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RESEARCH ARTICLE

A SUPRACLAVICULAR SWELLING REVEALING A FIBROUS DYSPLASIA

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Abstract

Fibrous dysplasia is a rare bone disease. It accounts for about 2.5% of bone diseases and 7% of bone tumors. We report the case of a 9years old girl admitted for management of a painful right supraclavicular swelling evolving in a context of conservation of general status and in whom the paraclinical assessment was in favor of fibrous dysplasia of the clavicle.

Introduction:-

Fibrous dysplasia or Jaffe-Lichtenstein disease is a sporadic condition, linked to a defect in the maturation of osteoblasts. It is associated with an abnormally high osteoclastic activity, leading to osteolysis and extension of the disease. We report a rare case of fibrous dysplasia of the clavicle.

Case report

Child K.H., 9 years old girl, who has had reoccurring pain in the right shoulder and fatiguefor 2 years, withsubsequent appearance of a right supraclavicular mass of hard consistency, gradually increasing in volume.

X-ray of both clavicles showed lytic lesion of the middle third of the right clavicle with a thinned aspect of the cortex.(Figure 1).

The scannographic complement showed an osteolytic process of the body and the medial extremity of the right clavicle evoking first an Ewing's sarcoma. (Figure 2). And the MRI was in favor of a suspicious right clavicular lesional process without invasion of the sternoclavicular or humeral joint, and no other visible bone localization. Associated cervical lymphadenopathy. (Figure 3)

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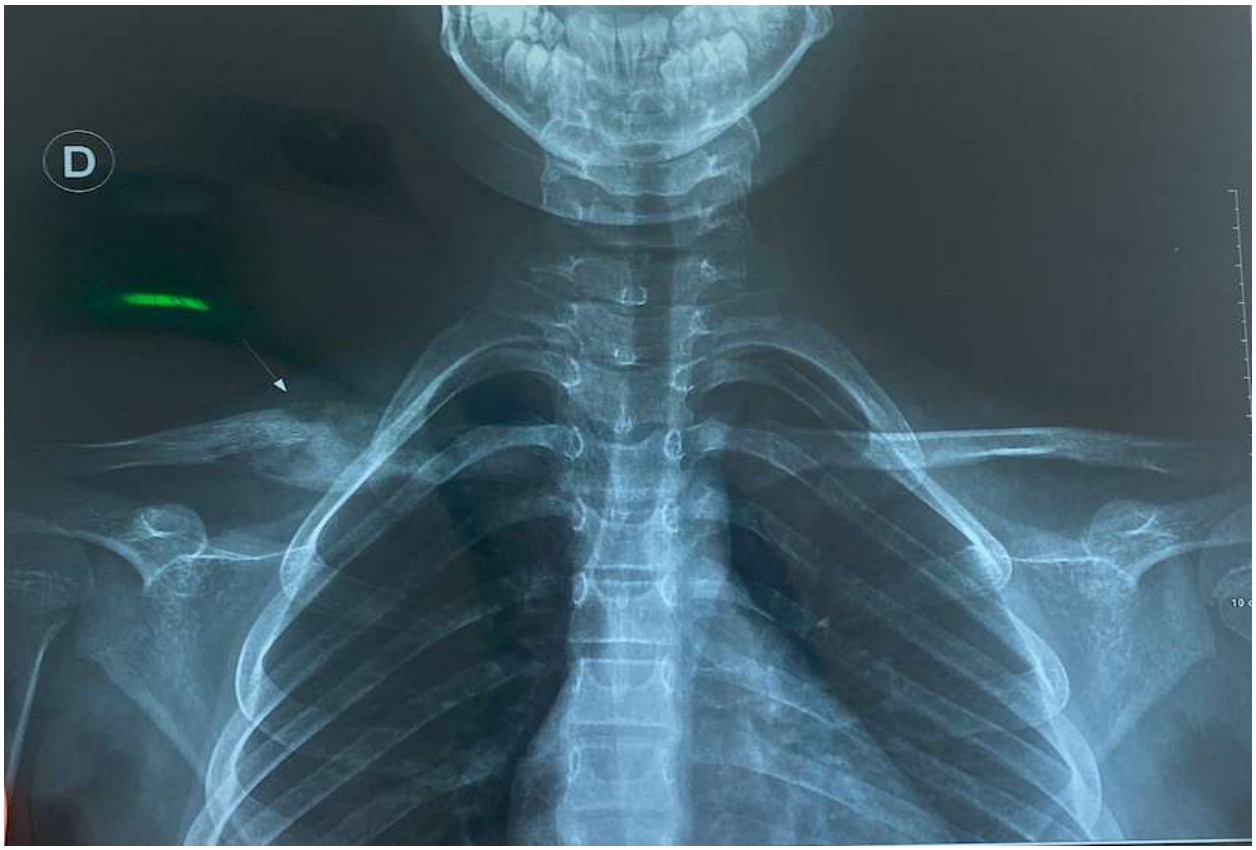

Figure 1:- X-ray showing the osteolytic process of the clavicle.

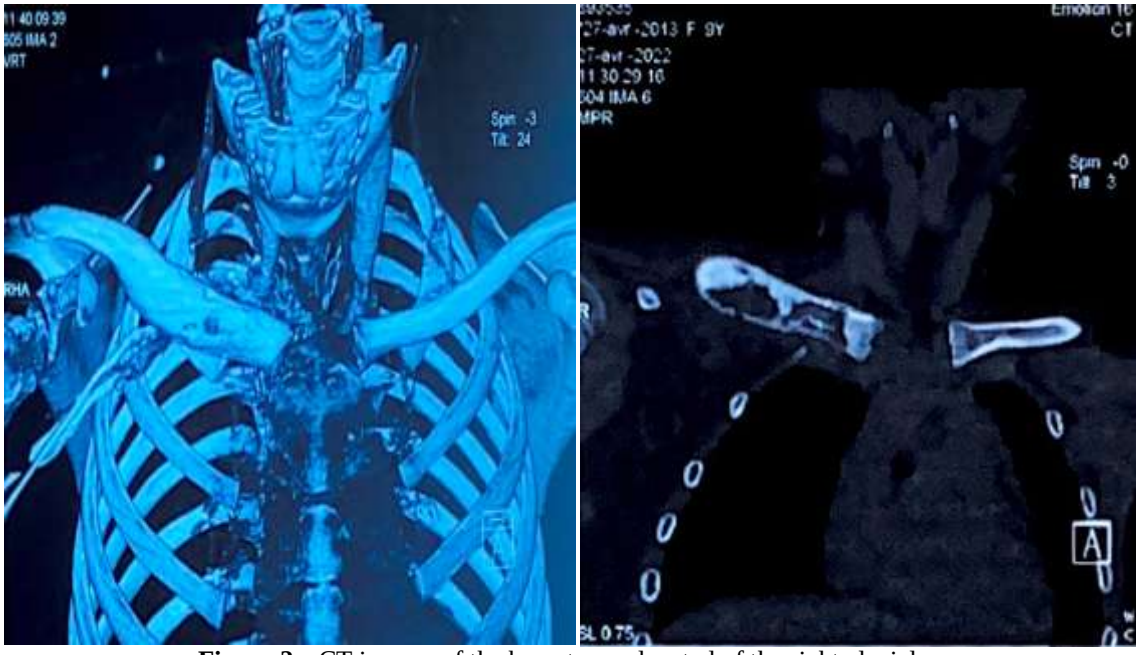

Figure 2:- CT images of the bone tumor located of the right clavicle.

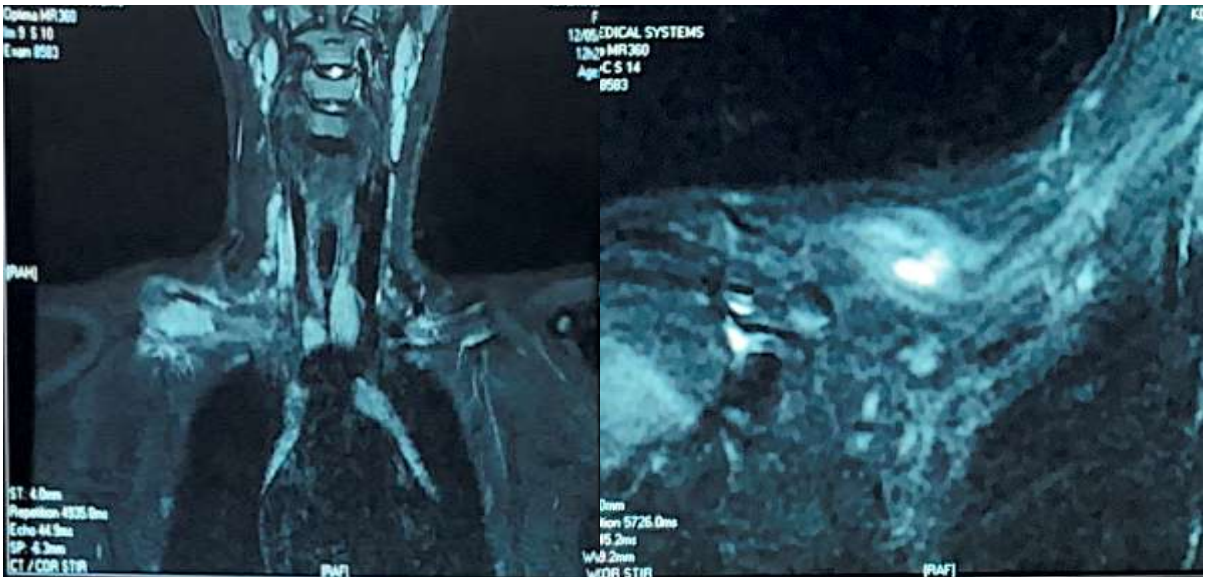

Figure 3:- MRI findings in our patient.

Bone scintigraphy (17/5) found intense uptake taking up the entire right clavicle corresponding to the mother tumor and with no suspicious localization at a distance. (Figure 4)

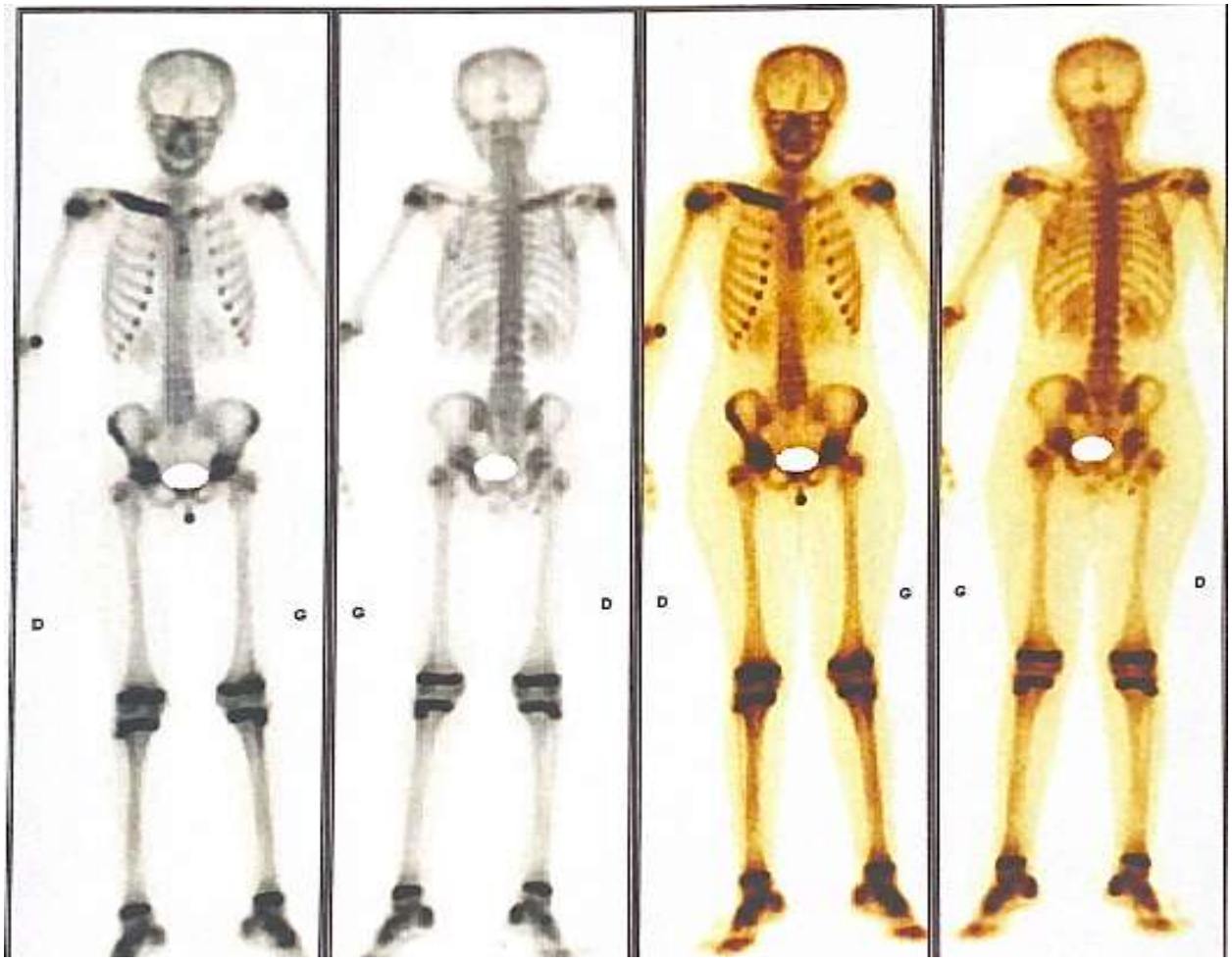

Figure 4:- Scintigraphic findings showing isolated intense uptake on the right clavicle.

The bone biopsy was performed. Interpretation was read and confirmed by two pathologists. The histological findings werein favor of fibrous dysplasia of the clavicle.

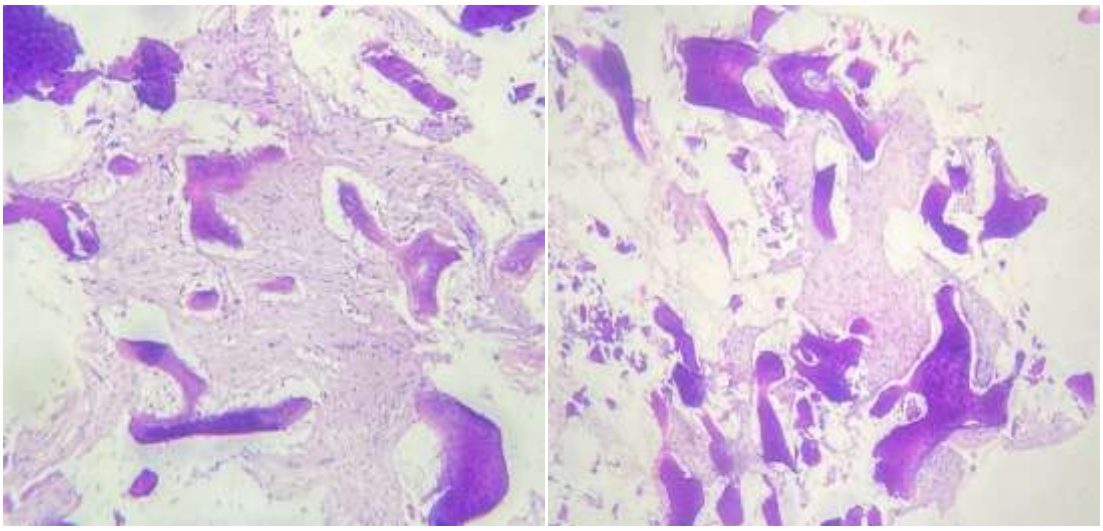

Figure 5:- Histological findings showing in favor of fibrous dysplasia.

The patient received symptomatic treatment consisting of immobilization, analgesics and anti-inflammatories, which contributed to thesedation of pain, in association with a 3 days Bisphosphonatestherapy and D-vitamin supplementation.

Discussion:-

Fibrous dysplasia is characterized by the following parameters: Sporadic and mosaic mode of transmission; Absent at birth;Association with the chromosomal locus 20 q13 and the GNAS gene [1]. It represents about 1% of primary bone tumors, and 7% ofbenign bone tumors [2,3]. It affects children and young adults during the 2nd decade of life [4,5].

Its monostotic form is six times more frequent than the polyostotic form. It can remain asymptomatic for a long time and oftenfortuitous discovery, often revealed by swelling or bone deformation [6,7].The female sex accounts for half of the cases of simple fibrous dysplasia. The maxillary bone, the proximal femur and the tibia are the bones mostfrequently affected, then, less frequently, the humerus, the radius and the iliac bone. The conventional radiological aspect is alytic lesion endomedullary, diaphyseal or metaphyseal with a border of marginal sclerosis and endosteal resorption, the diameter of the bone isgenerally increased, and its interior is made of a matrix having the density of ground glass. Lesions may contain arciform calcificationsreflecting progressive calcification of the cartilaginous nodules. These aspects are variable on the long bones and much more on the flat bones.

However, CT is useful in craniofacial lesions to better define the extent of the disease. MRI and bone scanfind an indication for detecting poorly visible lesions in conventional radiology. Bone scintigraphyis useful indetecting other asymptomatic sites.

The evolution is variable, but the monostotic forms detected in childhood are generally not very progressive. Surgical tissue resection is only indicated if the disease is symptomatic, it remains reserved for voluminous forms weakening the bone in order to preventpathological fractures and to stop the evolution of deformities.

Patient monitoring should be prolonged to detect sarcomatous degeneration of these lesions. The interest of bisphosphonates as a medical treatment remains to be proven.

Conclusion:-

Clavicular localization of monostotic fibrous dysplasia is exceptional. Imaging can be misleading, positive diagnosis is often determined by histology.

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