Classical intracranial chondrosarcoma: A case report

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Abstract. Intracranial chondrosarcoma is a rare malignant cartilage-forming tumor, with only a small number of cases in the posterior cranial fossa reported previously. The present study reports the case of a 40-year-old male patient who was admitted to Tianjin Huanhu Hospital with a progressive headache and dizziness that had lasted for 2 years. Physical and neurological examinations were normal. Radiography of the skull identified an opaque lesion in the left frontal region of the brain. Cranial computed tomography and magnetic resonance imaging revealed a lesion with calcification and homogenous contrast enhancement in the left frontal region. Subsequently, the patient underwent bicoronal craniotomy and gross total resection of the tumor. Pathological examination confirmed the diagnosis of classical intracranial chondrosarcoma. The patient was discharged 10 days after surgery, with no neurological deficit. One month after initial discharge, the patient underwent γ-knife treatment. A follow-up examination 9 months after surgery revealed that the patient was still alive and had returned to work, with no obvious symptoms or evidence of recurrence.

Introduction

Primary intracranial chondrosarcoma is an extremely rare malignant tumor of the central nervous system, which accounts for <0.16% of all primary intracranial tumors (1). The tumor most commonly arises from the skull base, however, cases originating from the choroid plexus, dura matter and brain parenchyma have also been reported (2). As a result of their slow growth rate, intracranial chondrosarcomas do not usually metastasize until the very late stages of the disease. The symptoms vary among patients, although a long-standing history of headaches and signs associated with intracranial pressure are the main symptoms. Furthermore, dizziness, tinnitus, sensory disturbances of the face, and decreased visual acuity have been reported in some cases (3).

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Cranial computed tomography (CT) and magnetic resonance imaging (MRI) can aid the diagnosis of these tumors, although a clinical pathological diagnosis is the gold-standard. Radical excision is the standard treatment for intracranial chondrosarcoma, and postoperative adjuvant radiotherapy is the preferred treatment for remnants of the lesion (4). The prognosis of patients with intracranial chondrosarcoma is strongly influenced by a number of factors, including the use of postoperative adjuvant radiotherapy, pathological patterns, previous treatment (surgery or radiotherapy) and the extent of tumor removal. The overall 5-year mortality rate among patients in a previous study was 11%, with an average survival time of 53.7 months (5).

Histologically, three variants of chondrosarcoma have been defined: Myxoid, mesenchymal and classic chondrosarcoma. In this study, the case of a patient with low-grade, classic intracranial chondrosarcoma in the left frontoparietal region, which was misdiagnosed as meningioma preoperatively, is presented.

Case report

A 40-year-old male patient was admitted to The Department of Neurology, Tianjin Huanhu Hospital (Tianjin, China) on February 24, 2014 with a progressive headache and dizziness that had lasted for 2 years. Neurological examinations (including for hemiparesis and sensory disturbance) were normal and pathological signs (such as the Babinski sign) were negative. All laboratory tests (including routine blood count tests and assessments of liver and kidney, immune and blood coagulation functions) were within the normal ranges. The patient had no history of trauma and no family history of any hereditary illness. Cranial CT and an MRI scan revealed a mass in the left frontal region. CT scan revealed an iso-/hyperdense mass with unclear boundaries located in the left frontoparietal region (Fig. 1). Multiple patchy calcification shadows were present at the margins of the mass. Volumetric contrast-enhanced MRI revealed a heterogeneous extra-axial mass with clear margins (Fig. 2). The superficial margin of the mass was broad and based along the inner table of the skull; the center of the mass exhibited hyperostosis. The lesion exhibited a signal intensity that was higher than the grey matter on T1-weighted images and T2-weighted images. Based on the results of preoperative imaging, left frontal meningioma was suspected. The patient underwent tumor resection using the left frontal approach on February 28, 2014. Grossly, the tumor was attached to the tentorium and adjacent dura and was brown in color, hypervascularized and hard with clear boundaries. A dural incision was

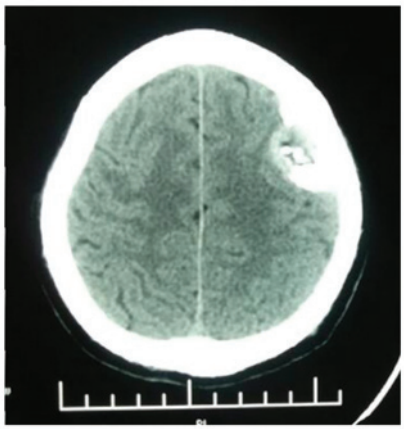

Figure 1. Pre-operative computed tomography revealed an iso-/hyperdense mass with unclear boundaries located in the left frontoparietal region.

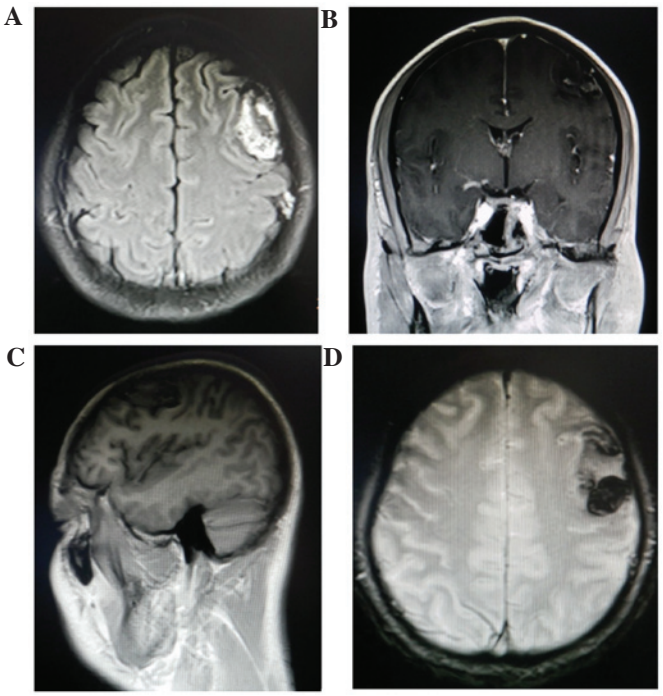

Figure 2. (A) Horizontal, (B) coronal and (C) sagittal pre-operative MRI scans of the left frontoparietal region revealed a mass with a signal intensity higher than that of the grey matter on T1-weighted images. (D) Postoperative cranial MRI revealed no residual tumor. MRI, magnetic resonance imaging.

made along the tumor-brain interface and the involved dura and tumor were completely removed. Analysis of a frozen tumor biopsy showed that the tumor tissues were lobulated, contained blood vessels and exhibited nuclear enlargement, which indicated meningioma.

On histopathological evaluation, the mass was composed of undifferentiated round or spindle-shaped cells and mature cartilaginous tissue (Fig. 3). Immunohistochemical examination revealed that the well-differentiated round cells were positive for vimentin (anti-vimentin antibody; cat. no. ab52942; 1:500; Abcam, Cambridge, UK) and S100 (anti-S100 antibody; cat. no. ab66041; 1:500; Abcam), and only scattered proliferating cells were positive for the proliferative marker Ki-67 (labeling index, 4.2%; anti-Ki-67 antibody; cat. no. ab833; 1:500; Abcam).

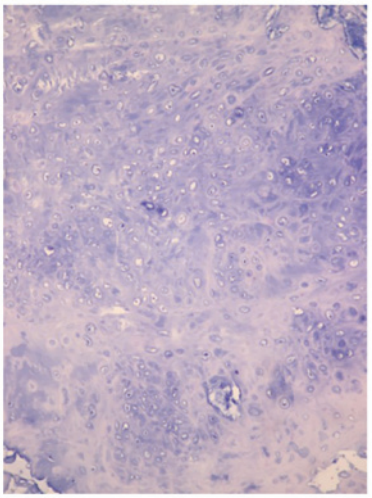

Figure 3. Histopathological evaluation revealed that the mass was composed of undifferentiated round or spindle-shaped cells and mature cartilaginous tissue (stain, hematoxylin and eosin; magnification, x200).

Thus, a pathological diagnosis of classical chondrosarcoma was confirmed. Postoperative cranial MRI identified no residual tumor (Fig. 2). The patient was discharged on the 10th postoperative day with no neurological deficits.

One month later, the patient underwent adjuvant γ-knife treatment (total dose, 60 Gy; 2 Gy once a day for 6 weeks). Follow-up MRI scans were taken at 1, 3 and 6 months after radiosurgery. A follow-up serial MRI performed 9 months postoperatively revealed no evidence of recurrence. The patient is currently alive and shows no obvious symptoms. Informed consent was obtained from the patient for the publication of this study.

Discussion

Intracranial chondrosarcoma, a subtype of chondrosarcoma, is a rare malignant cartilaginous tumor that was first reported by Mott in 1899 (6). Intracranial chondrosarcoma typically affects patients in the fourth and fifth decades of life, with no gender preference (7). The clinical presentation of chondrosarcomas has been extensively reported in the literature (8,9). Generally, patients present with an extensive history of headaches and symptoms associated with increased intracranial pressure. Histologically, intracranial chondrosarcomas are classified into three subtypes: Well-differentiated (classical type), intermediate (myxoid type) and undifferentiated (mesenchymal type) (10,11). In a review of 192 chondrosarcoma cases by Chandler *et al* (12), 62% were of the classical subtype, while the mesenchymal and myxoid types accounted for 30 and 8% of cases, respectively. Korten *et al* (8) reviewed 192 cases of chondrosarcoma and reported that in general, the mesenchymal type is malignant and occasionally spreads to distant areas, while the classical subtype is the most benign of the three subtypes. In this study, a case of classical type intracranial chondrosarcoma that occurred in the left frontal region of the skull was presented, which has rarely been reported in the literature to date. Generally, classical chondrosarcomas occur in the base of the cranium and affect patients between the fourth and sixth decades of life (3). In the review of chondrosarcomas by

Korten *et al* (8), 37% of tumors were located in the petrous bone, while 23% occurred in the occipital bone and clivus, 20% in the sphenoid bone and 14% in the frontal, ethmoidal and parietal bones; the remaining 6% were in dural tissue, which does not typically contain cartilage.

CT scans usually reveal an isodense/hyperdense mass with heterogeneous enhancement and varying degrees of calcification in patients with chondrosarcomas (3,13). MRI usually reveals a hypointense mass on T1-weighted images and an extremely hyperintense mass on T2-weighted images (1,14).

On immunological examination, chondrosarcomas usually exhibit significant positivity for S100 and vimentin, while only scattered proliferating cells exhibit positivity for the proliferative marker, Ki-67. These features differentiate chondrosarcoma from meningioma, hemangiopericytoma, metastasis and vascular malformations.

Radical excision is the standard treatment for intracranial chondrosarcoma. In addition, postoperative adjuvant radiotherapy has been reported to improve patient outcomes for intracranial chondrosarcoma (15). Due to the invasive nature of chondrosarcoma, adjuvant radiotherapy may be recommended even after successful radical resection (4). According to a previous study, the 5-year recurrence rate for chondrosarcoma patients treated with surgery alone is 44%, which is markedly reduced to 9% following the addition of adjuvant radiation therapy (16). Combined surgical and postoperative proton radiation therapy has also demonstrated promising results with regard to tumor control (17). In the present case, the tumor was located at the surface of the left frontal region, and exhibited good dissection margins from the surrounding tissue. In the present case, the patient was treated with adjuvant radiotherapy following surgery, and subsequently exhibited a favorable prognosis after total resection.

In conclusion, intracranial chondrosarcoma is a rare malignant cartilaginous tumor that generally arises from the base of the skull. Due to its rarity and similar imaging findings with meningioma, a differential diagnosis is often challenging. Pathological diagnosis is the gold standard and neurosurgical resection is the mainstay of therapy, although, as a result of its high propensity for recurrence, radiotherapy is often necessary. According to this rare case of a low-grade, classic intracranial chondrosarcoma, the present study has provided a more objective protocol for clinicians managing these patients.

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