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FIBROUS DYSPLASIA IN "JUST A NARROW EAC" PATIENT

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Introduction: Fibrous dysplasia (FD) is a non-hereditary benign disorder of skeletal development characterized by the replacement of spongy bone tissue by fibrous connective tissue due to abnormal fibroblast proliferation and poor osteoblast differentiation.

Methods and Materials: We present the case of an 18-year-old man with a history of recurrent otorrhoea. During the usual ENT controls in his childhood was observed a progressively narrower EAC until it was almost occlusive, in a CT study a typical pattern of FD was evidenced that compromises temporal, parietal and part of occipital. Due to the stability of the lesion in late adolescence, a follow-up was decided without other invasive measures.

Discussion: The FD was described in 1891 by Von Reklinhausen and was defined as FD by Lichtenstein in 1923. The etiology of FD is linked to the somatic mutation of the Gsa gene (GNAS1) on chromosome 20q11. There is an increase in adenylate cyclase that increases intracellular cAMP; This high concentration generates an increase in the proliferation and inappropriate differentiation of mutated cells, causing the formation of an immature and disorganized fibrous matrix that histologically appears as trabeculae of immature bone (thin, unconnected, curvilinear in a "Chinese letter" or " puzzle ") inserted in a fibrocellular stroma where scattered osteoclasts can be found. The most common form of presentation is monostotic, where a single bone is altered without systemic involvement (70-85% of cases). The polyostotic form is less common, 2 or more non-contiguous bones are affected, and endocrine compromise can be associated (Sd. McCune-Albright, Jaffe-Lichtenstein). The craniofacial form is rare, and consists with lesions of continuous bones from the craniofacial skeleton, without endocrine alterations. It usually presents with local pain, pathological bone deformities and fractures, hemorrhage, neurological compromises, hearing loss, diplopia, and rarely associated to osteosarcomas. They can also be found as a finding within another study. There is no consensus on treatment; management can be expectant or surgical when the lesion allows it. Pain management and the treatment of the endocrine disturbances when found is important to keep in mind. The most accepted treatment is bisphosphonates and lately the use of denosumab.

Conclusions: The FD is a benign disorder, potentially problematic if it compromises specific structures. If the lesion does not produce symptomatology, the follow- up of the patient is a valid decision.