Monostotic Fronto-orbital Fibrous Dysplasia With Convulsion

—Case Report—

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Abstract

A 28-year-old man presented with monostotic fronto-orbital fibrous dysplasia associated with convulsions. Signs of meningeal irritation were observed. Computed tomography (CT) showed right frontal sinusitis, and destruction from the inner to outer table with expansion of the diploic space. T₁- and T₂-weighted magnetic resonance imaging showed an abnormal low-intensity mass, with heterogeneous gadolinium enhancement. Although the meningitis resolved, signs of infection continued for 2 months due to sinusitis. Treatment of the right frontal sinusitis was undertaken, accompanied by open biopsy. The histological diagnosis was fibrous dysplasia. Once the infection had completely resolved, orbitofrontal reconstruction was undertaken. Cranioplasty was carried out using cranial bone cement. Three-dimensional CT was valuable to show the likely postoperative result.

Key words: monostotic fibrous dysplasia, convulsion, three-dimensional computed tomography

Introduction

Fibrous dysplasia is a congenital, non-hereditary disease and little is known about the origin in bone disorders or the outcome.^{5,9)} The disease is characterized by chronic changes in the normal medullary bone tissue structure, resulting in complete growth failure and progressive fibrous tissue infiltration by combined collagen, fibroblasts, and osteoid tissue.^{8,15)}

Fibrous dysplasia frequently occurs in the facial bone and skull base. The disease is benign, but surgical treatment is indicated to prevent cranial nerve compression which may result in visual disturbances, severe cosmetic deformity requiring intervention, or malignant transformation.⁶⁾

Here, we describe a rare case of fronto-orbital monostotic-type fibrous dysplasia associated with convulsions, which was treated surgically with artificial bone cement.

Case Report

A 28-year-old man was admitted to our hospital with convulsive attacks on January 8, 1999. Asymmetrical protrusion of the right fronto-orbital region had been present since his youth, but had not progressed. A common cold had persisted from January 4, 1999. He had received medication, but convulsions had occurred twice.

On admission, radiological examination revealed a right fronto-orbital mass. Neurological examination revealed consciousness disturbance with signs of meningeal irritation. Measurement of hematological, biochemical, and cerebrospinal parameters revealed mild meningitis (cerebrospinal fluid examination: protein 59 mg/dl, sugar 76 mg/dl, cells 292/3 fields [leukocytes 52, lymphocytes 240]). Metabolic evaluation found no abnormalities, including the level of alkaline phosphatase. Skull radiography showed a ground-glass appearance and a well-circumscribed lesion in the right fronto-orbital bone (Fig. 1 left). Computed tomography (CT) of the skull bones showed pneumocephalus, right ethmoidal sinusitis, and destruction from the inner to outer table with expansion of the diploic space (Figs. 1 right and 2 left). Three-dimensional (3D) CT clearly

Fig. 1 *left*: Skull radiograph showing a ground-glass appearance and a well-circumscribed lesion in the right fronto-orbital bone (arrow). *right*: Bone image computed tomography scan showing pneumocephalus (arrowhead) with destruction from the inner to outer table with expansion of the diploic space.

Fig. 2 *left*: Coronal bone image computed tomography (CT) scan showing right sinusitis at a fronto-ethmoidal lesion. *right*: Three-dimensional CT scan emphasizing the frontal asymmetrical protrusion (arrow).

demonstrated the fronto-orbital asymmetrical protrusion (Fig. 2 *right*). Magnetic resonance (MR) imaging revealed a low-intensity mass on T₁- and T₂-weighted images, which was heterogeneously enhanced by contrast medium in the diploic space (Fig. 3A, C). T₂-weighted MR imaging showed a high-intensity area adjacent to the lesion, and local encephalitis was suspected (Fig. 3B). Technetium-99m-labeled methylene diphosphonate bone scintigraphy showed an abnormal accumulation corresponding to the right fronto-orbital region (Fig. 3D). Electroencephalography (EEG) showed small spikes in the right frontal area. No other abnormalities were observed in the rest of the body. The preoperative diagnosis was fibrous dysplasia. Following admission and treatment, the meningitis, encephalitis,

Fig. 3 *A*: Axial T₁-weighted magnetic resonance (MR) image showing a low-intensity mass in the right fronto-orbital region. *B*: Axial T₂-weighted MR image showing a high-intensity mass and suspected encephalitis (arrowhead). *C*: Axial MR image with gadolinium showing high enhancement. *D*: Technetium-99m-labeled methylene diphosphonate bone scan showing an area of accumulated abnormalities corresponding to the fronto-orbital region (arrow).

and pneumocephalus resolved by the middle of March. EEG showed no further spikes. However, infectious signs continued due to sinusitis.

Surgical treatment of the right frontal sinus was undertaken using the Killian method, accompanied by open biopsy (Fig. 4 *upper*). The tumor was found in the perifocal area and was elastic-hard without surrounding infection. The tumor included trabeculae of bone with irregular margins within fibrous connective tissue (Fig. 4 *lower*). No normal bone elements were present in the field. The histological diagnosis was monostotic-type fronto-orbital fibrous dysplasia.

The sinusitis and general condition had completely resolved by the middle of April. Orbito-frontal reconstruction with dural plasty was then undertaken to confirm whether a connection between the intradural and extradural spaces was present. Intraoperative findings showed that the borderline between the lesion and the normal lateral orbital wall

was unclear. The dura mater adjacent to the lesion was very thick, but no connection was found. Cranioplasty was performed using CEMEX isoplastic (TECRES spa, Verona, Italy). Postoperative skull radiography showed adequate reconstruction (Fig. 5A, B), and 3D CT showed acceptable recontouring of the fronto-orbital bone (Fig. 5C). The postoperative course was uneventful, without further convulsive attacks.

Discussion

Fibrous dysplasia appears as an expansile osseous lesion, associated with thinned but intact cortical bone. There is no periosteal reaction in the absence of fractures.⁴⁾ The differential radiographic diagnoses are ossifying fibroma, histiocytosis, Paget's disease, meningioma of the en plaque type, aneurysmal bone cyst, giant cell tumor, giant cell reparative granuloma, and the "brown tumor" of hyperparathyroidism.^{4,7,12,13)} Ossifying fibroma is the most difficult to distinguish from fibrous dysplasia. Compared to fibrous dysplasia, ossifying fibroma is a better demarcated expansile lesion with smooth margins, and tends to behave more aggressively, encroaching on the nasal fossae, orbits, and paranasal sinuses.⁶⁾ In our case, the diagnosis was monostotic-type fibrous dysplasia based on the presence of a solitary mass which was not subcutaneously invasive and the intact endocrinological characteristics.

The convulsive attacks were probably due to the pneumocephalus and meningitis. The lesion due to the pneumocephalus had caused the dura mater to thicken. However, there was no infection because the sinusitis and its symptoms had already been

Fig. 4 upper: Intraoperative photograph (Killian method) showing that the tumor is elastic and hard, with no surrounding infection. lower: Photomicrograph showing trabecular bone with irregular margins within fibrous connective tissue. HE stain, $\times 20$.

Fig. 5 A, B: Anteroposterior (A) and lateral (B) postoperative skull radiographs showing adequate reconstruction. C: Three-dimensional computed tomography scan showing recontouring of the fronto-orbital bone.

completely treated. Fibrous dysplasia rarely invades the inner table. We suspected that the worsening sinusitis had caused the pneumocephalus.

Surgical treatment of fibrous dysplasia^{11,14} indicated in the case of progressive cranial nerve compression,² severe functional disorders, or malignant transformation. Treatment should be conservative in the absence of neurological deficits. There is a 25% incidence of recurrence if partial resection is performed.¹ Radiation can induce malignant transformation and therefore is not recommended.¹⁰ In our case, after the inflammation had been completely treated, we decided to undertake cranioplasty to repair the dura and treat the pneumocephalus.

3D CT and MR imaging allow radical ablation in the presence of a suitable anatomic structure. In particular, 3D CT images are valuable in selected cases of craniofacial fibrous dysplasia in which bone contouring is planned to achieve facial symmetry.³

CEMEX isoplastic cement has a high viscosity and is easy to handle during reconstruction. The permeability is almost the same as skull bone, and so allows effective cosmetic evaluation of the laterality of the orbito-frontal reconstruction. This artificial bone cement is highly effective for use in cranioplasty, including correction of the surrounding orbital region.

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