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 transphenoidal 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 syndrome10. The monostotic FD usually involves the temporal bone, orbit, mandible, and para nasal 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 literature6,10.

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 magnetic 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 detail and implication for the role of surgery is also discussed.

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 diphosphonate 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 transphenoidal 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
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. 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. 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. 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. Furthermore, update report did not find presentation in adulthood to be rare, because 39-50% of patients in that series became symptomatic during adulthood. Continued growth into adulthood occurs and is more often observed in males than females. 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 extent of the abnormality. A localized painless expansion of the skull on one side is the commonest difference noted first by patients with craniofacial FD. This bone swelling can lead to
intracranial extension and central neurological manifesta-
tions, neurovascular entrapment, orbito-sino-nasal obstruction, 
recurrent sinusitis, epiphora, proptosis, malocclusion, and 
cosmetic deformity.2,7,29. On involvement of basal cranium 
by FD, compressive cranial neuropathy that mainly affect the 
facial, auditory, trigeminal, olfactory, and visual function 
occur from thickening and narrowing of neural canals and 
foramina.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.2 On rare instances, however, this is sympto-
matic, and presented with headache, neck pain, isolated 
sphenoid sinus tumor, craniofacial instability, or with hypo-
glossal nerve palsy.10. 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. De-
pending 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.10,21. 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 appear-
ance of FD is determined by the collagen content, overall 
cellularity, bony trabeculae, degree of mineralization, disease 
activity, and cyst formation. Almost all FDs show hypointen-
sity on T1-weighted images, while the signal intensity on 
T2-weighted images varies from high to intermediate or 
low.21,22. On the contrary, most common abnormalities occup-
ing the clivus such as chordoma, plasmyctoma, chordosar-
coma, giant cell tumor, hemangio, lymphoma, carcinoma, 
Paget’s disease, and sclerotic metastases, exhibit hypointen-
sity on T1- and hyperintensity on T2-weighted images.2,28. 
In addition, clival bone lesions showing low or intermediate 
signal intensity on T2-weighted MR imaging are rare. Con-
sequently, differential diagnostic possibilities above men-
tioned could easily be excluded by radiographic characteris-
tics observed in the present case, the "ground-glass" appear-
ance on CT scan and hypointensity in T1- and T2-weighted 
MR imaging. Unusually, foci of actively proliferating and 
growing portion within the lesion,22,28, as clearly depicted on 
our PET-CT scanning, might be responsible for iso- or hy-
perintensity on T2 MR sequence.

The management of craniofacial FDs is strictly surgical 
when they cause cosmetic deformation, intractable pain, dis-
ease progression, compressing cranial neuropathy, association 
with aneurysmal bone cyst, or malignant degeneration.2,10. 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 sur-
ery is recommended.2. 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 appro-
priately selected patients with cranial FD.2,20.

Because of its very slow potential growth, most FD of the 
divus, 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.2. As literature stated, the diagnosis is generally clear 
after a proper radiological and scintigraphic workup, most 
cases do not require the confirmatory biopsy.2,28. 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.1,2,21. 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.3. 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 incom-
pletely.3. Furthermore, since the disorder is more extensive 
than it appears on clinical examination, navigation-guided 
surgical strategies are indicated to learn the individual’s ana-
tomy and the intraoperative orientation.3. Some FD lesions 
need to be treated by extended transphenoidal 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 monostotic 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.

References