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Case Report

Fibrous Dysplasia of the Clivus

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Fibrous dysplasia (FD) of craniofacial structures is well documented, however, its involvement of the clivus is seldom described. We report a case of clival FD in a young man who presented with headache localized to the occipital area. The radiological studies revealed a monostotic disease confined to the clivus, with typical findings of hypointensity on magnetic resonance images and ground-glass density on computed tomography. The diagnosis of FD was confirmed on pathological examination of specimens taken through transsphenoidal surgery. The patient showed reduction of symptoms and no change of residual lesion on follow-up imaging taken 2.5 years later after surgery. This study includes clinical aspect, radiographic appearance, differential diagnosis and treatment strategy of this rare skull base lesion.

KEY WORDS : Skull base· Clivus· Fibrous dysplasia· Magnetic resonance imaging.

INTRODUCTION

Fibrous dysplasia (FD) is a developmental disorder caused by abnormal proliferation of fibroblasts resulting in replacement of normal cancellous bone by structurally weak, immature osseous tissue. This process of unknown etiology, which mainly affects the younger population, may be monostotic, polyostotic or associated with McCune-Albright syndrome[14]. The monostotic FD usually involves the temporal bone, orbit, mandible, and paranasal sinuses in craniofacial region, but FD with isolated clivus involvement is exceptional. Previously, only a few cases of FD of the clivus have been reported in the literature[4,10].

FD might be confused with other disease involving the clivus, for which a more aggressive surgical approach is generally advised. Furthermore, with advancement and wide use of computed tomography (CT) and magentic resonance (MR) imaging, many of clival FD can be detected incidentally. The awareness of and differentiating it from other skull base lesions is central in managing the patients with FD in the clivus. In this report, clinical, radiological, and histopathological features of clival FD are reviewed in detai and implication for the role of surgery is also discussed.

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CASE REPORT

A 22-year-old man visited emergency room with complaint of severe headache on occipital area that worsened on moving his neck. He was intact neurologically and physically. Plain skull radiographs demonstrated an expansile bony pathology that filled the posterior sphenoid sinus and the clivus, extending to the occipital condyle. CT scan showed a dense mass measuring 35×28×43 mm with a typical ground-glass opacification and bony sclerosis. This lesion was appeared as hypointense and a mixture of hyperintense and hypointense areas on T1-weighted and T2-weighted MR images respectively, with moderate contrast enhancement (Fig. 1). Whole body bone imaging with technetium-99m methyl diphsophonate revealed an increased radioactivity in the central skull base. Fluorodeoxyglucose positron emission tomography (PET)-CT indicated a focal area of high uptake of tracer within the lesion (Fig. 2). A transsphenoidal approach for establishing the pathologic diagnosis and partly excising the clival tumor was attempted. The tumor was white-grayish in color, rubbery or bony hard in consistency, and was removed subtotally with blade and diamond drill. Cyst was not seen. The tumor was fibrotic and osseous in nature on gross and microscopic examination (Fig. 3). Postoperatively, the patient experienced resolution of his headaches. Suboccipital and

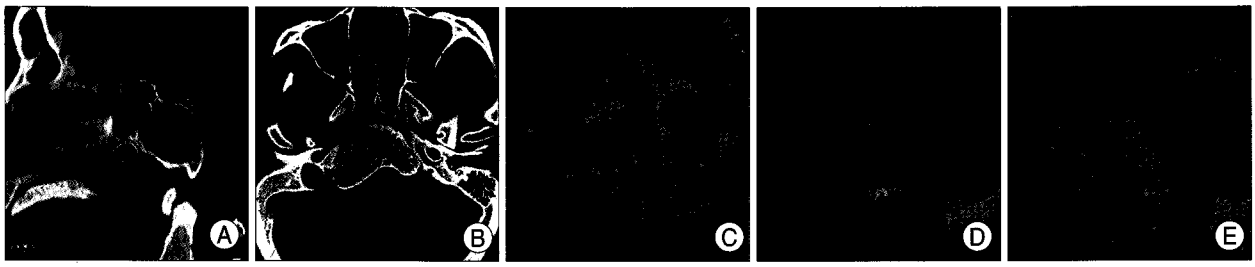

Fig. 1. Computed tomography scans and magnetic resonance images of a young adult with clival fibrous dysplasia. Note the intact cortical tables and the ground-glass appearance (A and B). The lesion is partially hyperintense on T2-weighted image (C), and hypointense and moderately enhanced on T1-weighted images (D and E).

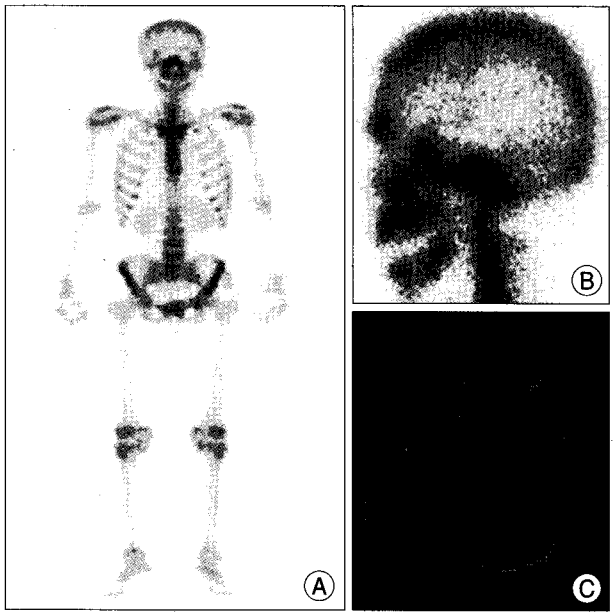

Fig. 2. Nuclear medicine studies of solitary fibrous dysplasia. Radionuclide scan confirms that involvement is confined to the clivus without evidence of disseminated disease (A). There is intensely increased uptake in the lesion (B). Positron emission tomography scan with fluorodeoxyglucose detects accumulation in the lesion (C).

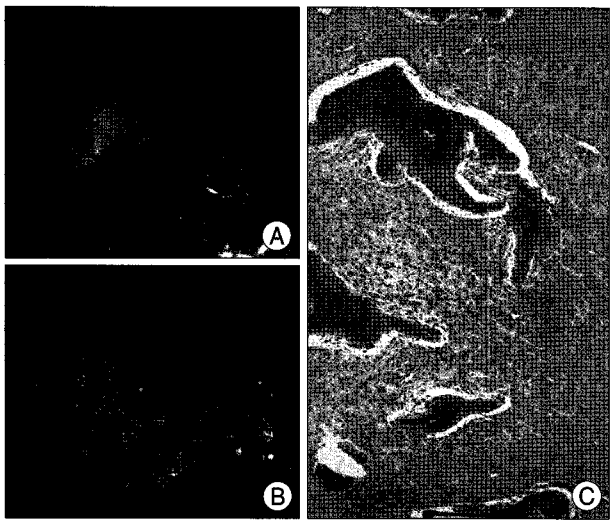

Fig. 3. Operative and photomicrographic features of the fibrous dysplasia. The tumor is smooth-surfaced, white and of a firm, rubbery consistency (A and B). The specimen shows mainly a fibrous matrix and immature bony trabeculae mimicking Chinese letters (C).

nuchal pain was also relieved by head immobilization and applying the neck brace. After more than 2.5 years of follow-up, the patient was symptom-free and the residual clival lesion did not grow.

DISCUSSION

Histopathogenetic studies of FD recently proposed that a defect in the osteoblastic differentiation and maturation, linked to a genetic mutation in the chromosome 20, results in the anomaly of the mesenchymal precursor of bone[24]. FD is classified as a non-heritable, non-neoplastic disease of the bone, however, it might be locally invasive and destructive, so can lead to bony expansion, weakness, and distortion. Craniofacial skeletons are affected in 30% of patients with monostotic disease and in 50% of patients with polyostotic FD, with a predilection for the orbit, sphenoid, frontal, ethmoid, temporal, and maxilla bones[11,14]. Clival involvement by monostotic FD, often misdiagnosed entity, is extremely rare, and there are only eight cases even in the largest series reported on patients with FD of the clivus[1]. But, among them just four clival lesions were confirmed pathologically.

In cranial FD, as a developmental disorder with a benign clinical course, monostotic lesion usually ceases to progress or evolves after adolescence. However, in exceptional case, it continues to grow even after this period, and it is unclear under what kind of conditions this growth continues[17]. Furthermore, update report did not find presentation in adulthood to be rare, because 39-50% of patients in that series became symptomatic during adulthood[1,6,9]. Continued growth into adulthood occurs and is more often observed in males than females[6]. Therefore, despite of the stable disease after subtotal excision, the present case need to be more carefully followed clinically and radiographically.

For the patients with FD, clinical symptoms and signs result from the progressive bone deformation that develops several years, and depend on the location and extension of the abnormality. A localized painless expansion of the skull on one side is the commonest difference noted first by patients with craniofacial FD[14,24]. This bone swelling can lead to

intracranial extension and central neurological manifestations, neurovascular entrapment, orbito-sino-nasal obstruction, recurrent sinusitis, epiphora, proptosis, malocclusion, and cosmetic deformity[3,5,17,25]. On involvement of basal cranium by FD, compressive cranial neuropathy that mainly affect the facial, auditive, trigeminal, olfactory, and visual function occurs from thickening and narrowing of neural canals and foramina[1,2,9,16,18]. Majority of the monostotic clival dysplasia do not appear to produce any functional limitations and aesthetic problems, and are mostly identified incidentally in early adulthood[3]. On rare instances, however, this is symptomatic, and presented with headache, neck pain, isolated sphenoid sinus tumor, craniocervical instability, or with hypoglossal nerve palsy[1,15]. As shown in the our case and other reports, the presence of localized headache in occipital area correlated with the extension of the pathology process to the occipital condyle.

The present case highlights the need to consider FD in the differential diagnosis of isolated clival lesions. Its diagnosis relies on image examination and pathology. High-resolution CT is the imaging tool of choice for evaluation of FD. Depending on the volume of osseous and fibrous tissues, three patterns have been extensively described in FD; sclerotic, cystic, and pagetoid. The FD involving the cranial base tends to be a sclerotic variety, where the inner table gets thickened, therefore, a peduncular or sessile intracranial extension is commonly seen[19,21]. A "ground-glass" opacity, ballooning and expansion of the affected bone, thinning of the cortex are the hallmarks of FD. These classic CT findings have been true in cases of clival FD on the prior studies[10,15]. The MR appearance of FD is determined by the collagen content, overall cellularity, bony trabeculae, degree of mineralization, disease activity, and cyst formation. Almost all FDs show hypointensity on T1-weighted images, while the signal intensity on T2-weighted images varies from high to intermediate or low[11,12]. On the contrary, most common abnormalities occupying the clivus such as chordoma, plasmacytoma, chondrosarcoma, giant cell tumor, hemangioma, lymphoma, carcinomas, Paget's disease, and sclerotic metastases, exhibit hypointensity on T1 and hyperintensity on T2-weighted images[19,20]. In addition, clival bone lesions showing low or intermediate signal intensity on T2-weighted MR imaging are rare. Consequently, differential diagnostic possibilities above mentioned could easily be excluded by radiographic characteristics observed in the present case, the "ground-glass" appearance on CT scan and hypointensity in T1- and T2-weighted MR imaging. Unusually, foci of actively proliferating and growing portion within the lesion[22,23], as clearly depicted on our PET-CT scanning, might be responsible for iso- or hyperintensity on T2 MR sequence.

The management of craniofacial FDs is strictly surgical when they cause cosmetic deformation, intractable pain, disease progression, compressing cranial neuropathy, association with aneurysmal bone cyst, or malignant degeneration[1,7]. It is best to perform bone contouring surgery subsequent to growth arrest for the patient with monostotic disease. When growth continues indefinitely in patients with the polyostotic type, recurrence of the disease and the resultant deformities are predicted, so complete resection and reconstructive surgery is recommended[9]. But, whole excision is rarely feasible since the lesions tend to be fibrotic and obscure anatomic landmarks. Bisphosphonate therapy may help to improve function, decrease pain, and lower fracture risk in appropriately selected patients with cranial FD[4,24].

Because of its very slow potential growth, most FD of the clivus, which is monostotic and asymptomatic, can be usually treated with clinical observation and patient education[2]. Nevertheless, management of clival FD is considered to be a great challenge because of its nonspecificity of the presenting symptoms and inaccessibility of the lesion during routine operation[3]. As literature stated, the diagnosis is generally clear after a proper radiological and scintigraphic workup, most cases do not require the confirmatory biopsy[1,14]. Infrequently, surgical intervention must be considered for the lesion which has an uncertainty in CT and MR images, despite the minor or no clinical findings, because the common clival lesions are neoplastic in nature and some are grave in prognosis[13,21]. In clinical practice, small isolated clival FD with no expansion usually will remain unchanged, therefore, annual survey for the lesion is recommended if the first follow-up imaging revealed no change in signal intensity or morphology[8]. But, in cases of cranial nerve impingement or extension to the condyles, surgery should be considered. Clival FD without extension to the condyles can be removed with a conventional or endoscopic endonasal approach. However, in a significant percentage of cases, dysplastic lesions were excised incompletely[5]. Furthermore, since the disorder is more extensive than it appears on clinical examination, navigation-guided surgical strategies are indicated to learn the individual's anatomy and the intraoperative orientation[18]. Some FD lesions need to be treated by extended transsphenoidal or subfrontal approach that have been used in the treatment of midline extra-axial cranial base tumors. Rarely, the patients presenting with severe headache and extension of the dysplasia to the condyle, craniovertebral fusion can be considered if there are signs of dynamic instability.

CONCLUSION

The authors report a rare case of monostic FD affecting the

clivus. This case emphasizes the importance of the differentiating this benign condition from other neoplastic clival lesions, based on clinical and radiological features. The conservative care with serial reevaluations is the key of the management strategy for FD in this area, however operative treatment is selectively indicated for patients with severe symptoms or disease progression.

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