Abstract: Nodular fasciitis is a benign proliferative spindle-cell lesion that presents as a rapidly growing mass frequently being mistaken for a sarcoma. We report in this observation a localization of this tumor in the angulomandibular region, which was problematic to us from the clinical and radiological diagnostic aspects. The biopsy or biopsy-excision must allow to respect the neighboring structures (vessels, nerves and muscular compartments). It is a benign lesion, which does not recur and can regress spontaneously.

Keywords: Nodular fasciitis, benign lesion, fibromatous pseudosarcoma, biopsy.

INTRODUCTION

Nodular fasciitis (FN) is a benign lesion with rapid proliferation of myofibroblastic cells, which develops at the expense of a muscular fascia within the subcutaneous tissue.

The usual locations are the torso (48%), the upper limb (20%) and 20% of the lesions are present in the cervicofacial region. The importance of this lesion comes from the fact that it can be confused with a sarcoma due to its rapid evolution, rich cellularity and mitotic activity.

We report in this observation a localization of this tumor in the angulomandibular region, which was problematic to us from the clinical and radiological diagnostic aspects.

CASE REPORT

A 3-year-old girl presented with a rapid onset tumefaction under the right mandibular-angle, evolving for 6 months.

On examination, a mass of approximately 5 cm in diameter was palpable at the mandibular-angle region. The lesion was firm, very mobile and could easily move under the labial mucosa. The clinical examination showed a mass that was not painful, well limited, firm, fixed, developed at the expense of the mandible (Fig: 1).

There was no loco-regional lymphadenopathy, the patient was afebrile. There were no biological inflammatory signs (leukocyte 6060 / mm and CRP 3mg/L).

Fig-1: tumefaction under the right mandibular-angle
Cervico-facial CT Scan was in favor of a right lateral cervical tumor mass under the right mandibular-angle with malignant mandibular bone involvement.

Surgical excision was performed, showing a richly vascularized tumor (Fig: 2) Macroscopic examination showed white, homogeneous, soft, indurated, non-encapsulated and poorly limited lesion.

**Fig-2: Tumor excision of a richly vascularized mass**

Incision 4 cm long, parallel to the basilar rim of the mandible. Two fingers across under the basilar rim in a cervical fold.

Detachment was done through the subcutaneous tissue to visualize the tetragonus muscle of the neck which is then incised (Fig: 3).

**Fig-3: Aspect after removal of the tumor**

**DISCUSSION**

Nodular fasciitis of the head and neck is rare in adults, but more common in young people. The cervicofacial region is by far the most affected area in children.

This tumor was first described as a fibromatous pseudosarcoma by Konwaler and al. in 1955 [12]. The etiology of the FN remains unknown. However, the literature frequently reports local inflammatory lesions or repeated trauma that may be responsible for this tumorous process [1,14,15].

The difficulty of diagnosis lies in the elimination of many differential diagnoses and especially in the assertion of the benign nature of the lesion. From a clinical point of view, the rapid evolution of the FN (from a few days to a few weeks) should alert us to a malignant process [1,14,15].

Three types can be differentiated:
- The subcutaneous type, corresponding to a well-defined spherical nodule attached to the fascia;
- The intramuscular type, generally larger than the subcutaneous form;
- The fascial type, centered by the fascia, often less well limited and developing along the interlobular septa, giving it a stellar appearance [16].

A paraclinical diagnosis procedure must be undertaken especially with the aid of an MRI despite the absence of systemic signs. The diagnosis of these myofibroblastic and fibroblastic tumors is difficult, in particular the recognition of benign or malignant character, due to the frequent absence of specific immune-histochemical markers. Diagnosis is most often based on a detailed morphological analysis interpreted according to the clinical context. The FN is a pseudosarcoma and most often a well-limited and non-encapsulated lesion of less than 3 cm.

Rare dermal cases have been reported. Under microscopy, the proliferation consists of myofibroblasts of increased size, immature appearance, resembling to fibroblasts in culture, arranged in short and irregular bundles. Fusiform cells have frequent mitoses without atypical form. Many radiated blood vessels leave the hemorrhagic center, forming a zone phenomenon explaining a richly vascularized appearance on ultrasound and the enhancement of the tumor by the MRI contrast medium. In immunohistochemistry, fusiform cells express smooth muscle actin. The Desmin, S100 protein and cytokeratins are not expressed. A recent study has shown an expression of HMGA2 (High Mobility Group A2) in 90% of nodular fasciitis [17]. This immunostaining appears as an aid to the differential diagnosis.
This benign lesion, which does not relapse (even in cases of incomplete excision), may regress spontaneously as Shimizu and al. in a series of 250 FN cases, and 150 Kijima and al. with a decline of eight years on these excised FN [2,14]. In case of recurrence, the diagnosis must be reviewed [11,8].

FN can be confused with many soft tissue lesions, benign or malignant. Thus, it is the benign mesenchymal lesion most often confused with sarcoma, because of rapid growth, high cellularity and frequent mitosis.

CONCLUSION

FN is a benign lesion, which is important not to be confused with sarcoma to avoid radical surgical treatment. Immunohistochemistry and biology can provide diagnostic assistance. Ultrasound can determine the nature of the lesion and its location in relation to the fascia, while MRI should be performed in case of suspicious lesions. The biopsy or biopsy- excision must allow to respect the neighboring structures (vessels, nerves and muscular compartments). It is a benign lesion, which does not recur and can regress spontaneously.

Contributions of the authors

All the authors contributed to the medical care of the patient, as well as writing this article they approved.

REFERENCES