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Concomitant occurrence of fibrous dysplasia and epilepsy: Report of an unusual case

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Abstract

Background: Fibrous dysplasia (FD) is a developmental anomaly in which normal bone is gradually replaced with fibrous connective tissue. There are two variants: monostotic and polyostotic. The diagnosis of fibrous dysplasia is generally made on the basis of clinical and radiographic findings. The association between epilepsy and craniofacial fibrous dysplasia is rather unusual. A case of fibrous dysplasia is presented here which was seen co-existing with epileptic seizures.

Case Presentation: An 18-year-old male patient reported to the department with the chief complaint of a painless swelling on the right side of the face since 4 years. He gave history of trauma secondary to a fall 4 years back and seizures since 2 years. On extra-oral examination, a diffuse swelling was seen on the right middle third of face extending medio-laterally from ala of nose to outer canthal line, superiorly involving the zygomatic arch. The swelling was bony hard and non-tender on palpation. Intra-orally, swelling was seen associated with right buccal vestibule extending from right lateral incisors to second molar region, with normal overlying surface. Based on the history and clinical presentation a provisional diagnosis of a benign lesion was made. Radiographic features were suggestive of a fibro-osseous lesion, most probably fibrous dysplasia. Considering the fact that fibrous dysplasia may sometimes lead to epilepsy, patient was referred for thorough neurological examination. A surgery was then planned for the patient for correction of the facial deformity. However, the patient refused any further treatment.

Conclusion: A clinician must be vigilant and cognizant of this association so that a prompt referral and treatment may be achieved. (El Med J 2:3; 2014)

Keywords: Fibrous Dysplasia, Epilepsy

Introduction

Fibrous dysplasia (FD) is an uncommon congenital developmental anomaly of bone where normal bone is replaced by fibrous connective tissue. It has two clinical variants: monostotic and polyostotic form. Craniofacial involvement in FD is seen in almost all cases of polyostotic form and around 30% of the polyostotic form [1]. The association between epilepsy and craniofacial fibrous dysplasia is unusual. Soler et al. have even reported epileptic seizures as the first sign of fibrous dysplasia in one patient [2]. A clinician must be vigilant and cognizant of this association so that a prompt referral and treatment may be achieved. Also, thorough clinical and radiographic examinations are essential prerequisites for the diagnosis of fibro-osseous lesions.

Case Report

An 18-year-old male patient reported to the department with the chief complaint of a swelling on the right side of the face since 4 years. The swelling was painless and had gradually increased to the present size. He gave history of trauma to the secondary to a fall 4 years back. He had consulted a dentist then, however was not rendered any treatment. Patient also complained of seizures since 2 years.

On extra-oral examination, a diffuse swelling was seen on the right middle third of face extending medio-laterally from ala of nose to outer canthal line, superiorly involving the zygomatic arch (Figure 1). The swelling was bony hard and non-tender on palpation. Intra-orally, swelling was seen associated with right buccal vestibule extending from right lateral incisors to second molar region, with normal overlying surface (Figure 2). The teeth present in the region appeared to be normal.

Figure 1: Diffuse swelling on the right middle third of face extending medio-laterally from ala of nose to outer canthal line, superiorly involving the zygomatic arch.

Figure 2: Swelling seen associated with right buccal vestibule extending from right lateral incisors to second molar region, with normal overlying surface.

Based on the history and clinical presentation a provisional diagnosis of a benign lesion was made. Thorough radiographic investigations were advised which included intra-oral periapical radiographs and

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panoramic radiograph. Computed tomography, although indicated, could not be done due financial constraints. Intraoral periapical radiographs showed diffuse radio-opacity in the periapical area with respect to right canine to second molar region [Figures 3(a) and 3(b)]. Panoramic radiograph also showed similar diffuse radio-opacity in the periapical regions of the associated teeth, extending superiorly to involve the zygomatic bone (Figure 4). The radio-opacity showed abnormal and irregularly shaped trabecular pattern with gradual blending of lesion with the normal trabeculae.

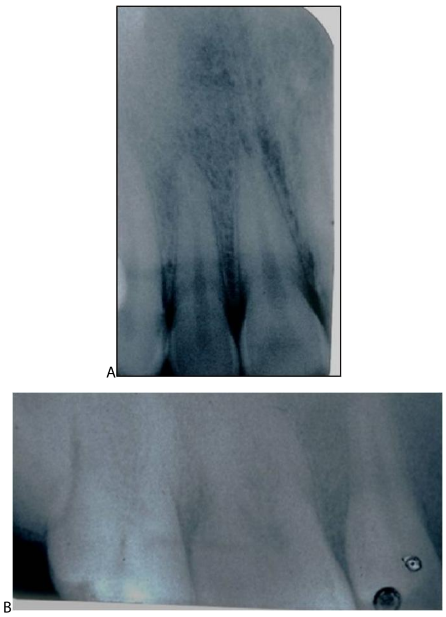

Figure 3: Intraoral periapical radiographs showing diffuse radio-opacity in the periapical area with respect to right canine to second molar region.

Figure 4: Panoramic radiograph showing similar diffuse radio-opacity in the periapical regions of the associated teeth, extending superiorly to involve the zygomatic bone.

The radiographic features were suggestive of a fibro-osseous lesion, most probably fibrous dysplasia. Considering the fact that fibrous dysplasia may sometimes lead to epilepsy, patient was referred for thorough neurological examination. A surgery was then planned for the patient for correction of the facial deformity. However, the patient refused any further treatment.

Patient was followed up telephonically up to 6 months after the initial visit. He was undergoing treatment for epileptic seizures at that time.

Discussion

The term fibro-osseous lesion (FOL) is a generic designation of a group of jaw disorders, characterized by the replacement of bone by a benign connective tissue matrix. This matrix displays varying degrees of mineralization in the form of woven bone or of cementum-like round acellular intensely basophilic structures [3, 4].

Fibrous dysplasia (FD) is a developmental anomaly in which normal bone is replaced with fibrous connective tissue. As the lesion matures, the fibrous connective tissue is replaced with irregularly patterned trabecular bone [5]. It is a localized abnormality, which can involve one (monostotic) or multiple (polyostotic) bones. With initial development of fibrous dysplasia, the patient usually reports facial swellings and asymmetries. Although the lesion is usually asymptomatic, encroachment on canals and foramina, as well as limitations of movement, may lead to complaints of pain and discomfort [6]. In general, males and females are thought to be affected evenly, although recent research has shown a slight female preponderance. The lesions of fibrous dysplasia are twice as common in the maxilla as the mandible, and the posterior aspects of the jaw are more frequently affected than the anterior.

Maxillofacial FOLs are of particular interest to the radiologist because they emphasize the central role of the radiologist in the diagnostic process. The late Charles Waldron wrote "In absence of good clinical and radiologic information a pathologist can only state that a given biopsy is consistent with a FOL. With adequate clinical and radiologic information most lesions can be assigned with reasonable certainty into one of several categories" [7].

The monostotic form accounts for 80–85% of cases of FD. Although the term "monostotic" can be readily applied to cases of FD affecting the mandible alone, this is generally not so for FD affecting the maxilla or face. There, FD can affect contiguous bones such as the zygoma and the sphenoid. These cases have been called "craniofacial FD" [8].

The association of epilepsy and fibrous dysplasia is quite rare. Soler et al in 1999 suggested that epilepsy might be an initial symptom in fibrous dysplasia [2]. The mechanism for the production of epileptic seizures may not be related to compression phenomenon or local ischemia, but may be secondary to alteration in the mechanism of cAMP as the secondary messenger of the cerebral cortex. However, much needs to be explored in this field. Patients with fibrous dysplasia hence should undergo neurological examination to rule out any such possibility.

The diagnosis in this case was made based on the characteristic clinical and radiographic features. Fibrous dysplasia may sometimes be confused with other fibro-osseous lesions such as Paget's disease. In addition to the predilection of Paget's disease for an older population, certain radiographic and clinical features help to distinguish this lesion from other radiographically similar lesions. These features include thickening of the cortices and cotton wool appearance of the involved bone. The most useful clinical feature for distinguishing Paget's disease from fibrous dysplasia is that the former tends to occur bilaterally in the jaws, whereas the latter affects only one side.

Treatment of fibrous dysplasia usually involves bony recontouring at the affected site to improve aesthetics and function. The lesions can show surprising growth potential if they are surgically altered during their active growth phase [5].

Conclusion

A clinician must be vigilant and cognizant of this association so that a prompt referral and treatment may be achieved.

Competing interests: The authors declare that no competing interests exist.
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