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RESEARCH ARTICLE

FIBROUS DYSPLASIA OF THE MIDDLE TURBINATE: RARE LOCALIZATION

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Abstract

Fibrous dysplasia is a rare bone pathology and it can often go unnoticed due to its few symptoms.Despite being a benign process, it can behave in aggressively by its local growth, and sometimes degenerate to osteosarcoma. We report a case of fibrous dysplasia which is atypical due to its exclusive location in the middle turbinate.The diagnostic and therapeutic problems that this entails are raised.

Introduction:-

Fibrous dysplasia is an infrequent abnormality of bone development characterized by the replacement of bone marrow by fibro-osseous tissue. It accounts for 7% of all benign bone neoplasms that can affect the entire human body. (1)

Involvement of the craniofacial bones is common, but involvement of the nasal turbinate, either alone or with adjacent bones, remains very rare.

We report the case of fibrous dysplasia of the middle turbinate in a patient followed and treated in our department.

The aim of this work is to provide an atypical case of fibrous dysplasia exclusively localized at the level of the middle turbinate, and to discuss the diagnostic and therapeutic problems that arise due to this special location.

Clinical case:

This is a 38-year-old man, in good health, with no medical or surgical history, who was admitted for moderate episodes of left epistaxis, evolving for 05 months, with intermittent frontal headaches for 01 month.; all evolving in a context of apyrexia and conservation of the general state

On admission, the inspection did not find any facial asymmetry or proptosis.

A nasal endoscopy showed a right deviation of the nasal septum and a left middle turbinal hypertrophy with a normal-looking mucous membrane. The cavum was free

A naso-sinus computed tomography (CT) showed a lesional process measuring 37*16*27mm (AP*T*H) involving the left middle turbinate, with regular contours, osteocondensing density in ground glass not enhanced after injection of product contrast, and exerting a mass effect on the nasal septum, suggesting fibrous dysplasia of the middle turbinate. (Figure 1)

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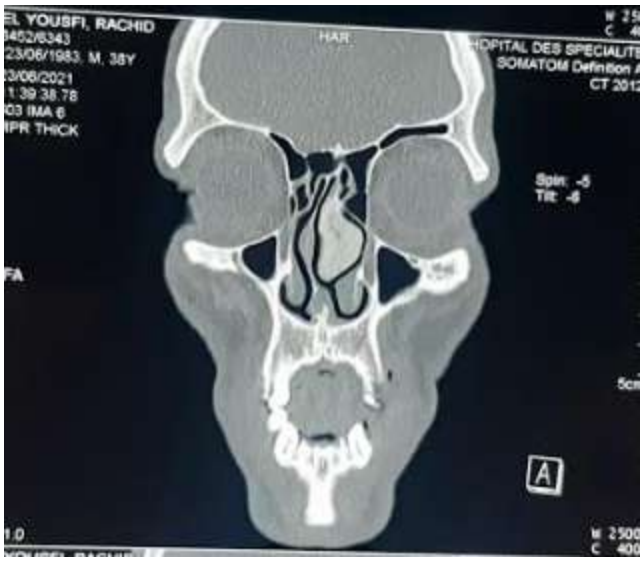

Figure 1:- Naso-sinus CT showing the fibrous dysplasia of the left middle turbinate.

The patient underwent a left middle turbinectomy under general anesthesia and under endonasal endoscopic guidance; subluxation of the left middle turbinate, resection from its root and extraction from the left nasal fossa was performed. (Figure 2)

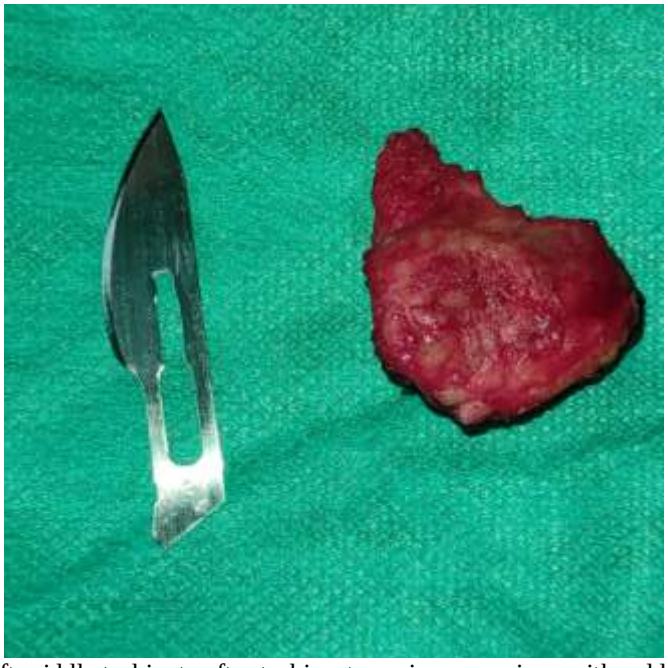

Figure 2:- Left middle turbinate after turbinectomy in comparison with cold blade N° 24.

The definitive anatomopathological examination objectified a morphological aspect of a fibrous dysplasia without histological signs of malignancy. (Figure 3)

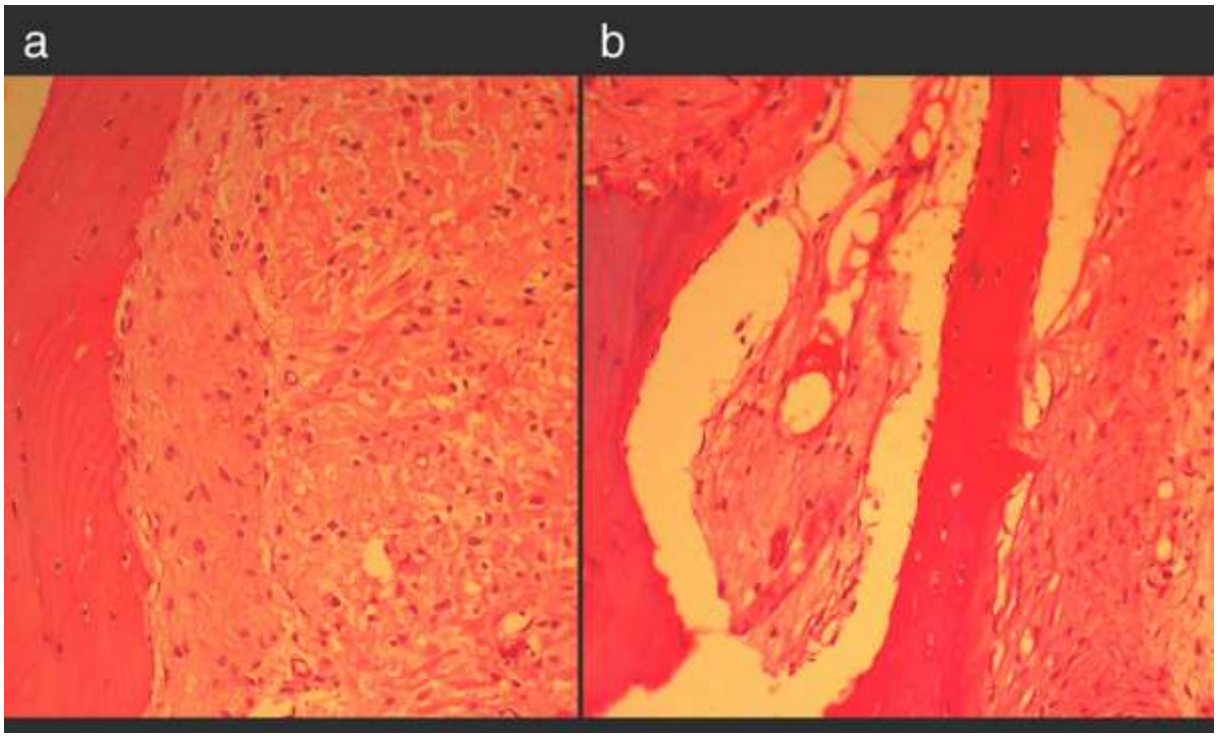

Figure 3:-Haematoxylin and eosin-stained photomicrograph (a) shows isolated trabeculae of woven bone without osteoblastic rimming and intervening cellular fibrous tissue comprising benign spindle to polygonal cells with no atypical features. (b) Additional photograph demonstrates sheets of mature bone separated by focal cellular areas with formation of fibrous and hyaline cartilage nodules.

The clinical and endoscopic follow-up of the patient was satisfactory without recurrence with a follow-up of one year.

Discussion:-

Fibrous dysplasia is an infrequent benign bone pathology, characterized by a slowly progressive replacement of the bone marrow by fibro-osseous tissue.(1)(2)

It is a congenital sporadic lesion which represents 1% of general bone tumors and 7% of benign bone tumors. (3)

About 25% of cases involve a craniofacial injury. (3) Very rarely, fibrous dysplasia is observed in the nasal cavities. (4)

First reported by Von Recklinghausen in 1891 in patients with neurofibromatosis, it was presented in detail by McCune and Brunch in 1937 and named by Lichtenstein in 1938. (5)

With a prevalence of less than 1/2000, probably underestimated due to the asymptomatic forms, the disease affects both sexes, but predominates in women (sex ratio=3F/1M).

It is more common in young adults in the second decade of life (average age = 28.6). (6), (7)

Depending on the number of bones involved and the secondary symptoms, three forms of the disease are described: monostotic, polyostotic, and complicated (Mc-cune Albright syndrome and Mazabraud syndrome)

The most common form is the monostotic form (70%) in which only one bone is involved. (5)

The most frequent sites in the craniofacial region are the maxilla and the mandible, followed by the frontal, parietal and occipital bones. (8) (9)

Our case presents an exclusive fibrous dysplasia at the level of the middle turbinate (monostotic) without any other location in a 38-year-old patient

The polyostotic form represents 30% and affects several bones. At least half of cases of polyostotic fibrous dysplasia involve craniofacial bones. The lesion can affect any bone of the face. (10)

McCune-Albright syndrome is a polyostotic form that combines precocious puberty, endocrine abnormalities such as thyroid nodules with hyperthyroidism, adrenal hyperplasia with hypercorticism, pituitary tumors with acromegaly or hyperprolactinemia. It accounts for about 3% of all cases.

Mazabraud's syndrome associates, in addition to polyostotic fibrous dysplasia, intramuscular myxomas which are generally located near the bone lesions, and tend to recur after surgical excision. (11)

Some cases of polyostotic forms and McCune-Albright syndrome continue to develop after adulthood; the reason for the continued bone growth has not yet been established. (12)

Fibrous dysplasia is characterized by the inability of bone tissue to produce mature lamellar bone. Histological examination shows an accumulation of immature bone tissue. Molecular studies have suggested that it is a non-inherited genetic disorder caused by a sporadic missense mutation in the GNAS1 gene on chromosome 20, which codes for the stimulatory G-protein-coupled receptor alpha subunit. Therefore, organs that have the stimulatory G protein-coupled receptors, such as bones, skin, ovaries, thyroid, and pituitary, are often affected. Therefore, the clinical spectrum of dysplasia is variable, depending on the stage of embryogenesis when the mutation occurred. (13), (14)

The clinical features of Craniofacial Fibrous Dysplasia are nonspecific, with painless facial and cranial deformities reported as the first signs. Involvement of the paranasal sinuses, orbits, and basal foramina of the skull may produce a variety of symptoms, including headache, especially frontal headache, visual signs (decreased visual acuity, proptosis, diplopia, epiphora) and rhinological signs (nasal obstruction, anosmia, epistaxis, mucoceles and recurrent rhino sinusitis)

The disease has a submucosal growth pattern, and nasal endoscopic examination may reveal normal mucosa with turbinate enlargement that may be confused with Concha bullosa. (9)

That said, the positive diagnosis of fibrous dysplasia must necessarily rely on imaging and anatomopathological examination. (15)

Computed tomography is strictly related to the degree of tissue mineralization, ranging from the radiolucent appearance of the lesion in the early stages to later ground-glass appearance.

The most common form is “frosted glass” (56%). (9)

Useful for detecting and defining neurovascular and ocular involvement, MRI can assess the compression of adjacent structures, especially the sinuses. It helps to differentiate inflammatory changes (retention of secretions) and persistence of residual tissue after surgery or malignant transformation of the disease. Finally, it makes it possible to identify the existence of infiltrations of the bone and/or hydro-aeric levels in the lesion. (5), (16)

Fibrous dysplasia may be associated with hormonal changes, in particular with abnormalities in phospho-calcium metabolism.

Laboratory tests such as serum calcium, alkaline phosphatase, urinary hydroxyproline and parathormone evaluation are mandatory to exclude McCune Albright syndrome. (17)

The differential diagnosis must be established mainly with ossifying fibroma, osteoma and chondromas, among others.

The clinical and radiographic results being often similar between these entities, the differentiation will be established mainly by the histological data.

Therapeutically, close clinical and endoscopic monitoring is the most accepted approach in patients with asymptomatic fibrous dysplasia,

However, if malignancy is suspected, a biopsy should be performed.

The regional anatomy determines the management of fibrous dysplasia involving the middle turbinate.

The configuration of the ethmoidal roof and the depth of the cribriform plate on CT were classified by Keros into three anatomical categories. (18)

The category associated with the deep olfactory fossa is the most vulnerable to iatrogenic damage during surgery due to a long lateral lamella and close proximity of the anterior fossa structures to the nasal cavity.

Additionally, the entry point of the anterior ethmoid artery is the thinnest in the region, predisposing to cerebrospinal fluid leakage and vascular injury leading to hemorrhage and orbital hematoma. (19)

Close clinical and endoscopic monitoring is the most accepted approach in patients with asymptomatic fibrous dysplasia, this is due to the fact that with the end of bone growth, the appearance of new lesions decreases, and over time they become more sclerotic.

Radiotherapy is contraindicated because it increases the risk of malignancy.

In our case, it was a symptomatic fibrous dysplasia of the middle turbinate since the patient presented episodes of left epistaxis for 5 months.
Thus, he benefited from a left middle turbinectomy from his root, respecting the roof of the ethmoid and the other adjacent structures.

We should suspect malignant transformation when Fibrous Dysplasia causes pain or spreads to soft tissue.

Spontaneous sarcomatous transformation is very rare (<1%), especially in patients who have received radiotherapy. In 50% of cases, this degeneration occurs in foci located in the craniofacial region. (16), (6), (15)

The prognosis of fibrous dysplasia is generally good, although the results are worse in young patients and those with polyostotic forms. (13)

Recurrence is rare in adults, but lesions may show unexpected growth potential if surgically modified during their active growth phase.

Conclusion:-

Fibrous dysplasia of the craniofacial bones is common, but involvement of the nasal turbinate, either alone or with adjacent bones, is very rare.

Involvement of the middle turbinate, particularly the lateral lamella, has clinical significance, as it poses a risk of injury to the cribriform plate and skull base during surgical or endoscopic intervention.

The differential diagnosis must be established with the typical pathologies of this anatomical structure of a bony nature.

It should be considered in the presence of any picture of unilateral nasal obstruction with epistaxis

Imaging (CT and MRI) with soft tissue and bone reconstruction and assessment of the area in multiple planes provides a comprehensive diagnosis.

Conflicts of Interest:-

The authors declare no conflicts of interest regarding the publication of this paper

Références:-

1. Delap TG, Kaberos A, Dowling P, et al. Cranio-facial fibro-osseous dysplasia. Acase report and overview of the management of the condition. Int J PediatrOtorhinolaryngol 1996;37:179–91.
2. Alba García JR, Armengot Carceller M, Pérez Fernández CA, Díaz Fernández A,Taleb C, Campos Catalá A, Basterra Alegría J. [An exceptional form of cranio-facial fibrous dysplasia: fibrous dysplasia of middle turbinate]. Acta Otorrinolaringol Esp. 2002;53:291-4.
3. Feller L, Wood NH, Khammissa RA, Lemmer J, Raubenheimer EJ. The nature of fibrous dysplasia. Head Face Med 2009; 5: 22.
4. Galvan O, Gassner EM, Neher A, et al. Fibro-osseous lesion of the middleturbinate: ossifying fibroma or fibrous dysplasia? J Laryngol Otol 2007;121:1201–3.
5. Sadeghi SM, Hosseini SN. Spontaneous conversion of fibrous dysplasia into osteosarcoma. J Craniofac Surg 2011; 22: 959–61.
6. Shapeero LG, Vanel D, Ackerman LV, et al. Aggressive fibrous dysplasia of the maxillary sinus. SkeletalRadiol1993; 22:563-568.
7. ZeinebAyadhi et al. Dysplasie fibreuse ethmoïdo-frontale: à propos d'un cas. Pan African Medical Journal. 2021;38(385). 10.11604/pamj.2021.38.385.27561.
8. Bhat V, Kansal K, Krishna SH, Pobbysetty R, Hassan S. Fibrous dysplasia of the middle nasal turbinate: imaging and clinical significance.BJR Case Rep 2016; 2: 20150296.
9. Apostolos Karligkiotis, Paola Terranova, Iacopo Dallan, Paolo Castelnuovo, Monostotic Fibrous Dysplasia of the Inferior Turbinate ; Otolaryngology–Head and Neck Surgery 146(6) 1035-1036 American Academy of Otolaryngology Head and Neck Surgery Foundation 2012.
10. Erdem LO, Erdem CZ, Kargi S. A case of monostotic fibrous dysplasia of themaxillary sinus. KulakBurunBogazIhtisDerg2003;10:208–11.
11. Mazabraud A, Girard J. Un cas particulier de dysplasie de localisations osseuses et tendineuses. Rev Rhum Mal Osteoartic. 1957 Sep-Oct;24(9-10):652–9.
12. Kusano T, Hirabayashi S, Eguchi T, et al. Treatment strategies for fibrous dyspla-sia. Craniofac Surg 2009;20:768–70.
13. Parekh SG, Donthineni-Rao R, Ricchetti E, Lackman RD. Fibrous dysplasia. J Am Acad Orthop Surg 2004; 12: 305–13.
14. Furin MM, Eisele DW, Carson BS. McCune-Albright syndrome (polyostotic fibrous dysplasia) with intracranial frontoethmoid mucocele. Otolayngol Head Neck Surg 1997; 116: 559-62.
15. Başak S, Karaman CZ, Akdilli A, Mutlu C, Odabaşi O, Erpek G, et al. Evaluation of some important anatomical variations and dangerous areas of the paranasal sinuses by CT for safer endonasal surgery. Rhinology 1998; 36: 162–7.
16. Marín Garrido C, Fernández Liesa R, Naya Gálvez MJ, et al. Displasiafibrosacraneofacial. A propósito de dos casos. ORL- IPS 2000; 28: 74-78.
17. Caudill R, Saltzman D, Gaum S, Granite E. Possible relationship of primary hyperparathyroidism and fibrous dysplasia: report of case. J Oral Surg. 1977;35:483-90.
18. Nair S. Importance of ethmoidal roof in endoscopic sinus surgery. Sci Rep 2012;
19. Hwang PH, Abdalkhani A. Embryology, anatomy and physiology of the nose and paranasal sinuses. In: Snow JB, Wackym PA, Ballenger JJ, eds. Ballenger's otorhinolaryngology: head and neck surgery. Revised edition. Shelton, CT: PMPH-USA; 2009. pp 455–63.