Oral Radiol (2009) 25:135–141
DOI 10.1007/s11282-009-0017-x

CASE REPORT

Fronto-orbital sphenoethmoidal fibrous dysplasia

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Received: 2 April 2009 / Accepted: 25 May 2009 / Published online: 25 June 2009
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Abstract Cranial fibrous dysplasias (FDs) are rare and comprise less than 1% of all primary bone lesions. They may produce cosmetic deformities, peripheral compressive cranial neuropathies, and compressive central neurologic manifestations. We describe an unusual presentation of a fronto-orbital sphenoethmoidal FD in a 32-year-old woman with conventional radiographic, CBCT, and MRI findings. In the head and neck examination, an asymmetry was noticed on the left side, without evidence of adenopathy, paresthesia, or motor nerve deficiency. Panoramic radiographs showed a radiopaque expanded bone in the region of the posterior maxillary sinus and orbita. Computed tomography with three-dimensional reconstruction demonstrated an expanding lesion of the cranial bones, involving the ethmoid and periorbital bone, producing a ground-glass appearance. After the radiologic examination, the patient was referred for surgery with a diagnosis of cranial FD and underwent a cranioplasty. The CT and MRI features were typical for FD, but physicians and dental professionals should be aware of this diagnosis, even if no symptom is apparent and the patient came in only for a routine dental examination. Maxillofacial radiologists should also consider that the signal intensity on both T1- and T2-weighted images and the degree of contrast enhancement on T1-weighted images depend on the amount and degree of bony trabeculae, cellularity, collagen, and cystic and hemorrhagic changes.

Keywords Fibrous dysplasia · Cranium · Computed tomography · Magnetic resonance imaging

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Introduction

Fibrous dysplasia (FD) is the abnormal development of bony tissue that leads to fibrous tissue proliferation and imperfect osteogenesis [1]. Although benign, it is slowly progressive [2, 3] and usually manifests itself in the form of painless asymmetric enlargement and functional problems in the involved area [4]. FD may affect only one bone (monostotic) or more than one bone (polyostotic). A polyostotic lesion combined with cutaneous pigmentation, precocious puberty, and endocrine dysfunction is known as Albright syndrome. FD is caused by a somatic mutation in the protein transcript of the *GNAS1* gene, which leads to an increase in intracellular cyclic adenosine monophosphate. In general, it affects 1 in 4,000 to 10,000 individuals and is rare, with hematomatous, locally expanding, destructive bone lesions comprising less than 2.5% of all primary skeletal lesions. The incidence of cranial FDs is even rarer among primary bone lesions, representing less than 1% [4]. Cosmetic fronto-orbito-facial deformities, sino-orbital and auditory involvement, and compressive neurological manifestations are the main surgical concerns [4–6]. Until recently, many such lesions were largely inaccessible, particularly when they were primarily located at the base of the skull. However, with the development of neuroimaging facilities [magnetic resonance imaging (MRI) and

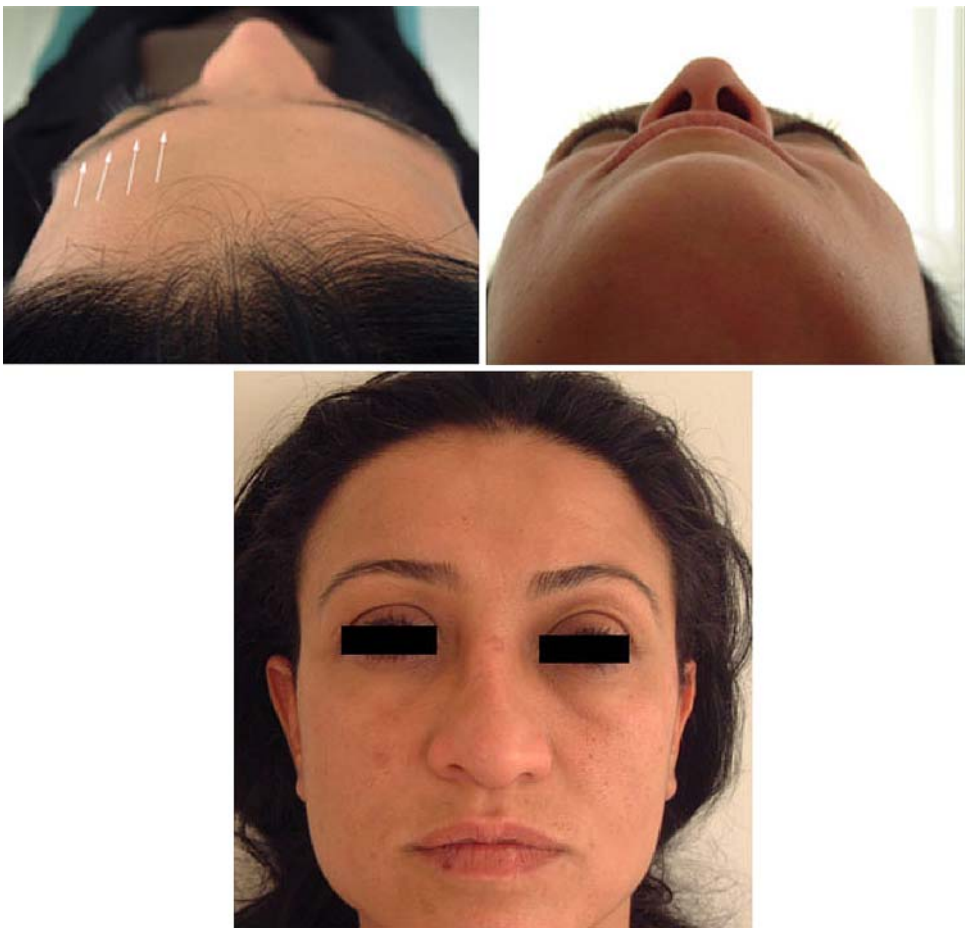

Fig. 1 Preoperative appearance of the patient with asymmetry on the left side

computed tomography (CT)] and the application of current skull-base approaches, more aggressive and radical surgical procedures are being undertaken.

In this presentation, we discuss an incidental finding of a cranial FD in a routine dental examination of a 32-year-old woman, with the results of various imaging methods [cone beam computed tomography (CBCT), MRI, plain radiograph].

Case report

A 32-year-old woman presented at our clinic with a chief complaint of pain in the maxilla from her crown and bridge restoration and in the left temporomandibular joint (TMJ) region, along with ipsilateral headache. The patient's general medical history was unremarkable, and she had no history of trauma. In a head and neck examination, an asymmetry was noticed on the left side (Fig. 1), but no evidence indicated lymphadenopathy, paresthesia, or motor nerve deficiency. Her visual acuity was within normal limits. Regarding the asymmetry, the patient had consulted an ophthalmologist approximately 10 years earlier when she first noticed the asymmetry, but no diagnosis was even made.

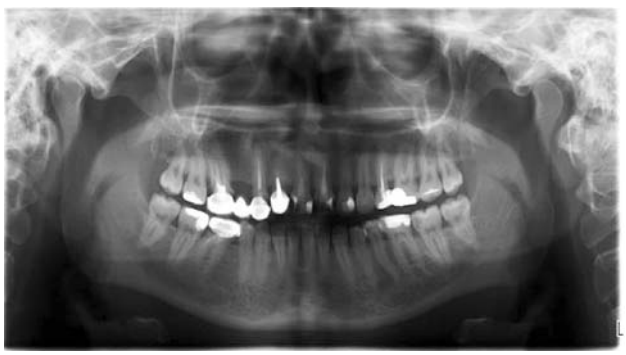

Fig. 2 Panoramic radiograph. The asymmetry is difficult to notice

Intraoral examination revealed multiple porcelain crowns without occlusal interference, and extraoral assessment indicated skin of normal appearance. Sensitivity to palpation was observed in the left TMJ region. The patient expressed no complaints in terms of restrictions on TMJ movement. Initially, a panoramic radiograph was made, which showed a crown and bridge restoration with radiolucencies at the apex region. No abnormality was detected in either TMJ joint (Fig. 2). However, to examine the asymmetry in detail, we decided to perform CBCT (NewTom 3G) with three-

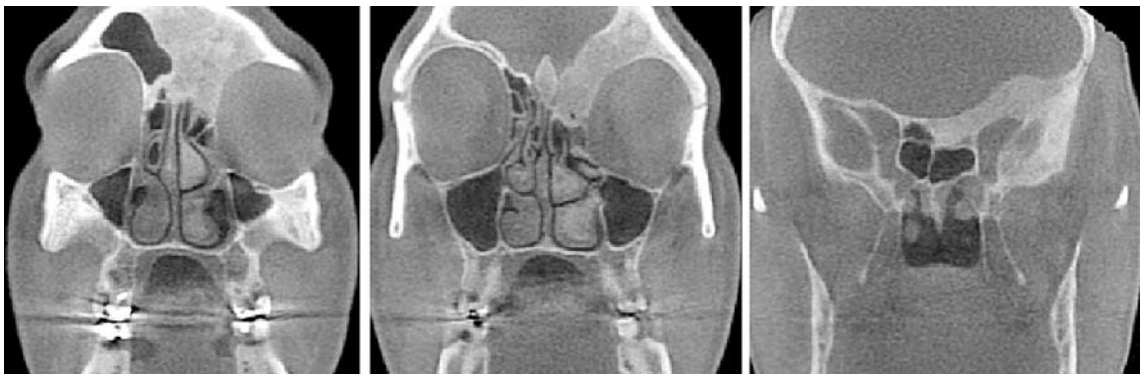

Fig. 3 Coronal CBCT images of the paranasal sinuses show thickening of the frontal bone

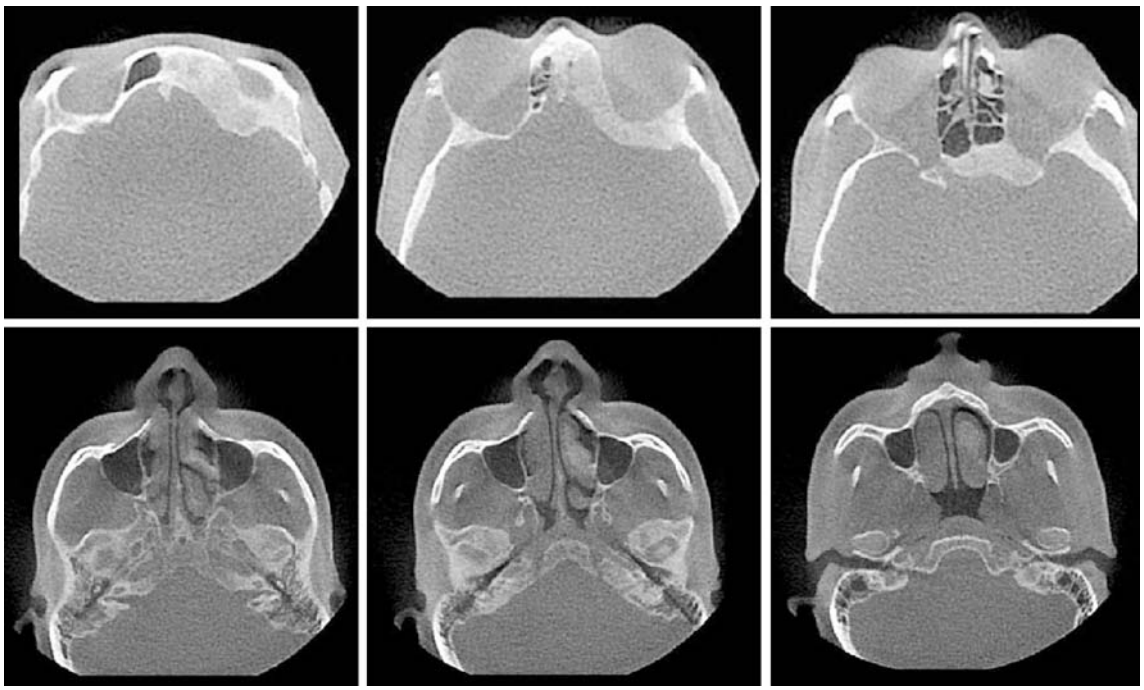

Fig. 4 Axial CBCT of the frontal to ethmoid sinus-level (*upper*) and maxillary sinus-level (*lower*) images show the ground-glass change, with thickened appearance in the frontal, ethmoid (including the crista galli), and sphenoid bones

dimensional (3D) reconstruction to obtain a more precise location and definition of the pathologic features. CBCT imaging demonstrated an expanding lesion of the cranial bones. Axial CBCT images at the frontal to ethmoid sinus level and at the maxillary sinus level showed ground-glass changes, with a thickened appearance in the frontal, ethmoid (including the crista galli), and sphenoid bones. Although the ethmoid bone was affected, the maxillary sinus seemed intact at the levels of these axial CTs (Figs. 3, 4).

However, when examining the maxillary sinus from higher slices of axial CTs (at the zygomatic process of the maxilla level), the superoposterior wall of the maxillary sinus was shown to be affected (Fig. 5a). Axial CT scan also revealed a narrowed left optic canal (Fig. 5b). As the mass involved some of the cranial bones, to detect any soft-tissue changes, the lesion was observed with T1- and T2-weighted and enhanced MRI, which showed increased signal intensity (SI) within the lesion, similar to the muscle SI on both T1- and T2-weighted images. Although T1-weighted images showed a homogenously increased SI, T2 MR images revealed variations in SI as a mixed hypointense-to-isointense signal. The lesion also showed a degree of contrast enhancement (Fig. 6). The optic nerve was intact, but the optic canal began to narrow (Fig. 7).

The patient was referred for surgery with an initial diagnosis of cranial FD. She underwent a cranioplasty, and histopathologic examination after the surgery confirmed the diagnosis of cranial FD. The histopathologic

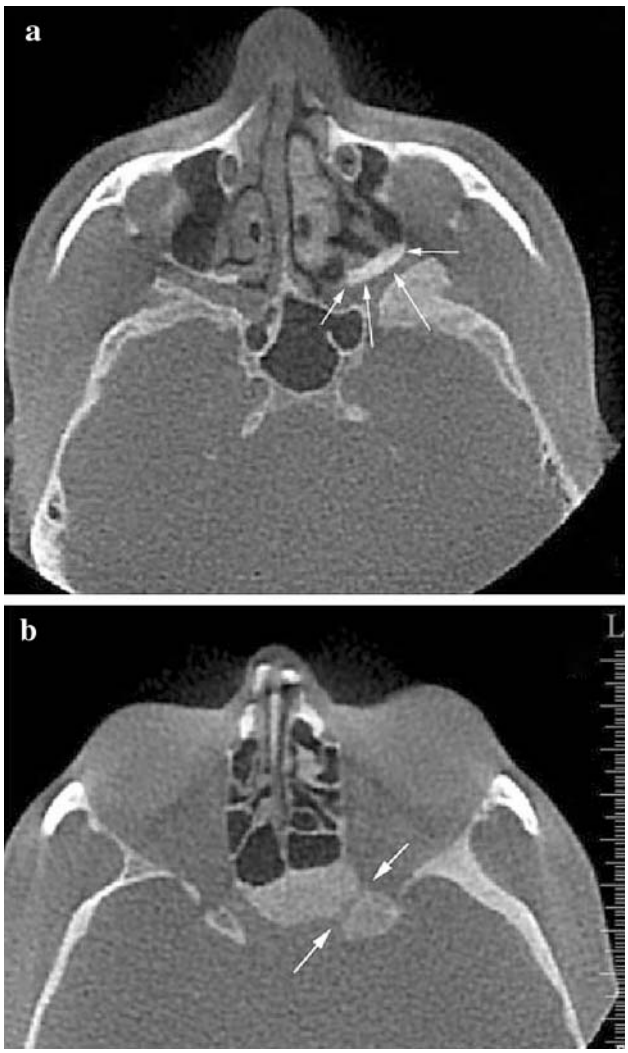

Fig. 5 Axial CBCT scan shows **a** affected bone on the left posterior maxillary sinus wall (*arrows*) and **b** narrowed left optic canal (*arrows*)

examination revealed irregular trabeculae of woven bone, intermixed with connective tissue stroma of differing amounts.

Discussion

FD is a disorder of unknown etiology in which the bone marrow is replaced and subsequently expanded by fibro-osseous tissue. The disease generally affects older children, adolescents, and young adults [3–7]. The more common monostotic form of FD, characterized by single-bone involvement primarily in the extremities and ribs, tends to become quiescent with cessation of growth. Craniofacial involvement, which occurs more frequently in the polyostotic disease, tends to have a typical hemicranial or hemifacial distribution that helps to distinguish FD from Paget's disease, which is often bilateral [1, 8]. Because FD primarily affects the bone marrow, the craniofacial form of the disease is characterized by expansion of the diploic space, with secondary displacement of the outer table and sparing of the inner table. The diploic space widened by fibrocellular connective tissue has a characteristic ground-glass appearance, which is better appreciated with CT than on radiographs. Small sclerotic and lytic areas may be distributed throughout the ground-glass background of the diploic space.

The initial diagnosis in our patient was established on the basis of physical and radiologic examinations. However, a definitive diagnosis was made only after surgery and histopathological examinations. To make an initial diagnosis, the patient's first physical examination was carefully conducted. The slight facial asymmetry on the left side of the front was a clue for the diagnosis of a bone-related disease. In our patient, the lesion was asymptomatic, except the facial asymmetry, with neither headaches nor visual disturbances developing. However, in similar cases, to determine the origin of the lesion, further advanced radiological examinations (especially CT and MRI) need to be performed to rule out the involvement of important anatomic structures, such as the optic canal, which began to become narrowed in this case.

In this case, follow-up of the patient is continuing for the superoposterior wall of the maxillary sinus. This region is partly affected by the lesion and the progression of the lesion through the maxillary sinus should be followed carefully. In this instance, a decision was made to follow-up the maxillary sinus region via panoramic radiography, which can determine if the state of the posterior maxillary sinus wall has worsened.

Although the initial diagnosis in the present case demonstrated that CBCT images were superior in diagnostic efficacy to panoramic images for localizing the extension of the FD, for follow-up of the maxillary sinus region after surgery, CBCT images should not necessarily replace panoramic images. CBCT studies cause higher radiation exposures (4–20 times greater). From the standpoint of radiation risk, CBCT appears to have three to seven times the risk of a panoramic examination, depending on the area examined, the degree of collimation, and the acquisition software. Thus, the decision to select an imaging modality for diagnostic purposes regarding maxillary sinus follow-up should be based on the diagnostic yield expected and in accordance with the as-low-as-reasonably achievable (ALARA) principle [9, 10].

Bone thickening may lead to cosmetic problems, including frontal prominence and proptosis, diplopia, restricted ocular mobility, and several cranial nerve deficits, particularly visual and hearing problems secondary to narrowing of neural foramina [6]. Encroachment on the

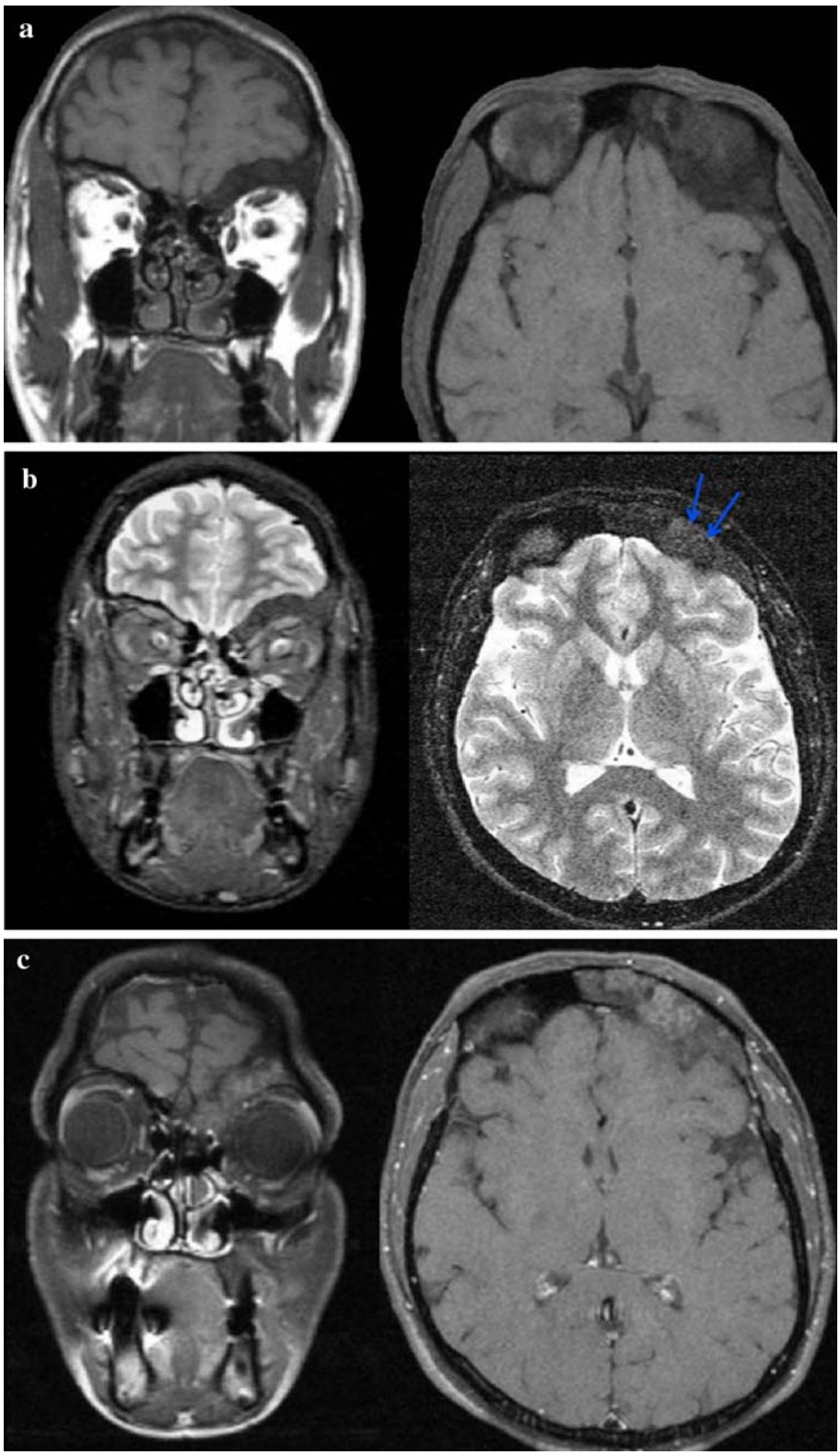

Fig. 6 **a** Axial and coronal T1-W scans reveal homogeneously increased signal intensity of fibrous dysplasia, similar to that in the muscles. **b** Axial and coronal T2-W scans also reveal increased signal intensity of fibrous dysplasia, similar to notable signal changes. **c** After gadolinium enhancement, the T1-W MR image reveals patchy irregular contrast enhancement involving the frontal and ethmoid bones

neural foramina is better evaluated with cross-sectional imaging methods such as CT, and preferably MRI. The affected bones usually have a low-to-intermediate SI on both T1- and T2-weighted sequences [1, 7, 8], probably due to the replacement of the marrow cavity by fibrous tissue. However, the lesions may also have variable SI on T2-weighted images, in which they may appear hyperintense, depending on the activity of the lesions. On

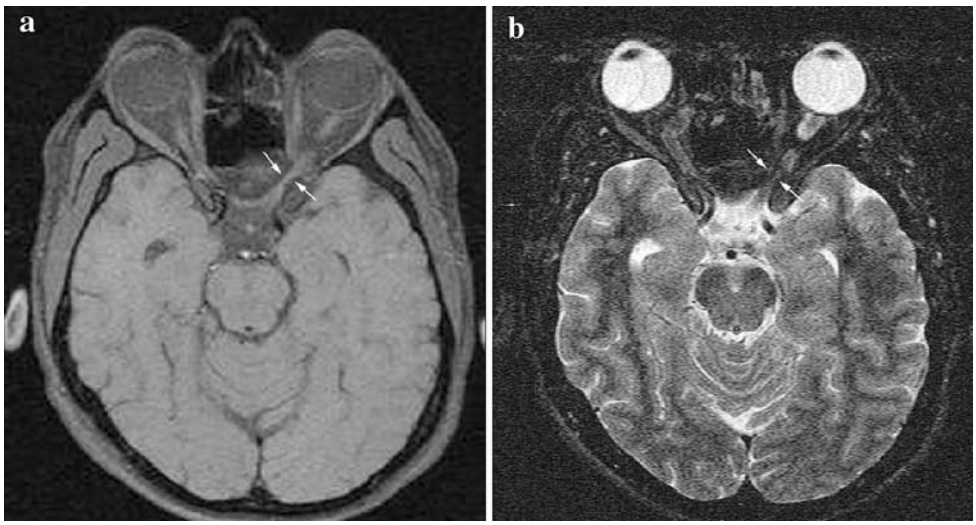

Fig. 7 **a** Axial T1-W and **b** T2-W images show the narrowed optic canal (*arrows*)

postgadolinium MR images, affected bones show variable enhancement, reflecting vascularity of the fibrocellular tissue. Marked enhancement, as seen in this patient, was demonstrated to occur in lesions that showed clinical and histological activity [7].

Chen and Noordhoff [11] classified craniofacial bone affected by FD into four major zones: zone 1, the facial area above the maxillary alveolar bone; zone 2, hair-bearing cranium; zone 3, central cranial base; and zone 4, teeth-bearing bone. The present case falls into zones 1 and 3.

Monostotic FD is about four times more frequent than its polyostotic counterpart, with skull involvement occurring in 50% of polyostotic patients. Any craniofacial skeletal bone may be affected, but 20% of patients have involvement of the fronto-orbital region [4]. According to the literature, if FD is encountered in the cranium, the ethmoid is most commonly involved (71%), followed by the sphenoid (43%), frontal (33%), maxilla (29%), temporal (24%) and occipital (5%) bones [12]. The present case had not only involvement of the fronto-orbital region, but also the sphenoethmoidal bones.

Differential diagnosis must include McCune–Albright syndrome, which presents with the polyostotic form and widespread skull-base involvement, but shows cutaneous pigmentations and endocrinological disorder, which were absent in the present case.

In summary, polyostotic craniofacial FD may rarely involve both sides of the cranium. In addition to the well-known obliteration of the paranasal sinuses, an unusual sinus-related complication such as a mucocele can sometimes be seen. The CT and MRI features were fairly typical for FD, but clinicians must be aware of the potential diagnosis even if no symptoms are observed and patients visit for a routine dental examination. Maxillofacial radiologists should keep in mind that the SI on both T1- and T2-weighted images and the degree of contrast enhancement on T1-W images depend on the amount and degree of bony trabeculae, cellularity, collagen, and cystic and hemorrhagic changes [7].

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