

Monostotic Fibrous Dysplasia of the Clivus: Imaging Findings

Klivusun Monostotik Fibröz Displazisi: Görüntüleme Bulguları

ABSTRACT

Fibrous dysplasia (FD) is an uncommon benign disorder of unknown etiology. It most likely represents a disorder of normal bone development. The disease can involve any bone in the body. In the head and neck, the skull and facial bones are involved in 10–25% of cases of monostotic FD. Involvement of the clivus in monostotic FD is extremely unusual, and has rarely been reported previously. The diagnosis is based on radiological images. We retrospectively reviewed our database and identified three patients with the diagnosis of FD of the clivus. We reviewed the clinical and radiological findings of these patients in this paper.

KEYWORDS: Fibrous Dysplasia, Monostotic; Tomography, Spiral Computed, Magnetic resonance imaging, Cranial Fossa, Posterior

ÖZ

Fibröz displazi (FD), etiyolojisi bilinmeyen nadir benign bir bozukluktur. Sıklıkla normal kemik gelişiminin bir bozukluğuna bağlıdır. Vücutta herhangi bir kemiği etkileyebilir. Baş ve boyun bölgesinde kafa ve yüz kemikleri, monostotik FD'li olguların %10-25'inde tutulabilir. Monostotik FD'de klival tutulum oldukça nadir olup seyrek olarak bildirilmiştir. Tanı, radyolojik görüntüler temelinde konulmaktadır. Biz geriye dönük olarak verilerimizi değerlendirdik ve klivus yerleşimli FD tanısı alan üç hasta saptadık. Bu yazıda klivus yerleşimli FD tanısı alan üç olgunun klinik ve radyolojik bulguları ortaya konuldu.

ANAHTAR SÖZCÜKLER: Fibröz displazi, Bilgisayarlı tomografi, Manyetik rezonans görüntüleme, Posterior kranial fossa

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INTRODUCTION

Lichtenstein coined the term fibrous dysplasia (FD) in 1938 to describe a disorder characterized by the progressive replacement of normal bone elements by fibrous tissue. It is a benign disease most often seen in the first two decades of life and rarely appears in adults. FD or fibro-osseous dysplasia is a developmental disorder of growing bones of unknown etiology. FD is a well-known disease that affects the craniofacial skeleton. It occurs in two distinct forms: polyostotic, involving several bones and accounts and making up 30% of the cases and monostotic, which involves a single bone and constitutes 70% of the cases (1,2,12).

Fibrous dysplasia of the clivus is an unrecognized entity and we could find only sixteen reported cases of clival FD in our review of the literature (1-4,8,9,11,12). We herein present three new cases of monostotic FD of the clivus with computed tomography (CT) and magnetic resonance imaging (MRI) findings.

REPORT of CASES

We identified three patients (two females, one male) with fibrous dysplasia of the clivus. The patients ranged in age from 13 year to 34 years. The written consent to analyze the patients' files was approved by our institutional review board for human investigation. The files of these patients were analyzed retrospectively. The patient's age, gender, and symptoms at presentation were noted. All patients underwent physical and neurological examinations. The most common presenting symptom was headache. One patient had vertigo and tinnitus. Laboratory tests, which included serum calcium, phosphorus, and alkaline phosphatase, were normal. No abnormality was found on clinical examination of the chest, abdomen, and cardiovascular system.

Cross-sectional imaging (including CT and/or MRI) examinations were performed in all patients to elucidate the underlying pathologic abnormality. CT scan was performed in three patients. Cranial CT examinations were performed with a spiral CT scanner (Picker, PQS, OH, USA). CT images were obtained in the axial plane. All patients underwent MRI. MRI examinations of the brain were performed on a 1.5 Tesla MR magnet (Excelart, Toshiba, Tokyo, Japan) with a standard head coil. The MR images



Figure 1: CT images. (A) 13-year-old boy: Noncontrast axial CT image on bone window shows an expansile hyperdense lesion involving the left occipital condyle (white arrows), (B) 22-year-old woman: Axial CT scan on bone window demonstrates asymmetrical extension of the disease to the right condyle and the characteristic "ground glass" appearance (white arrows).

were obtained in three planes (axial, sagittal, and coronal) with T1-weighted (T1W) spin echo (SE) (repetition time [TR] = 440 msec, echo time [TE] = 15 msec, flip angle [FA] = 90°), and T2-weighted (T2W) fast SE (FSE) (TR=5000 msec, TE =90 msec, FA=90°).

Table I: Clinical and radiological findings in three patients with fibrous dysplasia of the clivus.

Patient No.	Age (Yr)/Sex	Presentation	Neurological examination	Symmetry extension	Radiological findings				Radiological follow-up
					CT	MRI		Gd-enhanced MRI	
						T1W	T2W		
1	13 y, M	Headache	Normal	Asymmetric, to left condyle	Groundglass appearance	Homogeneous Hypo/isointense	Hypointense	Mild	No change on imaging
2	22 y, F	Headache, vertigo, tinnitus	Normal	Asymmetric, to left condyle	Groundglass appearance	Homogeneous Hypo/isointense	Hypointense	Mild	No change on imaging
3	34 y, F	Headache	Normal	Asymmetric, to right condyle	Groundglass appearance	Homogeneous Hypo/isointense	Hypointense	Moderate	No change on imaging

Yr: year, F: Female, M: Male, CT: computed tomography, MRI: magnetic resonance imaging, Gd: gadolinium.

Gadolinium-based contrast material [gadolinium diethylenetriaminepentaacetic acid (Gd-DTPA)] was administered. The MRI appearances were noted on T1W, T2W and postcontrast T1W images. CT and MRI scans were evaluated for symmetry of the lesion, sinus involvement, and appearance of cortical tables. All lesions had a ground-glass appearance on CT images. All the patients had asymmetrical lesions with extensions into one of the

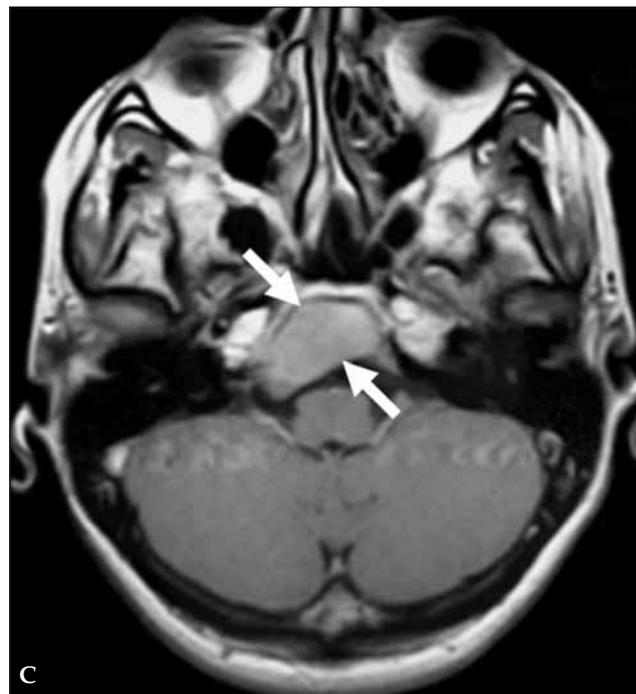
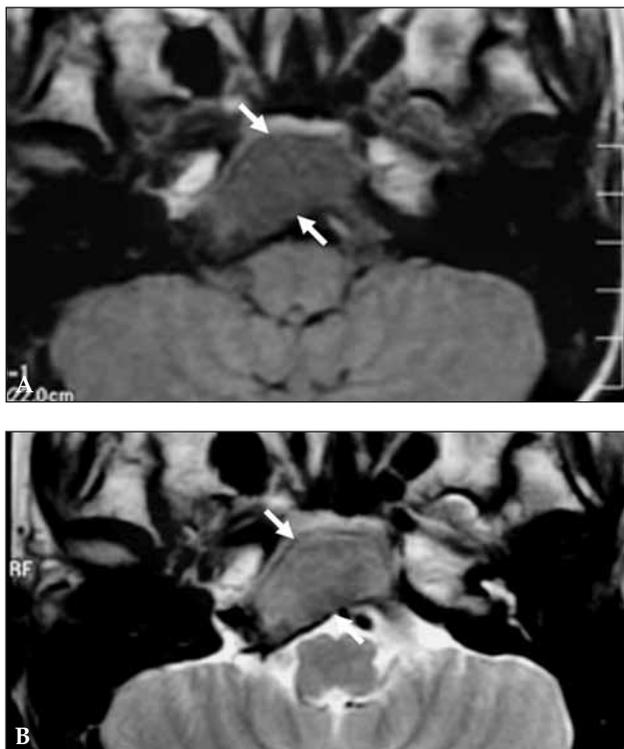


Figure 2: MR images of a 34-year-old female patient. (A) axial T1W image, (B) axial T2W image, and (C) axial gadolinium-enhanced T1W image. The lesion shows low signal on the T1W and T2W images, but increased signal after gadolinium enhancement compared with T1W images without enhancement (thick white arrows).

occipital condyles (Figures 1A,B). The posterior wall of the sphenoid sinus was involved in two cases. Both cortical tables were preserved in all the patients.

MRI demonstrated the lesions to be homogeneously hypo- and isointense to normal

bone and skeletal muscle on FSE T1W and hypointense to normal bone and skeletal muscle on SE T2W images in all patients (Figures 2A,B). All lesions showed some degree of enhancement on postcontrast T1W images (Figure 2C).

The patients refused a biopsy. No difference was detected in the follow-up CT imaging findings one year later. The demographic and clinicoradiological data on our patients are presented in (Table I).

DISCUSSION

FD is a developmental disorder caused by abnormal proliferation and maturation of fibroblasts resulting in replacement of mature bone by structurally weak, immature woven bone (12). FD may be 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 FD of the clivus is extremely rare. Large series of craniofacial FD have not reported the involvement of the clivus by this entity. Recently, neuroradiologists increasingly encounter patients with clival lesions discovered incidentally on CT or MRI scans. Surgical intervention must be considered despite the minor clinical findings as the common clival lesions are neoplastic in nature and some are grave in prognosis (5,6,10-12).

The imaging findings of FD have been well described by means of scintigraphy, radiography, and CT. CT scan shows the characteristic appearance of thinning of the cortical bone and expansion of the affected area with "ground glass" density (11). The MRI features of FD have also been reported in the literature (12). However, the MRI appearance may not be as specific as the CT. FD shows low signal intensity on T1W images, while the signal intensity on T2W images varies from high to intermediate or low. The variation in MRI appearances may be due to the overall cellularity, collagen content, extent of bone trabeculae, and cyst formation (11,12). In Jee et al.'s (5) study of 13 proven cases of FD, 38% showed hypointensity and 62% showed hyperintensity on T2W 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 (3). Metabolically active lesions are also expected to have a prolonged T2 relaxation time similar to that seen in other actively growing tissues. Our cases showed low signal intensity on T2W images, which resulted in the exclusion of many pathologic entities involving the clivus.

Our extensive literature search on monostotic FD of the clivus revealed only sixteen cases on this subject. Levy et al. (8), in their case report, described only the T1 characteristics of the lesion. In their case, the diagnosis of FD was confirmed by histopathologic examination. Maeda et al. (9) in 1993 reported a case with monostotic FD of clivus. A similar case has been reported by Sato et al. (11) in the same year. In 1998, Ham et al. (3) described two cases of craniofacial FD (one patient had a clival fibrous dysplasia, the remaining one had a FD of the sphenoid sinus). Itshayek et al. (4) reported a case with FD in combination with aneurysmal bone cyst of the occipital bone and the clivus. Sirvanci et al. (12) in 2002 described a case of monostotic FD of the clivus. A report by Adada and Al-Mefty (1) described eight patients who had experienced FD of the clivus. Four of the patients in their series had pathological confirmation. In 2006, Contratti et al. (2) presented a rare case of a polyostotic FD of the clivus in a young woman with clival and vertebral involvement.

The usual radiological differential diagnosis of clival lesions includes chordoma, chondrosarcoma, and plasmacytoma (1,12). Other disease processes known to involve the clivus are giant cell tumors, hemangiomas, lymphomas, carcinomas (adenocystic or nasopharyngeal), metastasis, and Paget's disease (1,10,12,13). Most abnormalities affecting the clivus exhibit low signal on T1W and high signal on T2W images. Echordosis physaliphora, which is a rare, benign lesion originating from notochordal anlage, has been described as hyperintense on both T1 and T2W images (12). 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 cannot be seen. Clival lesions showing low or intermediate

signal intensity on T2W images are rare. On T2W images, the signal intensity of the clivus can be low due to sclerotic reaction in patients with periclavial meningioma (12). 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. Lymphoma, which has a tendency to show lower signal intensity on T2W images, would also be unusual as a solitary lesion. Additionally, nonprogression of the lesion after an interval of a year easily excluded these two diagnostic possibilities (6,7,10).

The management of FD 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 FD 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. Asymptomatic patients should be followed regularly to safeguard against false diagnosis. The follow-up should be conducted yearly and should include a CT scan of the cranial base and clinical evaluation. Using flexion and extension x-rays of the cervical spine and thin-cut CT scans of the craniovertebral junction should be considered in patients with symptomatic involvement of the condyles (3,4,7,8). We think that it may not be necessary to obtain a biopsy in a lesion that looks typical for FD on CT and MR images. However, biopsy might be needed if the diagnosis is doubtful or malignant transformation is suspected.

In conclusion, CT is extremely useful for demonstration of the gross bony changes as well as anatomical details. However, MRI offers a higher contrast differentiation between intralésional components. We believe that the clinical and radiological diagnosis of FD of the clivus should be carefully and cautiously considered since it is a benign, slow growing lesion. Moreover, the patients should always be informed about requiring periodical follow-up of disease.

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