodular fasciitis, low-grade fibromyxoid sarcoma.
Therapeutically; given the risk of recurrence, the often mutilating nature of repeated excisions and the tendency for spontaneous stabilization of tumor growth after a growth phase, the current trend is to refrain from treatment.
Excision of the tumor may, however, be indicated when the tumor can be removed completely without mutilation.
On the other hand, if the excision is technically difficult or uncomfortable, simple surveillance or medical treatment is first used, with stabilization in 86% of cases [13-15].
Recent trials show that anti-angiogenic tyrosine kinase inhibitors (sorafenib, sunitinib) and anti-platelet growth factor (anti-PDGF) can stabilize or regress desmoid tumors [16, 17].

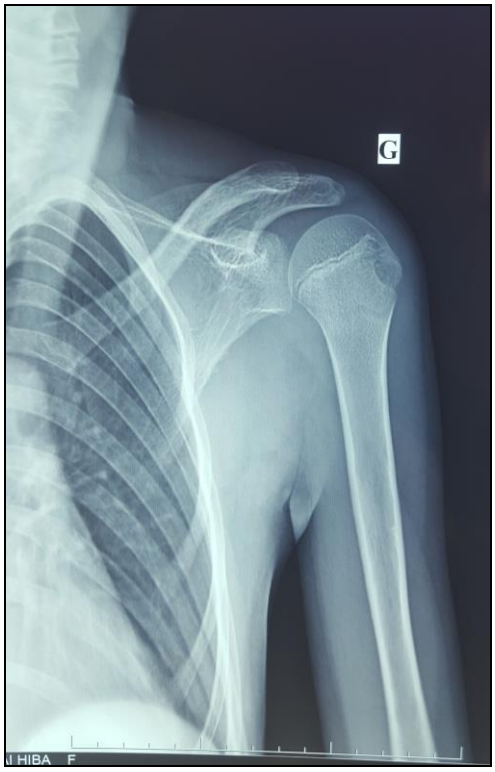

Fig 1: Standard X-Ray of the shoulder showing no bone involvement.

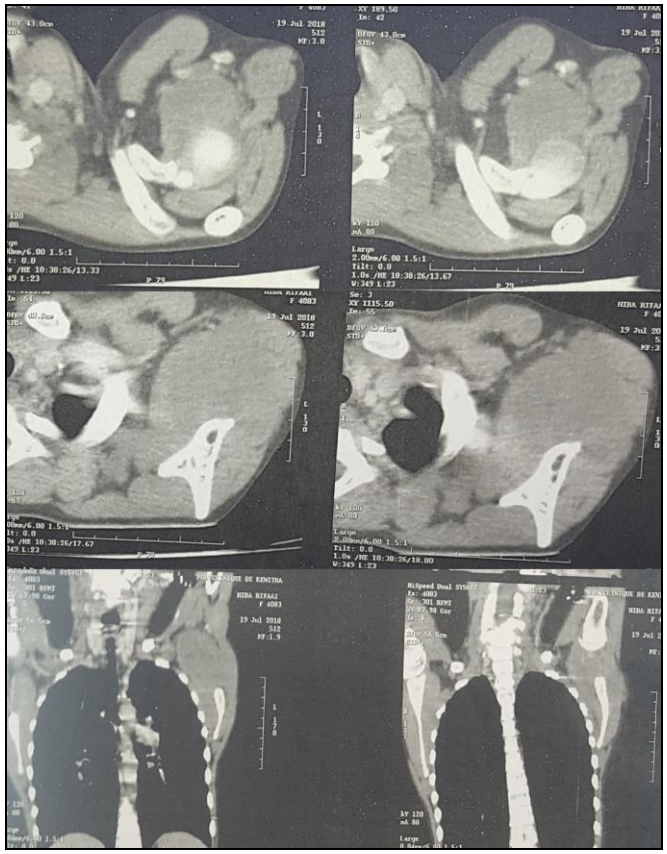

Fig 2: CT scan image of the left shoulder showing the process.

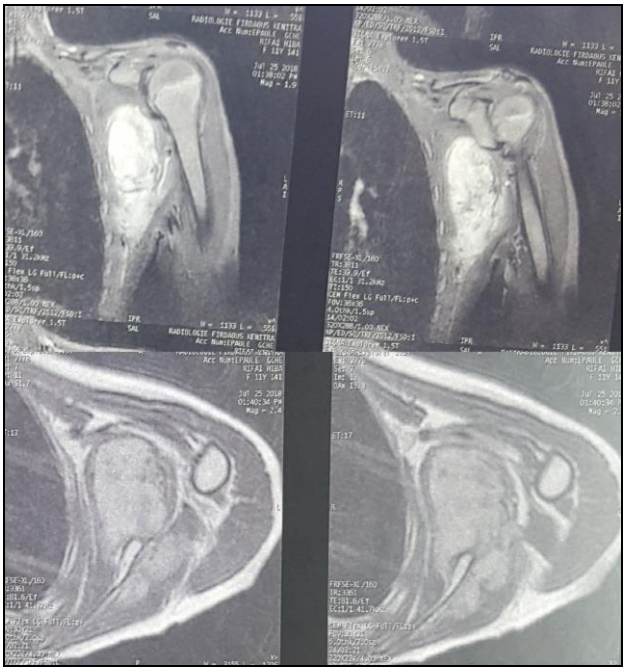

Fig 3: MRI of the left axillary region showing the tumoral process.

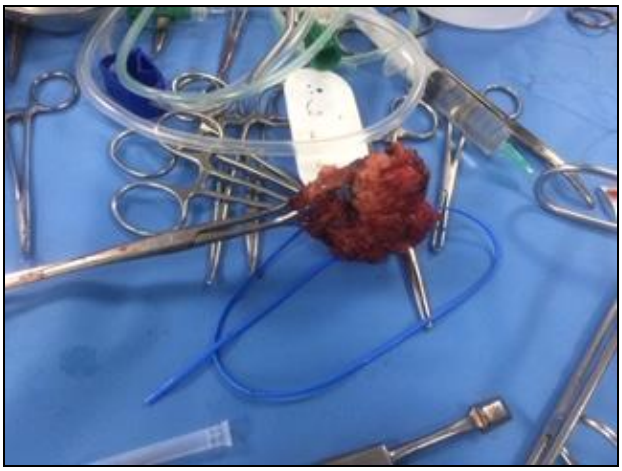

Fig 4: Peroperative image showing the resected retroscapular part.

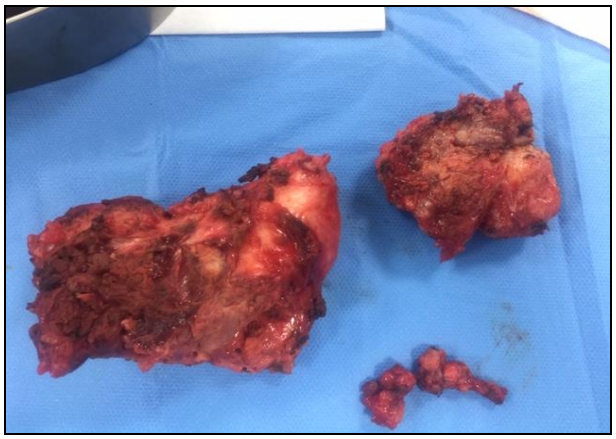

Fig 5: Image showing the entire mass resected.

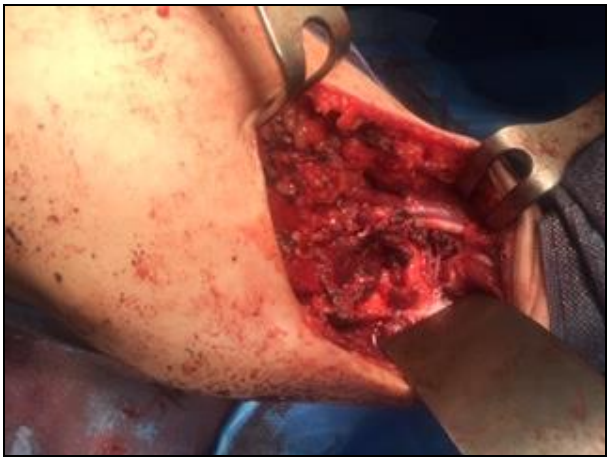

Fig 6: Preoperative image showing the residual cavity.

Conclusion

It is a rare tumor, very aggressive locally but without distant metastases.

There is no current consensus or consistent therapeutic attitude.

The surgical management is very mutilating with a major recurrence rate and possibility of several surgical times.

Conflict of interest

All authors declare that they have no conflict of interest.

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