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Case Report

Fibrous Dysplasia of the Paranasal Sinuses - A Rare Presentation

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Abstract

Fibrous Dysplasia in the head neck region generally have a predilection for the maxilla and the mandible. Presentation of FD as a paranasal sinus lesion is very rarely reported in literature. We present such a case of paranasal sinus FD discovered incidentally during the ENT examination in a patient presenting with mucosal disease of the middle ear.

Keywords: Fibrous Dysplasia; paranasal sinus

Introduction

Fibrous Dysplasia is a benign condition of the bone in which normal bone is replaced by abnormal fibrous (connective) tissue. This abnormal bone is quite fragile and prone to fracture. Pain may occur at the presenting site. But symptoms are variable and it may be completely asymptomatic, apart from the cosmetic deformity due to the abnormal bone growth. Fibrous Dysplasia generally affects the long bones of the legs like the Tibia and the Fibula. Common sites of Fibrous Dysplasia in the head neck region are the maxillary bone and the mandibular bone [1]. However, we came across an abnormal presentation of FD of the Nasal and Paranasal bones which is described below.

Case History

An 18 year old male patient came to ENT outpatient department with chief complaints of discharge from the left ear since past 8 months. The discharge was insidious in onset, mucopurulent, whitish, non-blood stained, non-foul smelling, present continuously, temporarily decreased on taking medications. He also complained of decreased hearing from the left ear which was gradual in onset and progression and present since past 4 months. We noticed that the patient had some sort of facial asymmetry of the head region but it was asymptomatic (fig.1). Pure tone audiometry showed moderate conductive hearing loss.

On examination it was found that the patient had granulations over the left tympanic membrane with moderate central perforation. He was simultaneously posted for endoscopic evaluation of nose to rule out other foci of infection. On endoscopic assessment it was found that the patient had abnormal thickening of the septum with inferior turbinate hypertrophy (abnormally enlarged inferior turbinate). The patient was advised CT scan of the paranasal sinuses and HRCT temporal bone, which revealed some interesting results.

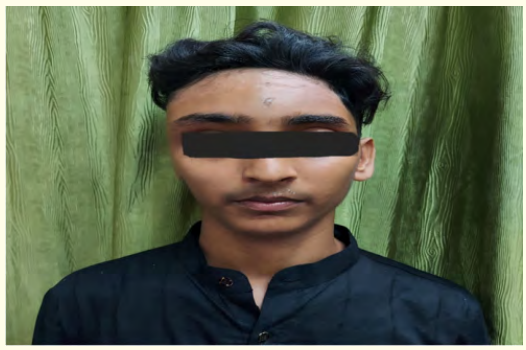

Figure 1

The HRCT of temporal bone was suggestive of soft tissue thickening in the left middle ear and mastoid cavity which was suggestive of chronic suppurative otitis media. The Paranasal Sinuses CT scan showed the presence of bony expansion of bilateral frontal, nasal bones, nasal septum, ethmoid bone, sphenoid bone, posterolateral part of right orbit and nasal turbinates, along with sclerosis of the sinuses. This bony expansion had a typical ground glass appearance. Also, there was obliteration of corticomedullary distinction at the above affected sites (fig 2, 3). This typical ground glass appearance with obliteration of corticomedullary distinction was highly suggestive of Craniofacial fibrous dysplasia of the paranasal sinuses.

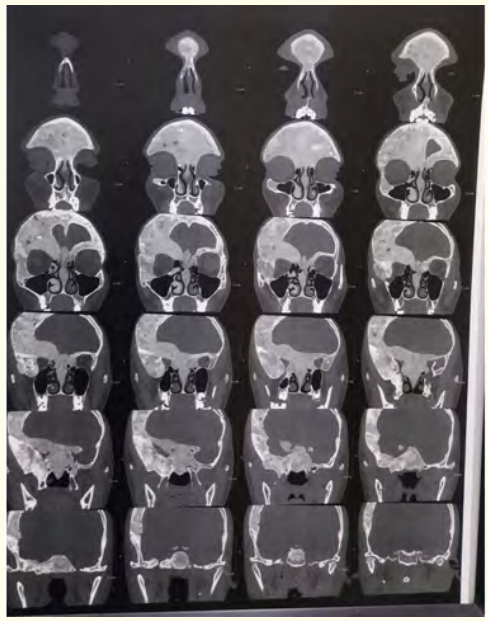

Figure 2

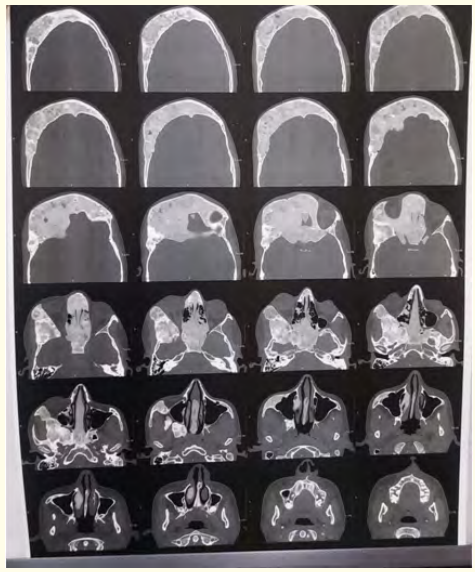

Figure 3

A complete physical examination did not reveal any cafe au-lait spots, there were no features of acromegaly or Cushing's disease. Also, there was no bony abnormality anywhere else on the body. Both his gonads were descended on genital examination. Opthalmic examination also did not reveal any functional abnormality or nerve compression.

Laboratory investigations revealed increased levels of serum Alkaline Phosphatase (525 IU/L). His serum calcium, phosphorous and parathyroid hormone levels were well within normal range. His thyroid function tests were also normal. Radiological examination of the lower limbs was also within normal limits. Since the patient was asymptomatic, he was counselled regarding his condition and the need for regular follow up.

Case Discussion

The term fibrous dysplasia was given by Lichtenstein in the year 1938 [2]. But the pathological description of this disorder was given long ago by von Recklinghausen in 1891 [3]. The aetiology of this disease is said to be due to post-zygotic, somatic mutation of the GNAS gene, which encodes the α subunit of the ubiquitous stimulatory G protein [4].

Typically fibrous dysplasia has been described into three variants, monostotic, polyostotic and as a part of McCune-Albright syndrome. Craniofacial involvement may occur both as true craniofacial fibrous dysplasia, considered a form of monostotic fibrous dysplasia which accounts for 70% of the cases. The craniofacial bones are affected in up to 70-80% of polyostotic cases which is more common than monostotic type which is affected in 20% of cases [1]. Occasionally it is seen in the setting of McCune-Albright syndrome which has polyostotic fibrous dysplasia with extra-skeletal manifestations as skin pigmentation and endocrine abnormality. Within the skull bones, the anterior skull base is more likely to be involved than the lateral skull base. The incidence is found to be more in frontal, ethmoidal and sphenoid sinus compared to temporal bone. In a study done by Lee et al concluded that the mean age at the time of diagnosis of paranasal sinuses was very high 45.9 years [5]. The most common site of occurrence was the sphenoid sinus 71.4% [5]. Isolated Fibrous dysplasia of paranasal sinuses in a very rare occurrence. Also, fibrous dysplasia of paranasal sinuses does not necessarily mean that it originates from the sinuses. The reason for this being that the disease prefers the membranous bones of the face,

but the paranasal sinuses like the ethmoid and the sphenoid develop in a bone which is preceded by cartilage [6].

The entire sinus can be completely obliterated by FD, however the incidence of sinusitis is not greater compared to the general population. This happens because in the disease process, there is loss of air space and Schneider an membrane in an obliterated sinus which elimination of a source of infection. Though nasal congestion appears to be an important symptom in these patients, a history of sinusitis and facial pain/headaches does not correlate with the amount of craniofacial disease [7].

Radiographic findings in a case of fibrous dysplasia was described by Fries in 1957 which categorised them into three characteristic patterns- Pagetoid, Sclerotic and Cystic pattern. The Pagetoid is common occurrence, found in about 56% of cases which is characterised by areas of intermixed tissues of different densities with dense areas of sclerosis and radiolucent areas of fibrosis. The sclerotic form has dense homogenous regions seen in about 23% of the patients. While the cystic type has round radiolucent rings surrounded by dense bone seen in 21% of patients [8]. In our case the radiological appearance was clearly suggestive of pagetoid variety.

It was noted in literature that the biopsy is not always required [6] when the radiologic features are strongly suggestive of fibrous dysplasia. Biopsy is indicated when there is exacerbation of symptoms and sign [6] or when there are no sufficient confirmative radiologic signs. Rapidly growing lesion is also one important finding that needs biopsy. Although CT scan is the first choice of investigation in a suspected case of fibrous dysplasia, MRI can be used to assess the soft tissue components. In a case of Fibrous Dysplasia, the involved bones on T2 weighted images exhibit low signal intensity; on the other hand, Ossifying Fibromas will give out a high signal intensity [9].

Fibrous Dysplasia needs to be differentiated from other similar occurring lesions like osteoma, ossifying fibroma, Pagets disease, enchondromatosis (Ollier Disease), eosinophilic granuloma, McCune Albright syndrome. In our case, the radiological features like ground glass appearance of the frontal, sphenoid and ethmoid bones with schlerosis of the sinuses with loss of corticomedullary distinction was greatly suggestive towards fibrous dysplasia.

In Craniofacial Dysplasia, patient should be operated in case of neurological and ophthalmologic symptoms. The indication of surgeries includes persistent nasal blockage, or sinonasal tract blockage, intracranial spread and in case of ophthalmic symptoms like visual impairment [6].

Bisphosphonates such as alendronate, pamidronate or zoledronic acid for craniofacial FD has been considered for pain reduction and to reduce the rate of growth of the lesion. Wang Y et-al has noted that the bisphosphanates supress the high bone turnover to partially control the activity of the disease. However, some publications has observed osteonecrosis of the jaw caused by bisphosphonate while treating Fibrous dysplasia. And there was no significant improvement radiologically in patients who received these drugs. New therapies are coming up that include RANK ligand (cell surface protein involved in many cellular processes) inhibition (i.e. denosumab) however their role in the treatment of FD-related pain or reduction in growth is still under studies [10]. Surgery is indicated in case of Fibrous Dysplasia when it causes pressure on the surrounding structures or it causes a cosmetic deformity. Surgical approach to paranasal sinus Fibrous Dysplasia can be either External or open approach or Endoscopic. External or open approaches used can be the Caldwell Luc approach, Lateral Rhinotomy approach, External Ethmoidectomy or sometimes even a Craniofacial Resection in case of a large lesion [9]. Endoscopic Transnasal approach are preferred for lesions limited to a single paranasal sinus [9]. Conservative surgery is advocated as the chances of malignant transformation are very less (0.5%) and generally seen only in polyostotic lesions and McCune Albright's syndrome [9].

Conclusion

Fibrous Dysplasia of the Paranasal Sinuses is a rare but benign condition. Malignant transformation is extremely rare and if present is observed only in polyostotic cases and in McCune Albright's syndrome. Active intervention in the form of surgery is recommended in case the lesion is large and causing pressure symptoms or a cosmetic deformity. Radical surgery is not recommended since the disease

follows a slow benign course. Conservative surgery with preservation of function is the preferred choice of management.

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Volume 3 Issue 1 July 2022