J Korean Neurosurg Soc 37 : 383-385, 2005

Case Report

Malignant Transformation of Fibrous Dysplasia on Anterior Skull Base

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Malignant transformation of fibrous dysplasia is a rare condition and the incidence is estimated at 0.4% for fibrous dysplasia and 4% for Albright's syndrome. The authors did not find a reported case of malignant change at skull base around the orbit in the literature. We experienced a case of fibrous dysplasia, in which neurologic symptoms were aggravated due to malignant change around the orbit, and report its favorable outcome obtained with total surgical removal.

KEY WORDS : Fibrous dysplasia · Skull base · Malignant transformation.

Introduction

Since fibrous dysplasia was first named by Leichtenstein[11,15], many studies have been conducted but definite treatment has not been established yet. Malignant transformation of fibrous dysplasia is a rare but an established complication of this disease. The first well-documented account was described by Coley and Stewart in 1945[7].

Fibrous dysplasia is a fibrous tumor, in which normal bone tissue is substituted by abnormal growth of fibrous connective tissue and causes deformation, enlargement, and aggravation of structural bone tissue. They are classified as monostotic and polyostotic type. The polyostotic form is less frequent than monostotic fibrous dysplasia. The most common locations of polyostotic type are the lower extremities and pelvis[2,13]. Fibrous dysplasia of the skull represents 10 to 27% of monostotic cases and occurs in all severe polyostotic forms. The most common presenting symptoms of craniofacial dysplasia are focal cranial swelling and exophthalmos or visual disturbance. Impaired vision is present in 20 to 35% of cases[1,4,5,13,14].

In fibrous dysplasia, malignant transformation is rarely occurred. This should be considered when recurrent or stable fibrous dysplasia produces pain or soft tissue extension or neurologic deficits[3,9,17]. In this case we should think the possiblity of the malignant change. And the presence of which can be established by radiologic examination and biopsy, particularly in an older patients. The most common malignant transformation reveals osteosarcoma, followed by fibrosarcoma and chondrosarcoma, giant cell sarcoma[9,15-17]. Prognosis for malignant change in fibrous dysplasia is poor. Most patients died with pulmonary metastasis, and the mean survival period was 3.4 years[17].

We experienced a case of fibrous dysplasia, in which neurologic symptom aggravated due to malignant change at skull base around the orbit, and total surgical removal is the only factor that has definite effect on the prognosis of the patient.

• Received : August 11, 2004 • Accepted : September 3, 2004
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Case Report

History

54-year-old male patient complained of headache, exophthalmos and decreased visual acuity was admitted. He had diagnosed polyostotic fibrous dysplasia and multiple bone involvement twenty years ago and had no specific treatments. Brain MRI showed heterogenous mass compressing optic nerve at apex of the orbit, with the bone change by fibrous dysplasia(Fig. 1).

At the first operation, it was thought that all the tumor had been removed. But it recurred after 6 months postoperatively, then the second operation was done. After the second operation, remnant tumor was observed(Fig. 2A). Only chemotherapy (Cisplatin + Adriamycin) was done because the patient refused to take reoperation. After 6cycles of chemotherapy, decreased visual acuity and ocular pain were aggravated. Follow-up brain MRI showed enlargement of the mass compressing the optic nerve(Fig. 2B). Third operation was performed.

The histologic evaluation of the surgically removed tumor revealed typical osteoid matrix producing bone tumor showing proliferation of osteoblasts and focal cartilage formation(Fig. 3).

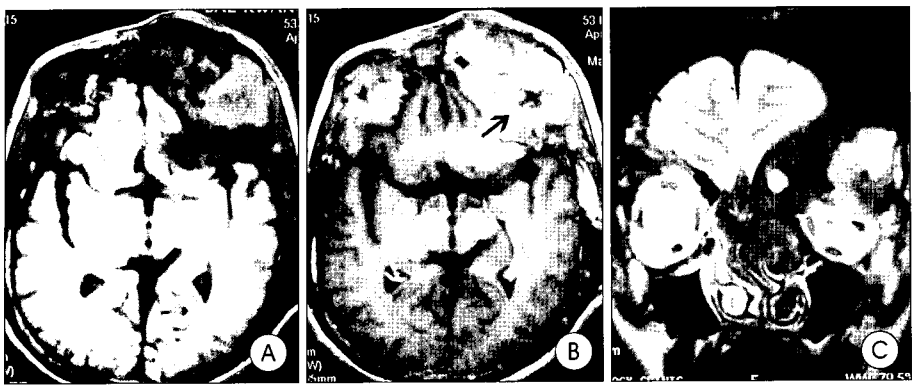

Fig. 1. T1-weighted axial magnetic resonance(MR) image of brain(A), T1-weighted axial enhanced MR image(B) and T1-weighted coronal MR image(C) show the well demarcated, slightly enhanced heterogenous mass lesion inside the fibrous dysplastic bone which was present before. The mass is compressing the left eye ball from its lateral side(arrow).

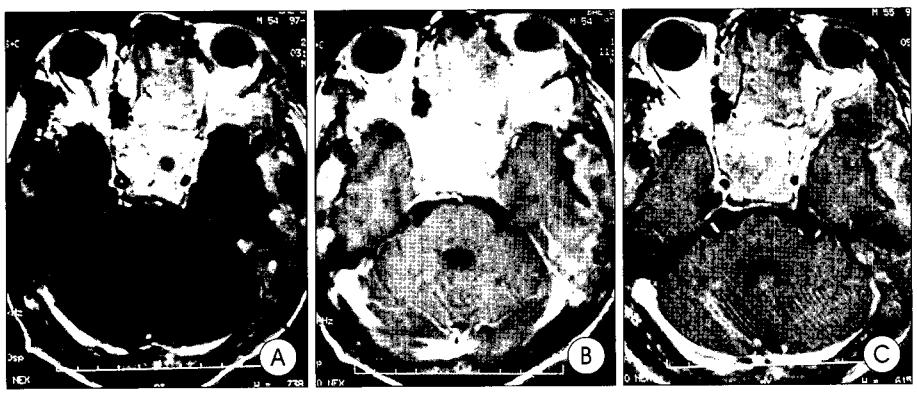

Fig. 2. Second postoperative T1-weighted axial enhanced magnetic resonance image shows that tumor is partially removed and the remnant tumor is visible. It still compresses optic nerve slightly(A). This means that the tumor is still growing after the chemotherapy(B). And after third operation, tumor is totally removed and there are no signs of other enhancing mass lesion(C).

Tumor had been completely removed with the third operation, and there was no further worsening of visual acuity. No evidence of tumor recurrence has been observed, and the patient is under follow up(Fig. 2C).

Discussion

Since fibrous dysplasia was first named by Leichtenstein[11,15], many studies have been conducted but definite treatment has not been established yet. As a rare tumor making up 2.5% of all bone tumors, and 7.5% of benign bone tumor, it is classified as monostotic and polyostotic type according to multiplicity of the lesion[6,12,14]. Polyostotic type, compared to monostotic type, tend to have deformation of bone tissue at multiple lesions, and nerve tissue compression, neurologic deficit, and cosmetic problems caused by it.

In the case of orbit and frontal bone involvement, surgical treatment is required when neurologic symptom, secondary deformation of other structures, complications due to fibrous dysplasia and cosmetic necessity are present. Treatment for fibrous dysplasia is sought for cosmetic and functional reasons. In the past, conservative management was usually recommended. However, because of the uncertain outcome, radical excision of dysplastic bone with immediate reconstruction of the region has recently been suggested[5,7,11].

Malignant degeneration of fibrous dysplasia is a rare condition. The incidence is estimated at 4% for Albright's syndrome (skin pigmentation, endocrine disorders, precocious puberty)[2,3,8-10,18]. Ebata et. al[9] reported 89 cases of malignant degeneration occurring in fibrous dysplasia. The most common type is osteogenic sarcoma, occurring in 60.7% of the reported cases. Common sites were craniofacial bones, particularly the mandible and maxilla, femur, tibia and pelvis. In most reported cases, the diagnosis of fibrous dysplasia was made in childhood but the malignant tumor developed during the third or fourth decades of life[17]. The lag between the development of the fibrous dysplasia and

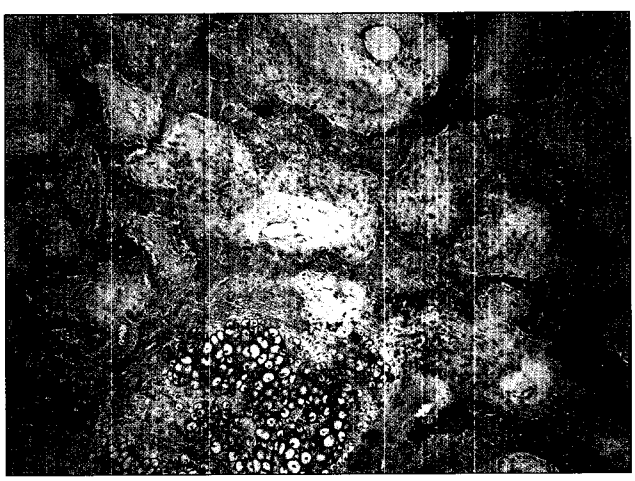

Fig. 3. On microphotographs of tumor tissue, histologically tumor consists of typical osteoid matrix producing bone tumor showing proliferation of osteoblasts and focal cartilage formation(H&E, ×100).

the sarcoma varied from a minimum of 2 years to a maximum of 30 years, the mean being 13.5 years. There was an equal number of males and females[15]. Malignant degeneration at skull base around the orbit is thought to be extremely rare. We could not find such a case in published articles. Twenty-three of the 83 cases were treated with local radiation[9,14,17]. It seems that irradiation provokes the fibrous dysplasia to undergo sarcomatous change[15]. Therefore, radiotherapy should not be used for the treatment of this case. Neither radiotherapy nor chemotherapy favorably influenced the clinical course of the tumor[15]. And our case does not reveal favorable response to chemotherapy. Total surgical removal is the only factor that has definite effect on the prognosis of the patient.

Conclusion

We experienced a case of fibrous dysplasia, in which neurologic symptom aggravated due to malignant change at skull base around the orbit, and report its favorable outcome obtained with total surgical removal.

In this cases, neither radiotherapy nor chemotherapy favorably influenced the clinical course. Total surgical removal is the only factor that has definite effect on the prognosis of the patient.

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