Skull Metastasis From Clear Cell Chondrosarcoma

—Case Report—

Takayuki KAWANO, Jun-ichiro HAMADA, Motohiro MORIOKA, Yosuke MIHARA*, Yasuji ISHIMARU**, Motoyuki KAKU*, Yukitaka USHIO, and Jun-ichi KURATSU

Department of Neurosurgery, Kumamoto University Hospital, Kumamoto; *Department of Neurosurgery, Shin-Beppu Hospital, Beppu, Oita; **Department of Anatomy and Pathology, Kumamoto University School of Health Sciences, Kumamoto

Abstract

A 67-year-old man presented with left lower cranial nerve paresis and dysfunction of the left cerebellar hemisphere 4 years after amputation of the left lower leg because of clear cell chondrosarcoma (CCC). Neuroimaging studies showed an osteolytic extradural mass with homogeneous enhancement in the left posterior fossa. Bone scintigraphy disclosed a single high-uptake lesion at the same site. The tumor was removed totally via a left suboccipital craniotomy. Histological examination found mainly clear cells arranged in a microlobular pattern separated by thin fibrovascular stroma. The nuclei were regular with few mitotic figures. Immunohistochemical staining showed the tumor cells reacted intensely for both S-100 protein and vimentin. Osteoclast-like multinucleated giant cells were found at the periphery of the lobules. The primary tumor showed the same findings and the metastatic tumor manifested no malignant change. The histological diagnosis was metastatic CCC. CCC is a very rare neoplasm with slow growth and low-grade malignancy. Distant metastasis is rare but can occur in the skull base bone despite radical resection of the primary tumor. Osteolytic findings of homogeneous enhancement on magnetic resonance imaging and a high uptake on bone scintigraphy are indicative of metastatic tumor from previous CCC.

Key words: clear cell chondrosarcoma, skull base, metastasis

Introduction

Clear cell chondrosarcoma (CCC), first described in 1976, is a cartilaginous neoplasm consisting largely or entirely of clear cells, in contrast to conventional chondrosarcoma. CCC is very rare, accounting for about 1–2% of chondrosarcomas of bone, and shows slow growth and low-grade malignancy. CCC may be discovered in the 2nd to the 9th decade, but mostly in the 3rd and 5th decade of life, in younger patients than those with conventional chondrosarcoma. The male:female ratio is 2–2.6:1. CCC manifests as radiological osteolytic destructive lesions. The proximal portion of the femur is the most common site (over 50% of cases), followed by the proximal portion of the humerus and the knee. The major symptom is localized pain and pathological fracture is not unusual. Complete removal of the lesion by adequate en bloc resection is desirable, as initial treatment by curettage results in local recurrence exceeding 80%, and overall mortality of 15%, although radical resection has led to improved mortality. CCC is less aggressive than conventional chondrosarcoma and distant metastasis is very rare.

We describe an extremely unusual case of metastatic CCC in the left posterior skull base bone.

Case Report

A 67-year-old man had undergone amputation of the left lower leg because of a malignant tumor at another hospital 4 years earlier. The histological diagnosis was CCC. Three months before reporting to our hospital, he developed hoarseness, numbness

Received September 14, 2004; Accepted January 14, 2005
of the left side of the tongue, and pain radiating to
the left occiput and shoulder. On admission, he had
hoarseness and dysphagia. His tongue deviated to
the left on protrusion and showed atrophy without
fasciculation. His left sternocleidomastoid muscle
also showed atrophy. Furthermore, he had dys-
diadochokinesia and dysfunctional coordination on
the left. These signs indicated disturbance of the left
lower cranial nerves (nerves IX–XII), and dysfunc-
tion of the left cerebellar hemisphere. Laboratory
examination of blood and serum, including serum
alkaline phosphatase, showed no abnormalities.

Computed tomography disclosed osteolytic
change with a tumor of the left occipital bone and
destruction of the left jugular foramen (Fig. 1A). T₁-
weighted magnetic resonance imaging with
gadolinium-diethylenetriaminepenta-acetic acid
showed homogeneous enhancement of the tumor lo-
cated below the left cerebellum, with good demarca-
tion from the cerebellar hemisphere, and extension
toward the left pyramidal bone (Fig. 1B, C). There
was a distinct border between the tumor and the
cerebellar hemisphere, suggesting an extradural
skull bone tumor. Angiography showed moderate
vascularity of the tumor supplied by the left
posterior meningeal artery (Fig. 1D). Bone scin-
tigraphy obtained 4 years earlier had shown a single
high-uptake lesion in the distal portion of the left
tibia (Fig. 1E); and on this admission a single high-
uptake lesion in the left posterior cranial fossa
(Fig. 1F).

Left suboccipital craniotomy was performed.
The left occipital bone was fragile and hemorrhagic.
The tumor was grayish and hard elastic, had invaded
the overlying muscles, and was tightly attached to
the dura mater. The feeding artery was coagulated
and the tumor was removed together with the
attached muscles. The fragile bone continued deep
into the occipital base and was removed up to the
jugular foramen. Opening of the dura showed that
the lower cranial nerves were in the normal
positions and appeared to be normal. The surface of
Fig. 2  A, B: Photomicrographs showing the tumor cells with a central vesicular nucleus surrounded by clear cytoplasm with distinct borders, and regular nuclei without mitotic figures. Osteoclast-like multinucleated giant cells are seen at the periphery of the lobules (arrows). HE stain, original magnification ×200.  C, D: Photomicrographs with toluidine blue staining showing the presence of cartilaginous tissue (C), and with periodic acid-Schiff staining suggesting the presence of glycogen particles in the cytoplasm (D). Original magnification ×200.  E–G: Immunohistochemical staining with antibodies against vimentin (E) and S-100 protein (F) showing positive reaction in the cytoplasm, and with KP-1 antibody (G) showing positive reaction in the osteoclast-like multinucleated giant cells. Original magnification ×200.

Neurol Med Chir (Tokyo) 45, July, 2005

Discussion

Of the rare cases with distant metastasis from CCC, the most common metastatic site is the lungs, and few cases with metastasis in ribs, skeletal bones, and vertebrae were reported. Radical resection decreased the local recurrence rate, but distant metastasis occurred even after radical resection. In our patient, metastasis occurred even after amputation of the lower leg. Three cases of CCC with metastatic lesions included dedifferentiated components. Although dedifferentiated change may explain the occurrence of metastasis after radical resection, our patient lacked dedifferentiated components. The mechanism(s)
underlying CCC metastasis remain unclear.

A high level of serum alkaline phosphatase was a marker of primary CCC in three of six cases, but our patient had normal levels. Interestingly, whole-body bone scintigraphy demonstrated the metastatic lesion in our case, suggesting that periodic bone scintigraphy is valuable for the long-term follow-up of patients treated by radical resection of primary CCC.

The metastatic tumor could be removed completely because no invasion or tight adhesion to adjacent tissue were present. Although radical resection of metastatic CCC in the skull region is recommended, the presence of important structures such as the internal carotid artery may make this approach impossible. Radiation therapy is not currently performed for CCC and the effectiveness of chemotherapy remains to be established, so the development of adjuvant therapy is necessary to protect patients with CCC from recurrence and metastasis.

References

9) Nakamori K, Murota K, Tomita Y: [Immunohistochemical and ultrastructural aspects of two cases of clear cell chondrosarcoma]. Byori To Rinsho 11: 361–366, 1993 (Jpn, with Eng abstract)

Address reprint requests to: M. Morioka, M.D., Department of Neurosurgery, Kumamoto University School of Medicine, 1–1–1 Honjo, Kumamoto 860–8556, Japan.

E-mail: morioka@kaiju.medic.kumamoto-u.ac.jp