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Orbital Cellulitis Revealing a Craniofacial Fibrous Dysplasia

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Abstract **Case Report**

Fibrous bone dysplasia (FOD) is a benign, rare, congenital condition, underestimated because it is often asymptomatic, and of unknown cause. It is a disease in which normal bone is replaced by fibro-osseous tissue resulting in deforming bone lesions. The disease is most often monostotic, much more rarely polyostotic. We report an observation of a 10-year-old female patient who presented with a painful periorbital swelling initially labeled as orbital cellulitis. Orbital CT confirmed bilateral orbital cellulitis but also made the diagnosis of craniofacial fibrous dysplasia on the bony windows. The patient was managed by oral antibiotic therapy with incision and drainage of the orbital cellulitis.

Keywords: Orbital cellulitis, fibrous bone dysplasia.

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INTRODUCTION

Fibrous dysplasia of bone, described by Lichtenstein [1], is a bone dystrophy involving one or more bones, resulting in the replacement of healthy bone by fibrous tissue. It is a rare benign pathology with a prevalence of less than 1/2000. It represents 2.5% of bone anomalies and 7% of benign bone tumors. There are two types of fibrous dysplasia: monostotic forms (70% of cases) which affect only one bone and polyostotic forms (30% of cases) which affect several bones [3]. The clinic is asymptomatic in the monostotic form and in the polyostotic form is revealed by limb deformities, craniofacial deformities, exophthalmos and sometimes accompanied by café au lait spots. As clinical and radiological data are not very helpful [4], the diagnosis is based on histological examination [5]. Because of the risk of recurrence, treatment should be conservative as much as possible, modulated according to the age of the patient and the type of fibrous dysplasia. Regular monitoring is necessary because of the small but real risk of sarcomatous transformation [6].

OBSERVATION

This is a 10-year-old girl referred by her pediatrician for a CT scan of the face because of the suspicion of orbital cellulitis. The patient presented clinically with a bilateral periorbital swelling, more marked on the left, painful and renal with a fluctuating area opposite the left medial cantus (Fig 1). The CT scan confirmed a bilateral orbital cellulitis and showed a speckled appearance on the bone window with dense areas of bone rarefaction involving the two maxillary bones, the mandible, the clivus, the medulla oblongata and the medulla oblongata, the clivus, the sphenoidal bone, the two temporal bones, the frontal bones and the walls of the orbits with hypertrophy of all the bones responsible for a filling of the sinuses by a ground glass opacity and an obstruction of the lacrimal canals (Figure 2 & 3). No signs of optic nerve compression were found. Given the absence of optic nerve compression, the proposed treatment was oral antibiotic therapy with incision and drainage of the orbital cellulitis and rigorous clinical and radiological follow-up.

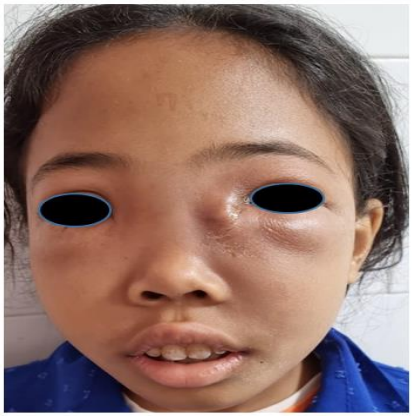

Figure 1

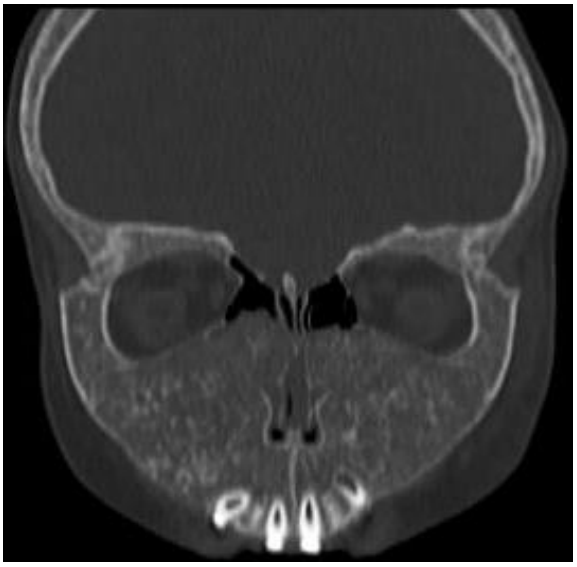

Figure 2: Bone window coronal section

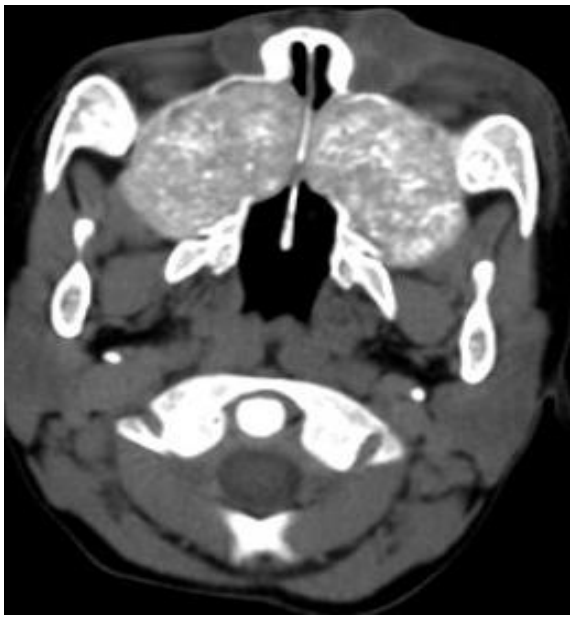

Figure 3: Parenchymal window with PDC injection in axial section

DISCUSSION

FOD is a congenital and non-hereditary disease characterized by fibrous hamartomatous proliferation in the bone marrow, with secondary metaplasia, mainly bone and sometimes cartilage. The frequency is low (0.8% of tumors for monostotic forms) [6]. It can present in 2 forms. The disease is most often monostotic, much more rarely polyostotic. All bones can be affected with a predominance of the femur 36%, the tibia 19%, the skull 17% and the ribs 10%. Polyostotic forms are usually unilateral or predominantly unilateral [6]. The size of the lesions is highly variable, usually larger in polyostotic forms. Monostotic forms are often asymptomatic and are discovered incidentally. Polyostotic forms can often be revealed in childhood by limb deformities, craniofacial deformities, exophthalmos, and sometimes by cranial nerve compression [6]. Polyostotic involvement may be associated with precocious puberty, hyperthyroidism, hyperparathyroidism, Cushing's syndrome, diabetes insipidus, and pigmented spots (irregular café-au-lait spots) constituting the Mc Cune - Albright syndrome. Café-au-lait spots are rare in monostotic forms. The etiopathogeny of fibrous dysplasia is unclear [7], several hypotheses have been advanced. In the congenital hypothesis, it would be a defect of the osteoforming mesenchyme. The traumatic hypothesis is still discussed: the lesion would result from post-traumatic intraosseous hematomas. An endocrine hypothesis (excessive synthesis of Releasing Factor) has also been proposed [8]. Biologically, there are few abnormalities in DFO, calcium and phosphorus are normal in the blood and urine, and alkaline phosphatases are sometimes discreetly elevated. Their very significant elevation should lead to a search for associated hyperparathyroidism. Radiographic and scintigraphic examinations should always be performed to look for other locations of the disease [9]. Imaging varies according to the location, and the degree of mineralization increases with the age of the lesion [3]. The typical appearance on standard radiography is a well-defined, moderately expansive lesion with a lytic "ground-glass" appearance with a border of sclerosis in the periphery that is sometimes discontinuous. When the bone component is more marked, the lesion is more condensed. Sometimes trabeculations of neoformed bone are found within the lesions. Intra-lesional cartilaginous islands may exist, sometimes ossified and responsible for flake or ring calcifications. The peripheral sclerosis may be thickened and produce a "ring sign" appearance. The CT scan allows to specify the extension and the nature of the lytic lesions, to evaluate the craniofacial lesions and to demonstrate a hemorrhagic complication. The appearance of the lesion is classically frosted glass of variable density, with clear contours, bordered by a thick osteocondensing border. The bone may take on an enlarged, expansive appearance with cortical thinning, especially in extensive forms. Heterogeneous bone islands can be seen in the medullary cavity, the density of which varies according to the fibrous or bony tissue content. The density is higher in fibrous dysplasia than in other lesions such as malignant tumors, histiocytosis, or osteomyelitis. There are forms that take on a cystic appearance more frequently in the long bones, whereas in cranial locations, the condensing form is classic, which may suggest Paget's disease .CT is an aid to standard radiography for the diagnosis of dysplasia; it allows the risk of fracture to be specified and cortical thinning to be better analysed. The MRI is not very useful for diagnosis, the signal of the lesion is often variable, but it allows, in addition to the CT scan, to establish a diagnosis of benignity and eliminate an aggressive lesion. It will make the diagnosis and the assessment of a malignant transformation. The scintigraphy reveals an aspecific hyperfixation and allows an assessment of the extension of polyostotic

forms by detecting the asymptomatic forms. It provides a "cartography" and directs to a complementary imaging (CT / MRI) if the fixation is located at the cephalic extremity. The monostotic or polyostotic FOD lesions may spread and worsen during childhood, but stabilize as a rule after puberty, with possible reactivation during pregnancy or estrogen treatment. Fractures may occur with minor trauma. A secondary aneurysmal cyst may develop on a monostotic or polyostotic form. Sarcomatous degeneration is rare (0.4 to 1 % of cases), more frequent in the polyostotic form. It becomes osteosarcoma, fibrosarcoma, more rarely chondrosarcoma. The differential diagnosis varies according to the type of lesion: essential cyst, aneurysmal bone cyst, chondroma, low-grade osteosarcoma, osteofibrous dysplasia, eosinophilic granuloma, Paget's disease, metastasis. The histological differential diagnosis is sometimes difficult, especially with ossifying fibroma where the characteristic osteoblastic front and the presence of lamellar bone are sought [10]. The evolution of fibrous dysplasia is slow and unpredictable. The therapeutic possibilities are discussed. It is essentially that of complications or their prevention. Compression of the auditory or optic nerve can be observed in craniofacial localizations. Spinal cord compression due to vertebral involvement is possible but rare. Radiotherapy increases the risk of sarcomatous transformation: it is therefore contraindicated [11] bisphosphonates associated with calcium supplementation would be effective in painful polyostotic forms. Surgery is indicated for severe and advanced forms with pain, deformity, fracture or risk of fracture. In all cases, the follow-up must be very strict because of this unpredictable evolution. In our case, the patient will be seen in regular consultation for a clinical and radiological assessment.

CONCLUSION

Craniofacial FOD is a rare bony pathology which can manifest itself by important aesthetic and functional disorders. It poses, in some cases, real difficulties of management and requires a rigorous follow-up because the risk of sarcomatous degeneration

Conflicts of Interest: The authors declare no conflicts of interest.

Authors' Contributions

All authors contributed to the conduct of this work. They have read and approved the final version of the manuscript.

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