Conservative Treatment of Huge Fibrous Dysplasia Causing Dysfunction in the Maxilla

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Abstract

Fibrous dysplasia (FD) is a non-malignant condition in which normal bone and marrow are replaced by fibrous tissue and haphazardly distributed woven bone. In the craniomaxillofacial skeleton, the maxilla is more frequently affected by FD than the mandible and generally affects adjacent bones such as the sphenoid, zygomatic and the frontal bones. The aim of this case report is to present the clinical, radiological, histological findings of patients diagnosed with FD in maxilla.

Keywords

Fibrous dysplasia, Maxilla, Conservative treatment

Introduction

Fibrous dysplasia (FD) is a skeletal anomaly in which deficiently organized and insufficiently mineralized immature bone and fibrous tissue takes the place of normal bone. FD may touch any bone in the body; nevertheless, the jaw, the skull, the pelvis, the long bones, the ribs are most generally affected [1]. The disease may involve a single bone (monostotic) (characterizing most cases of FD) or multiple bones (polyostotic) [2]. The polyostotic type may be correlated to the McCune-Albright syndrome in which characteristic café-au-lait spots are combined with skeletal abnormalities and endocrine abnormalities, as well as the Jaffe-Lichtenstein syndrome in which patients demonstrate the same symptoms, excluding endocrine abnormalities [3,4]. FD of bone evolves from activating missense mutations in Gs alpha gene in pluripotent embryonic stem cells. The inheritance of these mutations remains in a population of postnatal skeletal stem cells or mesenchymal stem cells which direct the formation of atypical bone in FD [5]. In the craniomaxillofacial skeleton, the maxilla is more frequently affected by FD than the mandible and generally affects adjacent bones such as the sphenoid, zygomatic, and the frontal bones [6]. The major clinical symptom of FD is painless swelling in the affected bones. The overgrowth of the bone can cause facial asymmetry, deformation, malocclusion caused by teeth migration and exophthalmos related to the anatomical sites and structures implicated [7]. Radiographic characteristics of FD differ from radiolucent (initial stages) to radiopaque (mature) lesions, in order to the combination of fibrous and osseous elements. The affected bone presents expansion and a ground-glass appearance is generally reported in progressive stages [8]. Histological examination is essential for definitive diagnosis. The aim of this case report is to present the clinical, radiological and histological findings of patients diagnosed with FD in maxilla.

Taking into consideration the gender, age, recurrence probability, size of lesion and involvement of adjacent anatomic structures, the ability to define the accurate diagnosis and plan the most appropriate treatment by the clinician is aimed.

Case Presentation

A systemically healthy, 41-year-old woman has referred to our clinic with a complaint of asymmetry and swelling in maxillary left posterior region for the past 2 years. A biopsy had been performed 2 years before the visit with clear diagnosis of FD. On the clinical examination, teeth migration, malocclusion and asymmetry were obviously visible. There was no swelling present elsewhere in the body, and café-au-lait spots were absent. Routine investigations such as hemogram,
FD is a non-malignant condition in which normal bone and marrow are replaced by fibrous tissue and haphazardly distributed woven bone [1]. All forms of fibrous dysplasia are seen equally in men and women without gender inequity. However, Kruse, et al. [9] reported that the disease was more common in women. It is often thought that lesions will stabilize with the completion of skeletal development in the first three decades of life. However, in this case the age of patients is over 40 years.

The FD has three radiological patterns as follows; the first type is pagetoid with bone expansion and alternate areas of radiodensity and radiolucency. The second pattern is sclerotic, with bone expansion and a homogenous radiodensity (a ground glass appearance). The third type is cyst-like, usually a round or oval lesion with a sclerotic border [10]. Our case matches with the first type of Fries’ descriptions.

MacDonald Jankowski [11] reported that tooth displacement might also occur and lamina dura could be absent in FD patients. Petrikowski, et al. [12] suggested that the loss of lamina dura could be used as an ancillary diagnostic feature for FD. In the present cases, the absence of lamina dura was observed and tooth displacement was present.

Regarding the treatment of fibrous dysplasia in literature there are various conservative and invasive methods [13]. Conservative treatments such as debulking, contour and
In older ages. The patient should be relieved that the lesion is not malignant. Patient controls should not be neglected in case of recurrence and malignant transformation risk, even though it is rare.

Acknowledgement
None.

Conflicts of Interest
There are no conflicts of interest to declare.

Consent
Consent for publication was taken from the patient.

Authors Declaration
The manuscript has not been published and is not under consideration for publication in any other journal. Instructions to the author were read. We accept all conditions and publication rights. All authors have approved the manuscript and its submission to the journal. We have no funding sources to declare.

References

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