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Monostotic fibrous dysplasia of the clivus: MRI and CT findings

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Abstract Fibrous dysplasia is a developmental disorder caused by abnormal proliferation and maturation of fibroblasts resulting in replacement of mature bone by structurally weak, immature woven bone. Clival involvement in monostotic fibrous dysplasia is extremely unusual, and has rarely been reported previously. We report a case of monostotic fibrous dysplasia of the clivus with special emphasis on the imaging findings and differential diagnosis.

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

Introduction

Fibrous dysplasia is a developmental bone disease first described by Lichtenstein in 1938. It is a benign disease most often seen in the first two decades of life and rarely appears in adults. Craniofacial fibrous dysplasia is a benign disease representing approximately 3% of all bone tumors and 7% of benign tumors. The disorder is usually classified into three forms: monostotic, involving only a single osseous site; polyostotic, involving two or more bones; and Albright-McCune-Sternberg syndrome, one-sided involvement of multiple bones, associated with hyperpigmentation of the skin, and endocrine dysfunction (hyperthyroidism and precocious menstruation in females). Clival involvement by monostotic fibrous dysplasia is rare and there are only two case reports in the literature [1, 2]. We report a new case of monostotic fibrous dysplasia of the clivus with T1- and T2-weighted, and contrast-enhanced magnetic resonance imaging (MRI) examination as well as computed tomographic (CT) findings.

Case report

The patient was a 23-year-old man who had suffered from vertigo and tinnitus for 5 months. Neurologic and physical examination was normal. MRI was performed, and an abnormal lesion was detected in the clivus. MRI indicated low signal intensity on both T1- and T2-weighted images (Figs. 1, 2). After intravenous injection of Gd-DTPA, marked, slightly heterogeneous enhancement of the lesion was demonstrated (Fig. 3). CT examination revealed the ground-glass appearance of the lesion with sclerotic margins without evidence of extra-osseous extension, and the normal appearance of the other craniofacial bones (Fig. 4). On the radionuclide examination there was mildly increased uptake in the lesion, and the involvement was confined to the clivus without evidence of disseminated disease (Fig. 5).

The patient refused a biopsy. One year later at follow-up MRI examination, no difference was detected in the imaging findings.

Discussion

Fibrous dysplasia is a developmental disorder caused by abnormal proliferation and maturation of fibroblasts resulting in replacement of mature bone by structurally weak, immature woven bone. Fibrous dysplasia may be

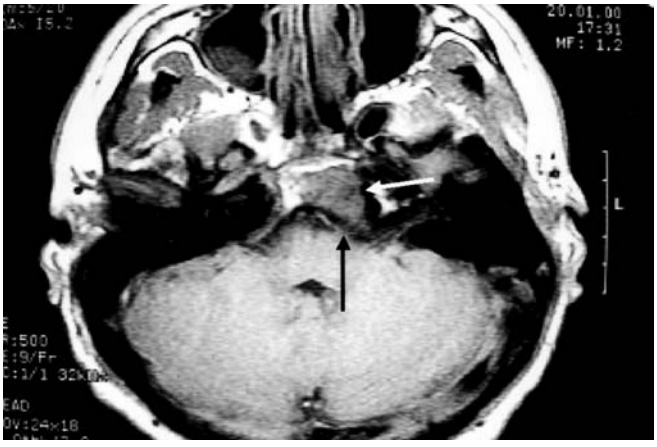

Fig. 1. Axial plane. T1-weighted SE image. There is replacement of the left part of the clivus with hypointense lesion (*arrows*). Note the bulging of the left lateral and posterior walls of the clivus due to the slightly expansile nature of the lesion. The right part of the clivus shows normal fatty marrow

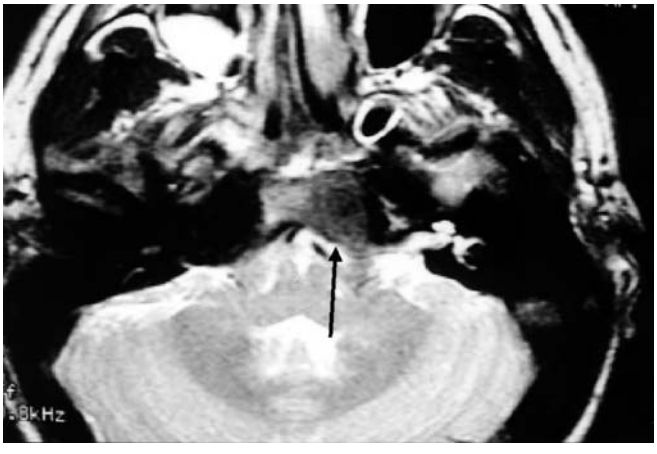

Fig. 2. Axial FSE T2-weighted image. The lesion contains an internal focus more hypointense than the rest of the lesion (*arrow*)

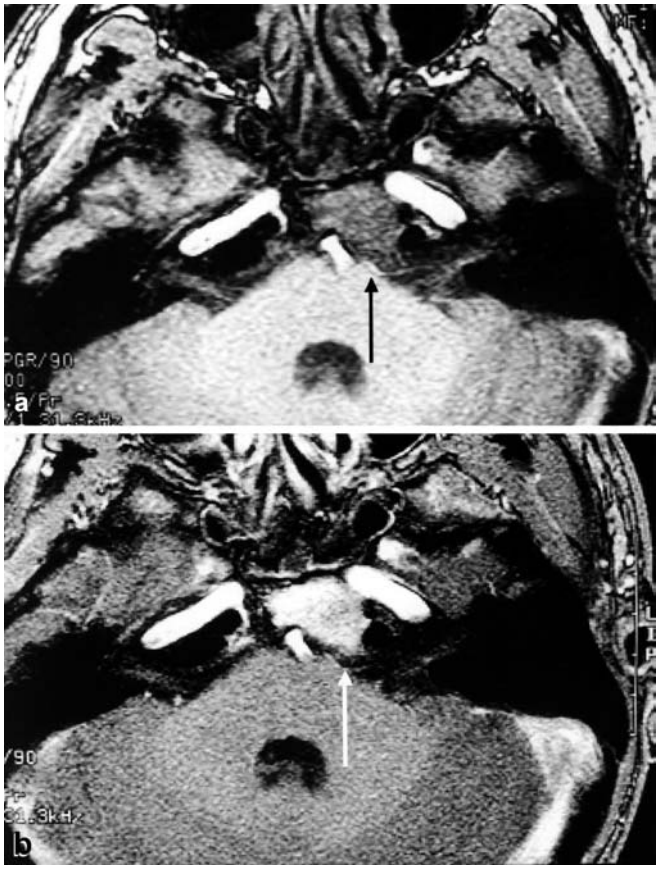

Fig. 3 a, b. Contrast enhancement. **a** Precontrast GE T1-weighted axial image. The lesion is isointense relative to grey matter (*arrow*). **b** Contrast-enhanced axial T1-weighted GE image showing marked enhancement. The left part of the lesion is slightly less enhanced than the right (*arrow*)

present in the monostotic (70%) or polyostotic form (30%) with or without Albright-McCune-Sternberg syndrome. The monostotic form involves the long bones, with most lesions located in the femur and tibia. The common sites affected by the monostotic form in craniofacial bones are the frontal, sphenoid, ethmoid, orbit, zygoma, maxilla, mandible, and temporal bones. However, monostotic fibrous dysplasia of the clivus is extremely rare, and has been reported only twice [1, 2].

The imaging findings of fibrous dysplasia have been well described by means of scintigraphy, radiography, and CT. The MRI features of fibrous dysplasia have also been reported in the literature [3, 4, 5]. Fibrous dysplasia shows low signal intensity on T1-weighted images, while the signal intensity on T2-weighted images varies from high to intermediate or low. This variation may reflect the overall cellularity, collagen content, extent of bone trabeculae, and cyst formation, all of which determine the MRI appearance of a lesion. In Jee et al.'s study of 13 proven cases of fibrous dysplasia, 38% showed hypointensity and 62% showed hyperintensity on T2-weighted images. Histopathologic examination of T2-hyperintense cases revealed fewer bony trabeculae, less cellularity and fewer collagen fibers than did the histopathologic examination of T2-hypointense cases [5]. Metabolically active lesions are also expected to have a prolonged T2 relaxation time similar to that seen in other actively growing tissues. Our case showed low signal intensity on T2-weighted images, which resulted in the exclusion of many pathologic entities involving the clivus (Fig. 2). After the administration of Gd-DTPA, marked and slightly homogeneous contrast fixation was detected (Fig. 3b).

To the best of our knowledge, only two cases of fibrous dysplasia confined to the clivus have been reported previously [1, 2]. Levy et al. [1], in their case report, described only the T1 characteristics of the

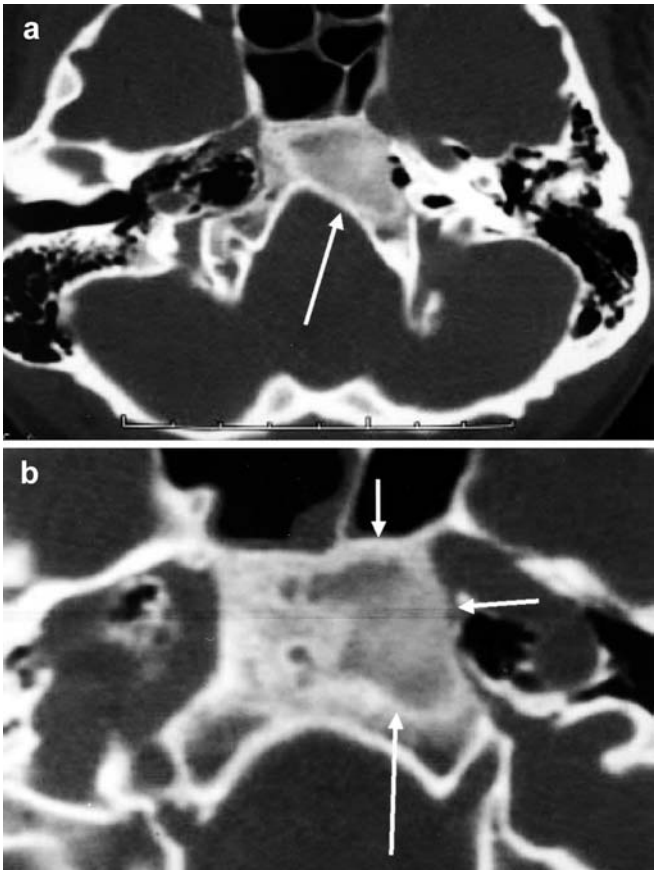

Fig. 4 a, b. CT scan. **a** Ground-glass appearance of the lesion with minimal expansion (*arrow*). **b** Image more cranial than image **a**. Note the sclerotic border of the lesion (*arrows*)

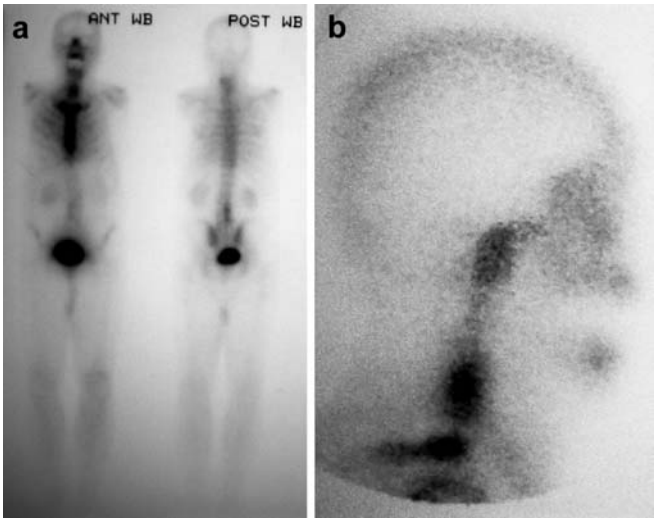

Fig. 5 a, b. Radionuclide evaluation. **a** Whole body bone scan confirming that the disease is not disseminated. **b** Lateral view reveals mild uptake of the tracer by the clival lesion

lesion. In their case, the diagnosis of fibrous dysplasia was confirmed by histopathologic examination [1]. T2 findings and contrast-enhanced images of monostotic fibrous dysplasia of the clivus have been reported in only one previous case [2]. The lesion in that report showed low signal intensity on T2-weighted images and revealed marked enhancement after intravenous injection of Gd-DTPA. Transnasal biopsy confirmed the diagnosis of fibrous dysplasia in that case. The case reported by Maeda et al. [6] in 1993, seems to be the identical case reported by Sato et al. [2] in the same year.

Most abnormalities affecting the clivus such as chordoma [7, 8], chondrosarcoma [8], giant cell tumor [9], cavernous hemangioma [10], lymphoma [7], and most metastases, exhibit low signal on T1-weighted and high signal on T2-weighted images. Ecchordosis physaliphora, which is a rare, benign lesion originating from notochordal anlage, has been described as hyperintense on both T1- and T2-weighted images [11]. Craniopharyngiomas may be infrasellar and primarily intraosseous, but they are not completely confined to clivus and do not show pure intraosseous localization. Pituitary adenomas may sometimes invade the sphenoid sinus and partly the clivus; however, similar to craniopharyngiomas, they do have extraclival components and a separate pituitary gland can not be seen.

Clival lesions showing low or intermediate signal intensity on T2-weighted images are rare. On T2-weighted images, the signal intensity of the clivus can be low due to sclerotic reaction in patients with periclival meningioma [7]. This differential diagnostic possibility could easily be eliminated in our patient by the lack of a lesion compatible with meningioma. Sclerotic metastasis is another consideration; however, solitary osseous metastasis in a 23-year-old patient without a known primary disease would be highly unlikely. Lymphoma, which has a tendency to show lower signal intensity on T2-weighted images, would also be unusual as a solitary lesion. Additionally, non-progression of the lesion after a the interval of a year easily excluded these two diagnostic possibilities.

For our patient, the differential diagnosis of fibrous dysplasia was primarily with ossifying fibroma. In ossifying fibroma, a peripheral rim of sclerosis is frequently present [12]. Histologically, ossifying fibroma and fibrous dysplasia may be indistinguishable, so we think that lack of histopathologic diagnosis was not a major failure in our case. Osteogenic sarcomas may affect the craniofacial region with jaw predilection [12]. Lack of change in the volume and characteristics of the lesion at follow-up MRI, which was done 1 year after the first examination, easily excluded this diagnostic possibility. We had recommended biopsy of the lesion to our patient because of the rarity of monostotic clival involvement of fibrous dysplasia; however, at present we think that it may not be necessary to obtain a biopsy in a lesion

which looks typical for fibrous dysplasia on CT images. Therefore, CT is of the utmost importance for clival lesions (especially T2-hypointense lesions) to obtain more specific information on the type of pathology.

The management of fibrous dysplasia is not surgical unless it causes unacceptable or progressive deformity, cranial nerve compromise, pain, or development of a malignancy. Usually small, non-expansile solitary lesions will remain unchanged. Although the prognosis of fibrous dysplasia is generally good, malignant degeneration and aggressive behavior have been described. The incidence of malignant transformation is highest for monostotic craniofacial lesions (0.05%). Osteosarcoma, fibrosarcoma, and chondrosarcoma are the malignancies reported in the literature. We recommended a yearly follow-up for our patient, and the first follow-up MRI study, requested by the patient's physician, revealed no change in signal intensity pattern or morphology. A CT follow-up, however, would be more appropriate for detection of any subtle changes in the morphology and internal structure of the lesion.

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