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Fibrous Dysplasia of the Clivus

Klivus Yerleşimli Fibröz Displazi

Mete İŞERİ, MD, Kadri İLA, MD, Deniz Özlem TOPDAĞ, MD, Murat TOPDAĞ, MD

Kocaeli University Faculty of Medicine, Department of Otolaryngology and Head and Neck Surgery, Kocaeli

ABSTRACT

Fibrous dysplasia is a localized benign disease of the mesenchyme, which occurs due to the replacement of normal medullary bone by structurally weak fibro-osseous tissue. Orbital, sphenoid, frontal, ethmoid, temporal and maxillary bones are the most commonly involved structures of the craniofacial region. Clinical symptoms like recurrent sinusitis, epiphora, proptosis, malocclusion and cosmetic deformation can be seen. The central neurologic signs and neurovascular compression symptoms can also be seen depending on abnormal extension and localization of the progressive bone deformation in the patients with craniofacial fibrous dysplasia. Fibrous dysplasia of the clivus is quite rare. Radiological imaging is usually adequate to make a diagnosis. The present paper introduced a case with fibrous dysplasia of the clivus presented with nasal obstruction and tremor.

Keywords

Fibrous dysplasia of bone; clivus; skull base neoplasm; endoscopy; nasal obstruction

ÖZET

Fibröz displazi, normal medüller kemiğin yapıca zayıf fibro-osseöz doku ile yer değiştirmesi sonucu oluşan lokalize, mezenkimal kökenli, benign bir hastalığıdır. Kraniofasiyal bölgede en sık orbita, sfenoid, frontal, etmoid, temporal ve maksiller kemik etkilenir. Kraniofasiyal fibröz displazili hastalarda progresif kemik deformasyonu sonucu anormal ekstansiyon ve lokalizasyona bağlı santral nörolojik belirti, nörovasküler bası, orbito-sino-nazal obstrüksiyon, rekürren sinüzit, epifora, propitozis, maloklüzyon ve kozmetik deformasyon gibi klinik semptomlar görülebilir. Klivus yerleşimli fibröz displazi olgusu çok nadirdir. Tanı için genellikle radyolojik görüntüleme yeterlidir. Bu makalede burun tıkanıklığı ve tremor şikayetiyle başvuran klivus yerleşimli fibröz displazi hastası sunuldu.

Anahtar Sözcükler

Kemiğin fibröz displazisi; klivus; kafa tabanı neoplazm; endoskopi; burun tıkanıklığı.

This study has been presented in 8th Turkish Rhinology Congress, 2012.

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Correspondence

Kadri İLA, MD

Kocaeli University Faculty of Medicine,
Department of Otolaryngology and Head and Neck Surgery, Kocaeli
dr.kila@gmail.com

INTRODUCTION

Fibrous dysplasia (FD) is a benign disorder in which medullary bone is replaced by fibroosseous tissue, and causes deformation and overgrowth of the involved bone.[1] It was 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.[2] FD classified into three forms: monostotic, involving only a single bone, which is the most common presentation (70% of patients); polyostotic, involving multiple bones involvement (30% of patient); and McCune-Albright syndrome, a rare variant of polystotic disease in which FD is associated with café-au-lait spots and multiple endocrine dysfunction such as hyperthyroidism and precocious menstruation in females.[2] The fibrous dysplasia of the clivus is a very rare disorder. We report a rare case of monostotic fibrous dysplasia of the clivus and discuss the management options.

CASE REPORT

A 51-year-old male patient, who has nasal obstruction for 10 years, presented to the neurology clinic with tremor in his hands. Magnetic resonance imaging (MRI) that was performed to eliminate cranial pathology revealed a 6x5cm mass beginning from the anterior aspect of the clivus, involving whole sphenoid bone including ala major and ala minor. It also involved the vomer and the lamina perpendicularis of the ethmoid bone expanding the nasal septum. The lesion showed heterogeneous signal intensity on T1 and T2 weighed images. In addition, a 3cm cystic area, probably indicating pooled secretion, was seen in the center (Figure 1). His maxillofacial computed tomography showed ground glass appearance (Figure 2). Cranial nerve examination of the patient, who had been radiologically diagnosed as fibrous dysplasia, was unremarkable. The patient was followed by the neurology clinic with the prediagnosis of Parkinson's disease. The patient was referred to our clinic because of fibrous dysplasia. Physical examination revealed a mass that deviated the nasal septum towards to the right and expanded from the dural plate to the nasal crest, lateralizing both inferior conchae (Figure 3). The results of routine biochemistry, complete blood count, T3, T4, TSH and parathormone analyses were within the normal levels. Neither Café au lait spots nor another skin lesion was detected. The patient was admitted to our clinic

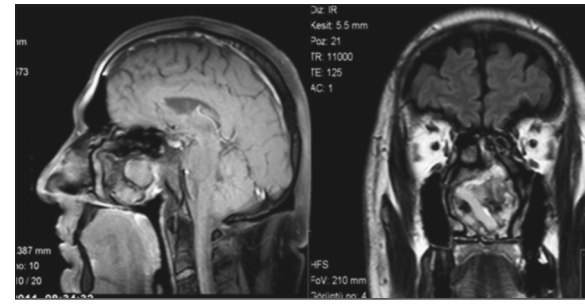

Figure 1. The 6 cm mass containing 3 cm cystic component probably indicating pooled secretion was revealed by cranial MRI. T1 and T2 weighed images show increased heterogeneous intensity including both the hypointense and hyperintense areas together.

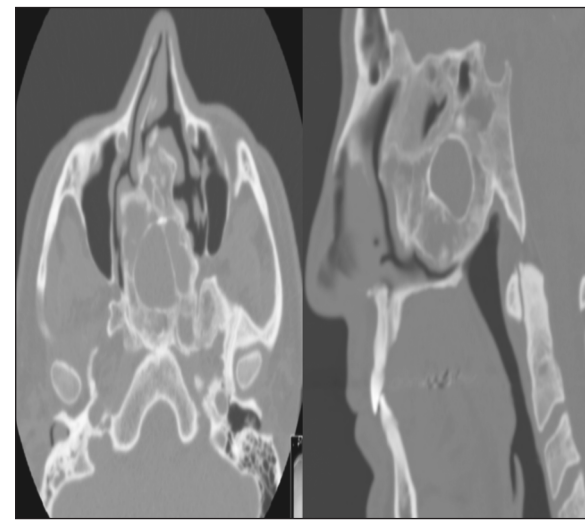

Figure 2. Approximately 6 cm mass with ground glass appearance in the nasal cavity consistent with fibrous dysplasia that enlarged the clivus was revealed by maxillofacial CT.

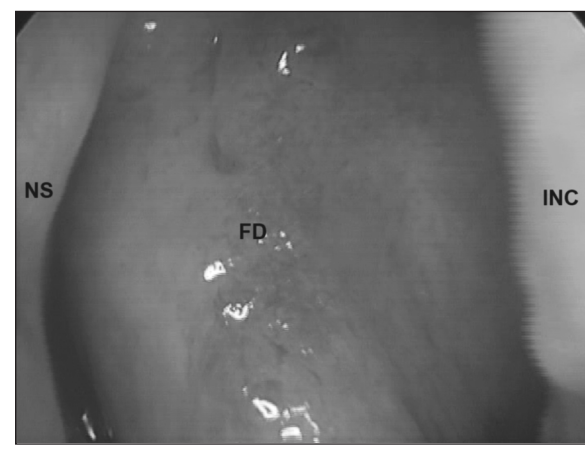

Figure 3. Endoscopic view of the mass from the left nasal cavity, which caused nasal obstruction and deviated the septum towards the right. NS (Septum), FD (fibrous dysplasia), and INC (inferior nasal concha) are seen.

for surgical intervention. Endoscopic endonasal approach was performed. After the mucoperichondrium elevated, it was observed that the mass was irregular and rich in bonny tissue. The part of the fibrous dysplasia in the nasal passage was removed with a diamond drill and a debrider. The patient underwent bilateral anterior and posterior ethmoidectomy. Both middle turbinates were rudimentary. Sphenoid sinus ostium was found and enlarged. The mass was drilled up to the level of the clivus (Figure 4). The nasal flap was overlaid on to the skull base creating a large nasal passage (Figure 5). Histopathological examination was reported as fibrous dysplasia. The lesion in the present case was localized in a single focus and considered as monostotic fibrous dysplasia.

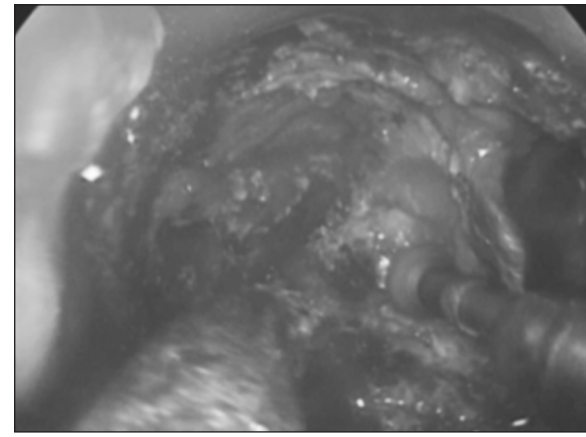

Figure 4. Removing of the fibrous dysplasia by drilling is seen.

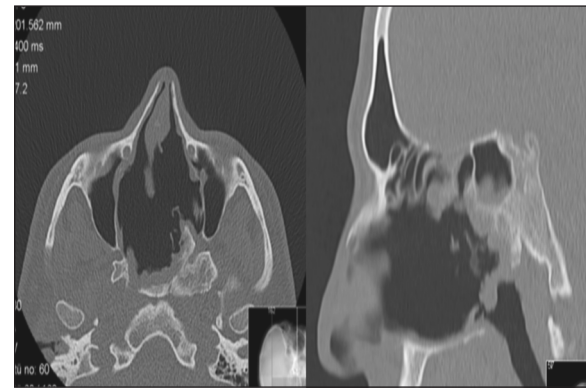

Figure 5. Maxillofacial CT examination showed postoperatif defect in the nasal cavity.

DISCUSSION

Fibrous dysplasia (FD) is the non-hereditary and non-neoplastic disease of the bone. Nevertheless, it may lead to enlargement, weakness and deformation of the bone because of local invasion and destruction. Craniofacial bones are involved in 30% of the monostotic fibrous dysplasia and in 50% of polyostotic fibrous dysplasia. Orbital, sphenoid, frontal, ethmoid, temporal and maxillary bones are the most commonly involved structures of the craniofacial region.[3] For patients with FD, clinical symptoms and signs result from progressive bone deformation that develops over several years, and depend on the location and extension of the abnormality. The great majority of the patients with craniofacial fibrous dysplasia present with unilateral, localized, painless cranial enlargement. This protuberance of the cranium may extend intracranialy causing central neurologic signs and neurovascular compression. Recurrent sinusitis, epiphora, proptosis, malocclusion and cosmetic deformations are other symptoms.[3,4] Patients with fibrous dysplasia which is located on the clivus may be asymptomatic or present with headache. If the fibrous dysplasia grows excessively on this region it may cause optic nerve and internal carotid artery compression, and the operation may be complicated. Because of this reason early treatment may be convenient.[5]

Fibrous dysplasia of the clivus is quite rare, to our knowledge there are only 26 cases have been reported in the literature.[1-3,5-11] The study conducted by Adada and Al-Mefty on eight cases with fibrous dysplasia of the clivus is the largest series reported until today.[5] Diagnosis had been made based on the pathological findings in four of these eight cases. The second largest study is three-case series with fibrous dysplasia of the clivus reported by Atalar and Ozum. It was reported that headache was the common complaint of these three patients.[8] The present patient case was diagnosed based on both radiological and pathological findings; nasal obstruction and history of tremor in hands were the presenting complaints. Literature review showed no case with fibrous dysplasia of the clivus extending far anteriorly causing nasal deformity and obstruction.

Radiological differential diagnosis of the clival lesions includes chordoma, chordosarcoma and plasmacytoma. Other diseases known to involve clivus are giant cell tumor, hemangioma, lymphoma, carcinoma, Paget's disease and metastasis.[5] Radiologic imaging is usually adequate to make a diagnosis. Ground glass appearance, thinning of cortical bone and protuberance of involved bone are the characteristic features of fibrous dysplasia on computed tomography.[6] Hypointensity is seen on T1 weighed magnetic resonance imaging, whereas hypointensity, isointensity and hyperintensity are seen on T2 weighed imaging. The degree of the cellularity, collagen content, bone trabeculation, calcification and cystic for-

mation are the reasons for the alterations seen on T2 imaging.[2,5,7] The cranial MRI of the present patient revealed 6 cm mass containing approximately 3 cm cystic area probably indicating pooled secretion, as well as increased heterogeneous intensity comprising both hypointense and hyperintense areas on the T1 and T2 weighted images.

Surgical approach for fibrous dysplasia should be considered in the presence of progressive deformity, cranial nerve compression, pain, and transformation into 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.[8] Various approaches are used for the treatment of midline skull-base tumors. Extra-axial cranial approach without cerebral and dural retraction is the best way. Ventral transoral-transpalatopharyngeal approach is effective for the treatment of fibrous dysplasia that causes compression in the cervicomedullary region.[6] Bicoronal approach can be applied when the orbita, anterior skull-base and sphenoethmoidal sinuses are involved. Midfacial degloving approach can be tried through a gingivobuccal incision for maxillary and midfacial involvements. Transconjunctival approach can be used during the surgical interventions of the inferior aspect of the orbita. Lateral rhinotomy and modified Weber Ferguson approaches are not preferred since they leave considerable scarring on the face.[12] Transsphenoethmoidal approach is used for 2/3 of the masses of superior clivus. The vascular soft tissues that are encountered inside the bone mass may complicate the surgery because of bleeding. Combined approach of sublabial transnasal surgery and partial resection of the nasal-base has been reported as well.[6] Endoscopic endonasal approach for skull-base tumors provides advantage with current technology. Endoscopic endonasal approach makes the treatment possible for small midline skull-base tumors, for which traditional transcranial approach was used.[6] Endoscopic endonasal transsphenoidal approach is superior to other approaches due to its wider point of view, avoiding blind curettage and short duration of surgical procedure. Its other advantage is providing adequate control of the clivus because of close view of the anatomical structures. Finally, endoscopic endonasal approach provides more aesthetical outcomes, is less traumatic, and provides post-operative comfort for the patients.[6]

Despite the good prognosis of fibrous dysplasia, malignant transformation has been reported as well. The prevalence of malignant transformation is 0.05% for monostotic craniofacial fibrous dysplasia.[7] Transformation of FD into osteosarcoma, fibrosarcoma and chondrosarcoma was reported in the literature.[7] Computed tomography is the most appropriate method for follow-up since it is able to detect any morphological or internal structural change.[13]

To sum up for differential diagnosis of clival lesion, CT and MRI should be performed together. Transsphenoidal endonasal endoscopic approach is a minimally invasive and useful technique for fibrous dysplasia of the clivus.

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