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Clinical and radiological observation in a surgical series of 36 cases of fibrous dysplasia of the skull

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ABSTRACT

Objectives: To describe the clinical and radiological findings in a consecutive series of patients diagnosed with fibrous dysplasia of the skull.
Patients and methods: A retrospective analysis of collected data for 36 patients with histopathologically confirmed fibrous dysplasia involving the skull is presented. The demographic data, clinical presentation, radiographic characteristics, and the management of these patients were reviewed.
Results: All 36 patients in this review were diagnosed with fibrous dysplasia involving at least part of the skull. In this study, the most commonly involved area of the skull was the frontal bone (52.78% of patients). The next most common area of skull was the temporal bone (30.56% of patients), followed by the sphenoid bone (25% of patients), the parietal bone (19.44% of patients), and orbital bone (13.89% of patients). The principal clinical presentation included headache, local lump, exophthalmos, visual disorder, cranial nerve paralysis, and facial malformation. These patients were treated by surgical treatment, and several of our patients underwent various degrees of reconstruction to optimize function.
Conclusions: Effective surgical treatment may improve the short-term outcome in these patients, and a "tailored" surgical approach is necessary.

1. Introduction

In 1891, Von Recklinghausen [1] described a number of individuals with striking bone lesions. Most are now known to have had the condition called neurofibromatosis, but some had fibrous dysplasia. McCune and Bruch [2] and Albright et al. [3] recognized in separate publications in 1937 the entity of osteodystrophia fibrosa disseminata, characterized by endocrinopathies, cutaneous hyperpigmentation, and precocious puberty in females. This severe form of fibrous dysplasia subsequently became known as the Albright triad or McCune–Albright syndrome [4]. The terms "fibrous dysplasia" and "polyostotic fibrous dysplasia" were first suggested by Lichtenstein [5].

Fibrous dysplasia is a relatively uncommon benign disorder of unknown cause, in which the normal bone structure is replaced by fibrous connective tissue with woven bony trabeculae. The disease can involve any bone in the body. Fibrous dysplasia can be monostotic or, less frequently, polyostotic. Involvement of the craniofacial bone results in noticeable facial deformity.

Recently, magnetic resonance (MR) imaging as well as radiography and computer tomography (CT) have been used for the diagnosis of fibrous dysplasia, which can help to reduce the possibility of misdiagnosing fibrous dysplasia of the skull for neoplastic diseases. We retrospectively studied 36 cases with histopathologically confirmed fibrous dysplasia (F-24, M-12; ages ranged from 6 to 59 years) involving the skull, who were cared for over a 21-year period (1988–2009).

2. Patients and methods

2.1. Patient population and general data

Between January 1988 and June 2009, 36 patients were admitted to the Department of Neurosurgery of Wuhan General Hospital and Wuhan Tongji Hospital with fibrous dysplasia involving at least part of the skull and subsequently treated. Patient information, including age at presentation, principal clinical symptoms, signs of the skull lesion, treatment, and clinical outcome, were collected. Radiographic evaluations including X-ray, computed tomography

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(CT) and magnetic resonance imaging (MRI) were obtained preoperatively and postoperatively when possible. In all patients, histopathological examination confirmed the diagnosis.

2.2. Statistical analysis

The results are expressed as the means ± standard deviation (SD). Statistical analyses were performed using SPSS Statistical Software (SPSS Inc., Chicago, IL).

3. Results

3.1. Clinical presentation

The main manifestations were atypical facial pain and local lumps. Some patients suffered from headache and dizziness: 5 presented with decreased visual acuity, 4 with exophthalmos, 7 with facial malformation, 2 with cranial nerve paralysis, and 1 with hearing loss; none had high intracranial pressure (see Table 1).

3.2. Radiological findings

All patients submitted to skull X-ray with serial films to assess disease progress. In total, 21 patients underwent computed tomography (CT), 4 patients had three-dimensional computed tomography (3D-CT), and 10 patients underwent magnetic resonance imaging (MRI). For patients in our series, plain film X-rays were usually sufficient to diagnose most cases of fibrous dysplasia. Radiographic findings demonstrated the classic ground-glass appearance on standard X-ray films. CT findings of the 21 patients in this study also showed characteristics of fibrous dysplasia and consisted of the following three varieties as described before [6]. The pagetoid, or ground-glass, pattern is most common (56%) and consists of a mixture of dense and radiolucent areas of fibrosis (Fig. 1 A, Patient 14). Sclerotic lesions (23%) display a massif that is homogeneously dense (Fig. 1B, Patient 23), whereas the cystic variety (21%) is characterized by a spherical or ovoid lucency surrounded by a dense, bony shell boundary (Fig. 1G, Patient 31). CT contrast-enhanced scans showed enhancement of the lesion (Fig. 1E, Patient 2). Three-dimensional computed tomography (3D-CT) demonstrated enlargement of the left anterior clinoid process and planum sphenoidale with gross destruction (Fig. 1C, Patient 23). MRIs demonstrated that the signal intensities of fibrous dysplasia were usually low on T1-weighted images (Fig. 2A, Patient 14). The signal intensity of fibrous dysplasia on T2-weighted images was often variable, ranging from low to high signals in some patients (Fig. 2B (Patient 14) and 2C (Patient 2)). Some lesions with a highly mineralized matrix showed correspondingly low signal intensities, whereas lesions with high fibrous tissue content and cystic spaces return high signal intensities. These high signal intensities on T2-weighted images corresponded to non-mineralized areas and to regions of cyst degeneration seen on CT (Figs. 1E and 2C, Patient 2).

3.3. Clinical results

The average age at presentation was 25 years (ranging from 6 to 59 years). Twenty-three patients were younger than 18 years, and two were more than 50 years old. There was a 1:2 male–female ratio in terms of incidence (12 males and 24 females). The course of disease varied from 10 days to 18 years, average (3.08 ± 3.66) years.

In 36 patients (0.5–11 years follow-up), the mean follow-up time was 4.7 years. Twenty-four patients have had at least 18 months of follow-up, and 15 have had 5 full years of follow-up. Mid-term and long-term follow-up data showed improvement of vision and relief of headaches. To our knowledge, no patients have experienced clinical deterioration. Improvement of vision was found in 3 eyes, stability of vision was found in 1 eye, and decreased vision was found in 1 patient. We obtained follow-up CT scans in three patients. None of them showed any changes or progression of the lesion upon radiological examination. Four patients (11%) had fibrous dysplasia recurrence and underwent a second operation for removal of a non-neoplastic tissue mass in the skull. Table 1 lists the clinical information, including gender, age, localization of the tumor, treatment procedure, clinical outcome, and time of follow-up for each patient.

3.4. Case reports illustrating tailored surgery

Non-surgical treatment of fibrous dysplasia yields results that are very disappointing. Surgical therapy has been quite helpful in the management of fibrous dysplasia. Obvious indications for surgery involve the correction of deformity, the treatment of pathological fractures, the relief of pain, and the arrest of excessive bone growth. Surgery may reduce the risks to vision, hearing, or speech caused by the encroachment of these tumors. Therefore we use the "cut, remove, remodel, and replace" method of dealing with fibrous dysplasia (see Table 1).

Patient 14 (Fig. 1A) was a 44-year-old woman who was admitted to our hospital with a 3-year history of atypical headache. Axial computed tomographic scan showed fibrous dysplasia of the ethmoid and sphenoid bones. Fibrous dysplasia extended medially into the ethmoids and sphenoid, which nearly completely obliterated the sinuses. Accordingly, through an endoscopic endonasal transsphenoidal approach, this patient underwent resection of the cystic component along with endoscopic drainage. At 6 months' follow-up, the patient was doing very well without evidence of tumor regrowth or new neurological complaints; sinusitis has not recurred.

Patient 31 (Fig. 1G and H) was a 16-year-old girl who presented to us with a 27-month history of progressive headaches and cranioaural asymmetry due to a left-sided protuberant parietal region and occipital bossing. CT scan revealed an extensive lytic process extending from the left parietal bone to the left occipital bone. A generous left parietal craniotomy was performed to completely remove the dysplastic bone. Reconstruction after radical surgery was achieved with titanium allegation implants in this patient. The dura remained intact throughout. Four years postoperatively, the patient's CT scan showed no abnormalities with persistence of the implants; the patient has an improved appearance and resolved headaches. In our patients in whom the involvement of frontal bones, parietal bones, and occipital bones is limited underwent complete removal of the dysplastic bone. The resulting defect was immediately reconstructed using large titanium allegation implants (Table 1). These implants have been well tolerated with good clinical results and lower morbidity than other methods.

At the age of 45 years, patient 23 was hospitalized with a slowly growing bony swelling over the left temporal area, which had been present for 6 months. For 3 months, the patient had noticed progressive, painless, axial proptosis of his left eye. There had been a decrease in his visual acuity (0.5 in the affected eye). His visual fields were normal. A left-sided frontotemporal craniotomy was performed to decompress the left optic nerve. The patient underwent partial resection of the fibrous dysplasia of the greater wing of the sphenoid bone, and the left optic canal was decompressed via an extradural pterional approach using a high-speed drill. The fibrous dysplastic lesion was firm and hemorrhagic. His visual acuity and visual field remained unchanged 1 year after surgery. At 1.5 years postoperatively, the patient had recurrent craniofacial fibrous dysplasia without any visual changes. He has undergone a second tumor resection. While vision in the affected eye