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Frontal sinus fibrous dysplasia with atypical radiological features: A case report and review of the literature

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ABSTRACT

Objectives: To present a case of frontal sinus fibrous dysplasia with atypical radiological features and to perform a literature review.
Case report: A 63 year old gentleman presented to clinic with a one year history of right eye proptosis. CT and MRI were performed and identified a mass filling the frontal sinus and eroding the orbital wall. The patient had a combined endoscopic and open approach to the right frontal sinus with the final diagnosis being fibrous dysplasia.
Study design: Case report and literature review.
Methods: Systematic literature review was conducted with the review focusing on the radiological features of fibrous dysplasia in the head and neck.
Results: Fibrous dysplasia can present in a variety of ways in the head and neck. There are certain features in its radiological appearance that can aid in early identification of the disease, thereby leading to the development of better management plans. In CT scan, the ground glass appearance is the most persistent presentation. While in MRI, the degree of mineralization of the lesion dictates its signal intensity.

Introduction

Fibrous dysplasia (FD) is a benign lesion that results from abnormality in bone metabolism [1].

The disease was first mentioned in 1891 when Von Recklinghausen described striking bony lesions that he reported on some of his patients. Some of those lesions are known now to be fibrous dysplasia [1,2]. McCune and Brunch described these changes in detail in 1937, with the term “fibrous dysplasia” first being used in 1938 by Lichtenstein [1,2]. Fibrous dysplasia (FD) may present as a solitary lesion known as a monocratic lesion, or it may present in the more severe polyostotic form. It may also present as part of McCune-Albright's syndrome which includes endocrinopathy and café au lait spots in addition to the polyostotic disease (1, 2, 3).

Although FD is a benign disease, it may be locally destructive. Its most common form is the monostotic form which represents 60% of all cases [4]. The average age of onset is in childhood or puberty with the polyostotic form usually diagnosed at an earlier age because of the more profound nature of the disease [3].

FD is a genetic non-inherited disease. It is attributed to a missense mutation in the GNAS1 (guanine nucleotide-binding protein, alpha-stimulating activity peptide 1) gene. This mutation is known to lead to abnormal proliferation and differentiation of pre-osteoblasts [4,5], with malignant change having relatively low occurrence [5].

Fibrous dysplasia has certain CT characteristics with the ground glass appearance being the most common persistent feature [6,7]. The disease may also present in a homogeneously dense pattern or with a cystic appearance [8].

In MRI, the lesion shows iso-to hypo-intensity in T1 imaging with more variable presentations in T2. The lesion's intensity is enhanced with gadolinium [2,8].

The following is a case report on a 63 years old male who presented with the monostotic form of the disease in the frontal sinus with confusing radiological features.

Case report

A 63 years old gentleman was referred to clinic with a one year history of progressive changes in his right eye. He complained of a droopy right upper eyelid and right sided proptosis. He also complained

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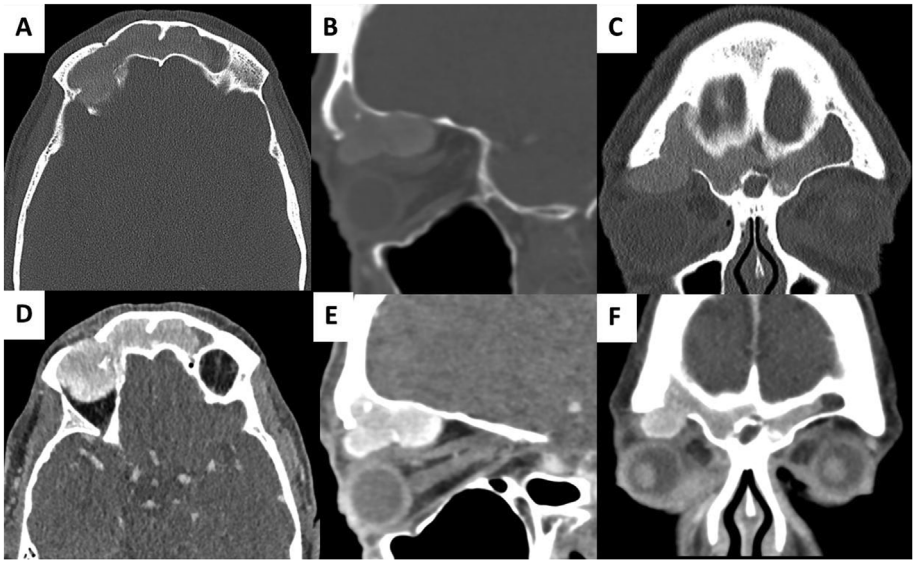

Fig. 1. CT images in the axial (A,D), sagittal (B,E) and coronal (C,F) planes in bone window without contrast (top row) and in soft tissue window after contrast (bottom row).

of vision restriction in upward gaze. The symptoms had progressively worsened over one year. He has a history of facial trauma that was sustained 17 years ago and which resulted in hairline fractures of the frontal sinus.

The patient had a CT scan that revealed a high density lesion in the right frontal sinus extending into the extraconal superior right orbit, the medial aspect of the frontal sinus and bilateral frontal recess. There was an old fracture deformity involving the posterior wall of the right frontal sinus. The most likely radiologic diagnosis was a mucocele.

MRI was performed and it revealed bilateral opacification of the frontal sinus which was hypointense in the T1 and T2 weighted sequence and which enhanced with contrast. The working diagnosis was right frontal sinus mucocele. The patient management plan involved an endoscopic approach to the frontal sinus with a possible orbitotomy in coordination with colleagues from the oculoplastic service.

The endoscopic approach was first attempted. Entry into the frontal recess was successful and subsequently confirmed using image guidance technology. A firm mass was located within the frontal recess. Some of the tissue was removed without mucus drainage. As the clinical findings did not match our pre-operative radiological diagnosis of presumed mucocele, The oculoplastic surgeons then performed a right anterior orbitotomy resulting in the excision of a 2.4 × 2.2 × 1.2 cm nodular firm mass which was completely excised and sent for histopathological examination.

The histological appearance was correlated with the radiological features and the diagnosis was confirmed to be fibrous dysplasia.

Discussion

Fibrous dysplasia is a benign bony lesion that arises as a result of replacement of normal bone by fibrous connective tissue with a characteristic histologic whorled pattern with trabeculae of immature non-lamellar bone [5].

Craniofacial FD is reported to be 30% of the total incidence of the disease [1]. The most common craniofacial form is the monostotic variety [9].

In a systemic review of 766 cases, MacDonald-Jankowski found that the most common presenting feature was a swelling [9]. In a recent case series of 32 patients, the most commonly involved bones were the maxillary, mandibular and sphenoid bones in descending order of likelihood [2].

Malignant transformation is very rare and tends to be increased with the polyostotic form and in McCune-Albright's syndrome. In spite of the benign nature of the disease, it is potentially locally destructive, hence the importance of recognizing and managing it.

CT is the gold standard for the diagnosis of FD. FD has certain characteristics on a CT scan that aid in identifying it. The most consistent feature is its ground glass appearance [2,9–11].

Fries reviewed the radiographic features of craniofacial fibrous dysplasia and described three patterns. The ground-glass pattern as the most common (56%) comprising of a mixture of dense and radiolucent areas of fibrosis. The second most common pattern is the sclerotic which appears homogeneous. The third cystic variant represents 21% and is characterized by a spherical or ovoid lucency surrounded by a dense bony shell [11]. Atalar et al. classified 60% of the cases in their series as being of sclerotic appearance [2].

Chen et al. presented CT finding in 46 patients. CT images appeared sclerotic or homogenous in 34%, heterogeneous in 55%, and cystic in 11% [12]. In a case series of 8 patients, two cases showed ill-defined margins whereas, other lesions were extended into and merged with the cortex of the involved bone. In this series, the internal structure of the bone involved displayed increased density and all 8 cases showed the characteristic ground glass appearance [10].

The radiographic appearance is more radiolucent and well defined in the early stages, and becomes mottled and more radio-opaque as the disease progresses [2]. In our case, the CT scan presented a lobulated, well-circumscribed high density mass (160 Hounsfield units) filling the frontal sinus, which demonstrated expansion of its bony walls. Osteolysis of the right orbital roof was identified through which the mass extended into the extraconal compartment of the orbit causing mass effect and displacing the superior rectus muscle inferiorly resulting in proptosis. The mass showed diffuse enhancement with iodinated contrast (Fig. 1).

On T1-weighted images, the FD shows low to intermediate signal intensity. The difference in signal intensity can be explained by the ratio of fibrous tissue to mineralized matrix of the lesions. Those with high fibrous tissue content display intermediate signal intensities, whereas lesions with highly mineralized stroma demonstrate lower signal intensities. The difference in the composition of the lesion is more clearly reflected on T2-weighted images. Some lesions with a highly mineralized matrix display low signal intensities, whereas lesions with high fibrous tissue content and cystic spaces display high signal intensities. As fibrous dysplasia is metabolically active, it typically shows high signal intensity in contrasted images [8].