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Craniofacial fibrous dysplasia

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ABSTRACT

Objective: The aim of the study was to report the clinical characteristics, radiological imaging methods, and management of patients with fibrous dysplasia. **Materials–Methods:** A retrospective review of 12 patients. Distribution of the cases according to the clinical and radiological features was described. **Results:** The age range was from 9 to 55. Sphenoid bone was the most common involved area in our cases. Simple cystic degeneration was observed in three cases and aneurysmal bone cyst in one case. **Conclusion:** Radiologic findings are characteristic but not pathognomonic. Our management is to follow nonsymptomatic cases or surgical intervention to stop progression of a lesion or to resolve compression symptoms.

1. Introduction

Fibrous dysplasias (FD) are nonhereditary benign pathologies in which immature bone and fibrous stroma replace normal medullary bone as a result of abnormal differentiation of osteoblasts. These mainly divide into two clinical types as monostotic and polystotic FD and are accounted for 7% of all the benign bone tumors. Males and females are equally affected. Most of the cases are diagnosed before the age of 30. Craniofacial region is involved in 25% of the FD cases. The most involved bone structures in the craniofacial region are maxillary and mandibular bones, while involvement of frontal (F), sphenoid (S), and ethmoid (E) bones is uncommon. The least involved structures are temporal (T) and occipital (O) bones. Any clinical symptom out of the facial asymmetry may not be seen for a long period in fibrous dysplasia. It leads to compression and obstruction in the adjacent structures secondary to the enlargement of the lesion and expansion of bone structure over time. Clinical symptoms such as blindness and deafness develop.

2. Materials and methods

Twelve FD patients with craniofacial bone involvement were defined according to the clinical symptoms and imaging methods. Ethical approval to perform the study was obtained from the Mustafa Kemal University's ethical committee. Directed graph and computed tomography (CT) were performed in 10 patients, magnetic resonance imaging (MRI) in 9, and scintigraphic examinations in 2 patients. All the patients were radiologically diagnosed as FD, while the diagnosis was confirmed in postoperative histopathological examinations in 4 patients. TOSHIBA Aquilon multidetector CT device was used for CT scans and 1.5 Tesla Philips device for MRI scans. Intravenous contrast media was used in 8 of 9 patients with MRI performed. Scintigraphic examination was performed in 2 patients for the extension and characterization of the lesion. We retrospectively evaluated these patients, and due to differential diagnosis of lesions in relevant sections, MRI and scintigraphy were requested to these patients. We also aimed to offer MRI and scintigraphy findings.

3. Results

Eight of our patients were females, and 4 were males, and 10 cases were under 30 years old, while the remaining 2 cases were 53 and 55 years old. Six of the cases were monostotic (Fig. 1A–F) and 6 were polystotic FD (Fig. 2A–D). On CT scans, a ground glass-sclerotic appearance was observed in 4 cases, ground glass in 5 cases, sclerotic in 1 case, and radiolucent appearance in 1 case. In nine cases with MRI scans performed, hypo and medium signal feature were dominant in T1- and T2-weighed sections, while a hyperintense appearance was noted in T2-weighed sections of the cases presenting cystic degeneration. Significant contrast media involvement was observed in six, and weak involvement in two cases from those was administered contrast media. There was prominent radioactive

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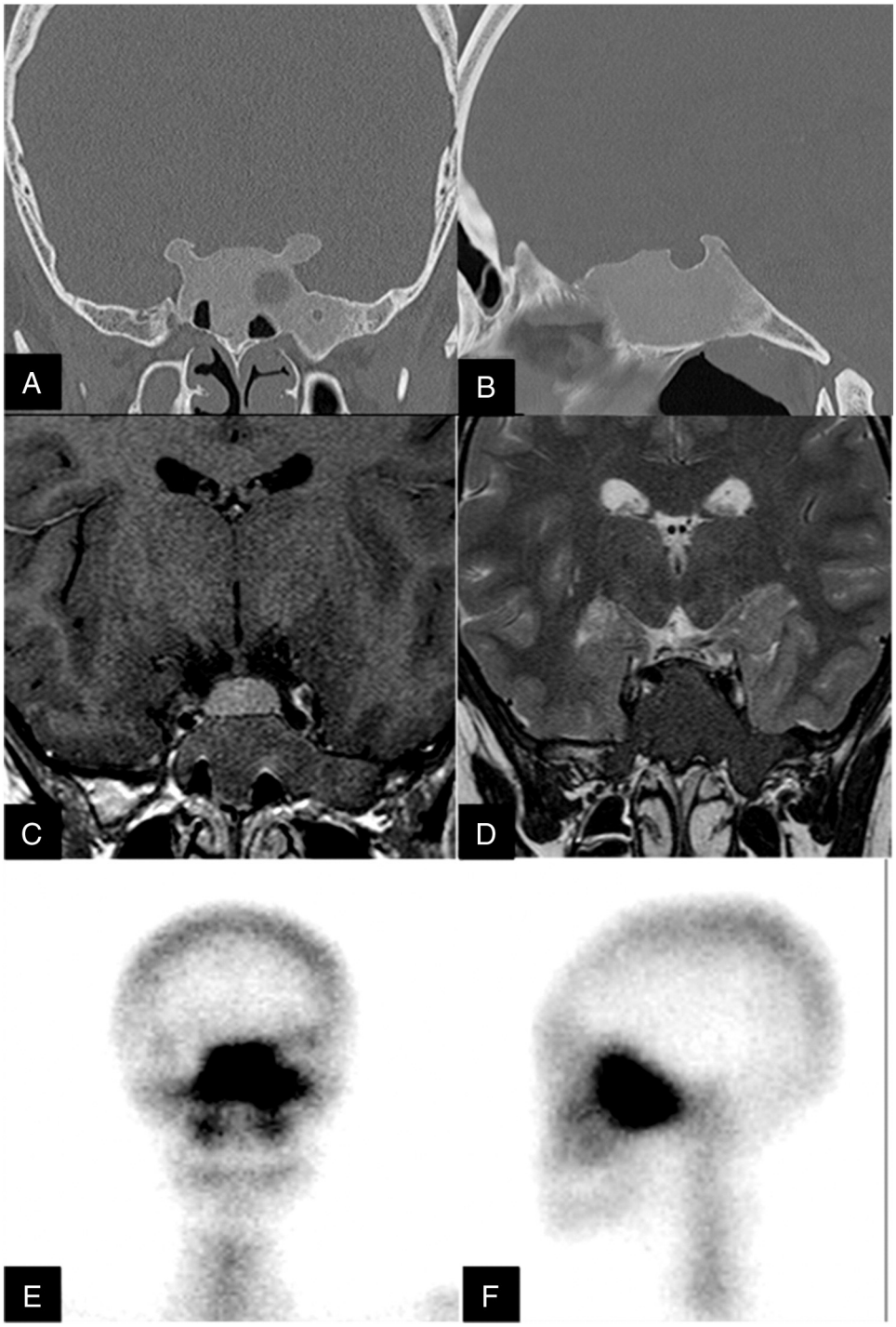

Fig. 1. (A–F) Fibrous dysplasia involving S corpus and left greater wing with ground glass appearance at coronal and sagittal planes from bone windows on CT (1A–B); hypointense signals with T1-weighed without contrast and T2-weighed coronal images on MRI due to fibrous and bone tissue (1C–D); overt radioactive media on the scintigraphic examination (1E–F).

substance involvement in two cases with a scintigraphic examination carried out (Fig. 1E–F). The most observed involvement was of the S bone with seven cases, and the other bone involvements observed were F bone in three cases (Fig. 3A–D), maxillary in five cases, T in two cases, parietal (P) in one case, O in one case, and palatine bone in one case. Simple cystic degeneration was observed in three cases and aneurysmal bone cyst (ABC) in one case (Fig. 4A–F). Maxillary sinus was partially or completely obliterated in five cases, sphenoidal sinus in four cases, ethmoidal in four cases, and F sinus in two cases. In one case, maxillary sinus was completely obliterated with mucocele formation observed (Fig. 5A–D). Perior-bital area was affected in seven cases, while orbital fossa was narrowed in four cases and narrowing of superior orbital fissure and the optic canal were observed in three cases. It led to significant narrowing of the adjacent nasal cavity in two cases. Distribution of the cases according to the clinical and radiological features is described in Table 1.

4. Discussion

FDs are benign fibro-osseous pathologies of developmental disorder of the growing bone in which weak fibrous and osseous