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Primary Osteosarcoma of the Sphenoid Bone with Extensive Periosteal Extension

—Case Report—

Takuro HAYASHI, Yoshiaki KUROSHIMA, Kazunari YOSHIDA, Takeshi KAWASE, Eiji IKEDA*, and Makio MUKAI*

*Departments of Neurosurgery and *Pathology, Keio University School of Medicine, Tokyo*

Abstract

A 28-year-old male presented with a primary osteosarcoma of the sphenoid bone with extensive periosteal extension manifesting as severe headache and right exophthalmos. Computed tomography (CT) and magnetic resonance imaging revealed an anterior middle cranial fossa tumor extending into both the orbit and the extracranial space. However, roentgenography and CT with bone windows showed no marked osteolytic changes of the sphenoid bone. Total removal of the tumor was performed via the orbitozygomatic approach. Surgery revealed that the tumor had extended periosteally without macroscopic bone destruction, but no obvious abnormalities of the skull. The histological diagnosis was osteosarcoma. The patient was treated with chemotherapy and radiation therapy, but died of tumor recurrence 10 months after the surgery.

Key words: osteosarcoma, sphenoid bone, osteoblastic change, osteolytic change

Introduction

Osteosarcoma is a highly malignant tumor which usually occurs in the long bones, and rarely affects the craniofacial bones. Osteosarcoma in the craniofacial bone accounts for 6–13% of all osteosarcomas,[12] and for less than 0.5% of all head and neck tumors.[1] Craniofacial osteosarcoma arises more frequently in the mandible and maxilla than in the calvaria and skull base.[12] Osteosarcoma arising from the sphenoid bone is extremely rare. We describe a case of primary osteosarcoma of the sphenoid bone without marked osteolytic or osteoblastic changes.

Case Report

A 28-year-old male was hospitalized with severe headache, right temporomandibular joint pain, and right exophthalmos. On admission, his vital signs were within normal limits and no neurological abnormality was recognized. Laboratory data were also within normal limits. Skull roentgenography showed no marked erosion of the sphenoid bone or the mandibular joint. Computed tomography (CT) and magnetic resonance (MR) imaging disclosed a ring-enhanced mass in the right temporal lobe measuring 4 cm in diameter with perifocal brain edema, an extracranial mass deep to the temporalis muscle, and an enhanced mass in the orbit. CT with bone windows did not show obvious marked bone destruction but indicated a slight hypertrophic change of the sphenoid bone (Fig. 1). Right carotid angiography showed that the lesion was hypovascular.

The patient underwent a right orbitozygomatic craniotomy with total removal of the tumors. Surgery revealed a tumor under the right temporalis muscle. Craniotomy exposed an epidural tumor, which had penetrated the dura mater and invaded into the temporal lobe, and a right intraorbital tumor. The normal shape of the sphenoid bone was preserved and no obvious macroscopic cortical abnormalities were observed.

Histological examination of the resected tumor and the sphenoid bone confirmed the diagnosis of osteosarcoma (Fig. 2). The tumor cells had proliferated mainly along the bone marrow spaces with minimal destruction of the cranial bone, and the tumor had grown across the dura (Fig. 3). Thoracic

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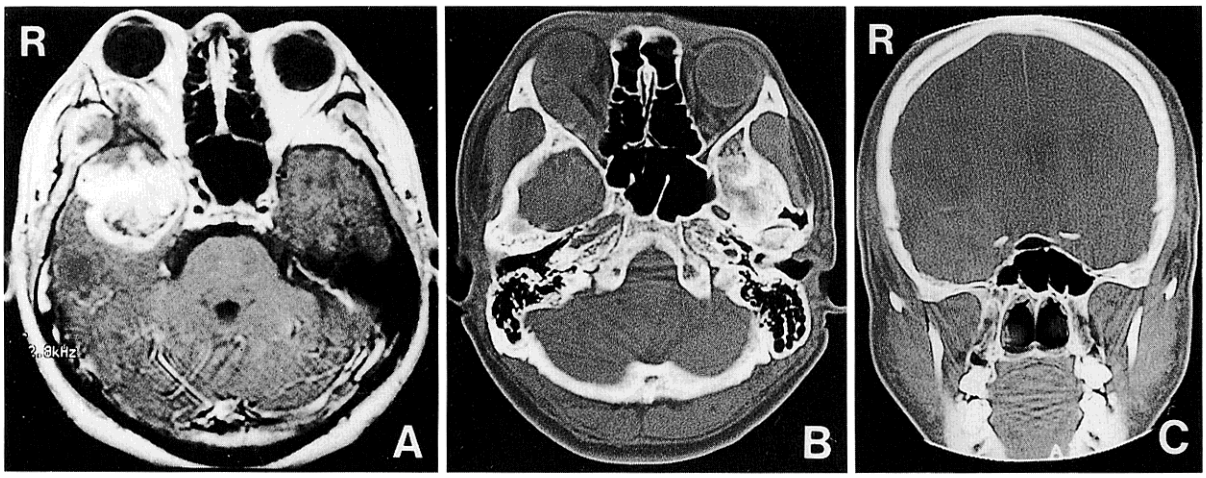

Fig. 1 A: Magnetic resonance image disclosing a ring-enhanced mass in the right temporal lobe measuring 4 cm in diameter with perifocal brain edema, an extracranial mass deep to the temporalis muscle, and an enhanced mass in the orbit. B, C: Axial (B) and coronal (C) computed tomography scans with bone windows showing no marked osteoblastic or osteolytic changes in the right sphenoid bone.

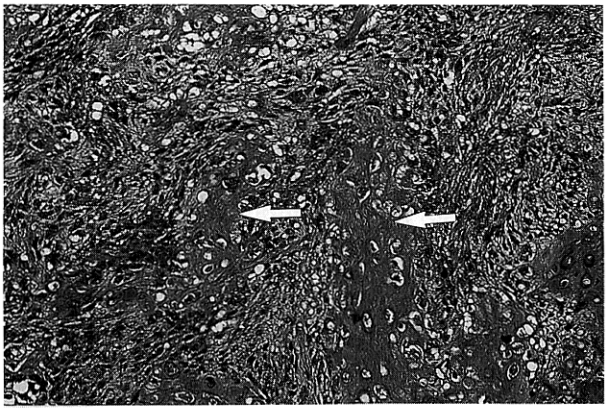

Fig. 2 Photomicrograph of the tumor showing diffuse proliferation of spindle-shaped or polygonal cells with nuclear atypia. The areas of osteoid formation (*arrows*) are characteristic of osteosarcoma. HE stain, original magnification ×66.

CT and gallium and bone scintigraphy detected no other primary or metastatic lesions.

Postoperatively, his symptoms were alleviated, and he underwent one course of methotrexate/cisplatin/tetrahydropyranyl-adriamycin intravenous chemotherapy and received radiotherapy (whole brain, total 50 Gy). MR imaging 2 months after the operation suggested recurrence of the tumor in the right orbit, and so further chemotherapy was discontinued. The patient was followed up at our outpatient department and serial MR imaging revealed enlargement of the recurrent tumor. The patient died 10 months after the operation.

Discussion

Osteosarcoma is extremely rare in the sphenoid bones, with only seven cases reported.[2,4,6,7,11,13] The mean age at onset of craniofacial osteosarcoma is the third decade of life, 10 years older than that for osteosarcoma in the long bones.[3,8,9,12] The cause of craniofacial osteosarcoma is unknown, but Paget's disease of the skull, previous radiotherapy, trauma, and fibrous dysplasia are associated with secondary osteosarcoma.[3,5,7,9] The most common primary osteosarcoma is the de novo type.[5] We believe that our case was the de novo type because the patient had no past history of osteosarcoma-associated diseases, and no other primary lesions were detected.

Roentgenography and CT with bone windows are the most useful method for the diagnosis of osteosarcoma. Sclerosis and lysis of the bones are the most common imaging features.[7,8,12] Radiological investigations frequently reveal destruction of the cortex of the skull bone. CT with contrast medium and MR imaging can define the extension into the soft tissue, for example, into the brain and muscle.[7] In our case, roentgenography and bone window CT demonstrated no typical findings except for slight hypertrophic change of the sphenoid bone, despite the detection of multiple enhanced masses, so no

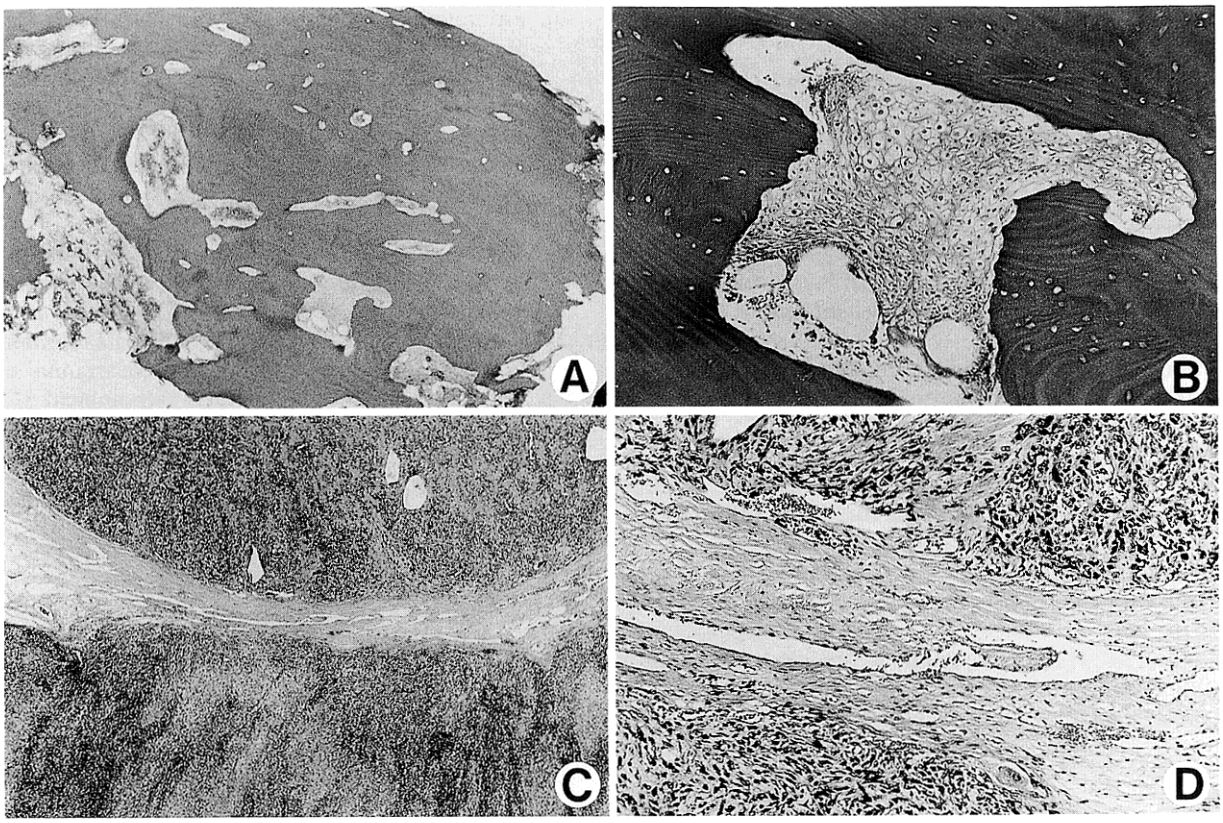

Fig. 3 A, B: Photomicrographs showing the tumor tissue in the right sphenoid bone, with proliferation of tumor cells mainly along the bone marrow spaces and minimal destruction of the preexisting trabecula of the lamellar bone. C, D: Photomicrographs showing the tumor penetrating the dura. HE stain, original magnification A: ×20, B: ×100, C: ×40, D: ×100.

firm preoperative diagnosis could be made. The operative findings also did not reveal obvious abnormalities of the sphenoid bone, such as deformities, erosions, or intraosseous masses. Histological examination of the surgical specimens also showed only the typical findings of osteosarcoma.

The most important differential diagnostic problem is centered on the origin of the tumor. In the present case, the tumor had invaded through both the sphenoid bone and the dura mater without visible deformity of the sphenoid bone, so we could not tell macroscopically whether the tumor originated from the bone or the dura mater. Therefore, the differential diagnosis should include the so-called "periosteal variant," which is a common subtype of osteosarcoma. The histological diagnosis was osteosarcoma. The tumor cells had proliferated mainly along the bone marrow spaces with minimal destruction of the preexisting trabecula of the lamellar bone. Therefore, we concluded that the tumor had originated from the sphenoid bone and extended periosteally with only slight intraosteal development which did not cause obvious cortical deformities. Consequently, the tumor was difficult to differentiate from multiple soft tissue tumors at the preoperative examination. This case presented with a unique developmental pattern for osteosarcoma.

The generally accepted treatments for craniofacial osteosarcoma are surgical resection, polychemotherapy, and radiotherapy.[5,7,10,12] Combined therapy has frequently been reported to improve the outcome. However, the prognosis for patients is not always satisfactory. Our patient was primarily treated with surgical resection for decompression and to establish the diagnosis. Chemotherapy and radiotherapy were administered as adjuvant treatments. However, the outcome for our patient was unfavorable, so further developments in treatment are required.

References

1) de Fries HO, Perlin E, Leibel SA: Treatment of os-

teogenic sarcoma of the mandible. *Arch Otolaryngol* 105: 358-359, 1979
2) Hazarika P, Nayak DR, Sahota JS, Rao D, Kapadia RD: Osteogenic sarcoma of sphenoid bone: an extended lateral skull base approach. *J Laryngol Otol* 109: 1101-1104, 1995
3) Huvos AG, Sunderson N, Bretsky SS, Butler A: Osteogenic sarcoma of the skull. A clinicopathologic study of 19 patients. *Cancer* 56: 1214-1221, 1985
4) Marks MP, Marks SC, Segall HD, Schwinn CP, Forrester DM: Case report 420. *Skeletal Radiol* 16: 246-251, 1987
5) Nora FE, Unni KK, Pritchard DJ, Dahlin DC: Osteosarcoma of the extragnathic craniofacial bones. *Mayo Clin Proc* 58: 268-272, 1983
6) Salvati M, Cervoni L, Ciappetta P, Raco A: Radiation-induced osteosarcomas of the skull: report of two cases and review of the literature. *Clin Neurol Neurosurg* 96: 226-229, 1994
7) Salvati M, Ciappetta P, Raco A: Osteosarcoma of the skull: Clinical remarks on 19 cases. *Cancer* 71: 2210-2216, 1993
8) Sharma SC, Handa KK, Panda N, Banerjee AK, Mann SBS: Osteogenic sarcoma of the temporal bone. *Am J Otolaryngol* 18: 220-223, 1997
9) Shinoda J, Kimura T, Funakoshi T, Iwata H, Tange K, Kasai K, Miyata Y: Primary osteosarcoma of the skull —a case report and review of the literature. *J Neurooncol* 17: 81-88, 1993
10) Smeele LE, Kostense PJ, van der Waal I, Snow GB: Effect of chemotherapy on survival of craniofacial osteosarcoma: a systematic review of 201 patients. *J Clin Oncol* 15: 363-367, 1997
11) Tanaka S, Nishio S, Morioka T, Fukui M, Kitamura K, Hikita K: Radiation-induced osteosarcoma of the sphenoid bone. *Neurosurgery* 25: 640-643, 1989
12) Vege DS, Borges AM, Aggrawal K, Balasubramaniam G, Parikh DM, Bhaser B: Osteosarcoma of the craniofacial bones. A clinico-pathological study. *J Craniomaxillofac Surg* 19: 90-93, 1991
13) Whitehead R, Melhem ER, Kasznika J, Eustace S: Telangiectatic osteosarcoma of the skull base. *AJNR Am J Neuroradiol* 19: 754-757, 1998

Address reprint requests to: T. Hayashi, M.D., Department of Neurosurgery, Keio University School of Medicine, 35 Shinano-machi, Shinjuku-ku, Tokyo 160-8582, Japan.