Solitary Plasmacytoma of the Skull Vault
—Case Report—

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Abstract
A 55-year-old female presented with a rare solitary plasmacytoma restricted to the skull vault without systemic myelomatosis. She had a 9-month history of a slowly growing soft mass in the right temporal region. Neuroradiological examination revealed a skull defect and an extradural tumor. The whole layer of the skull was destroyed by the tumor, but the dura was not involved. The tumor was totally removed, and postoperative radiotherapy was given. Follow-up examination after 2 years showed she was free from local recurrence or evidence of systemic involvement. Complete surgical resection with adjuvant radiation therapy is the treatment of choice. Although the prognosis is good, regular lifelong examinations for myelomatosis are required.

Key words: plasmacytoma, solitary plasmacytoma, skull vault

Introduction
Plasmacytoma of the skull is usually a manifestation of systemic myelomatosis or multiple myeloma, and occurs as multiple lesions. Solitary lesions are not unusual in the skull, but usually occur in systemic myelomatosis. True solitary plasmacytomas of the skull vault without signs of systemic myelomatosis are very rare, with only about 20 such cases reported. This excludes plasmacytomas involving the skull base since lesions originating from the nasopharynx, oropharynx, or paranasal sinuses, which are defined as extramedullary plasmacytomas, often invade the skull base secondarily, and are difficult to differentiate from true skull plasmacytomas. Furthermore, lesions infiltrating the skull base are unlikely to be solitary and carry a high risk of progression to multiple myeloma. In contrast, the prognosis for solitary plasmacytoma of the skull vault is good when there is no evidence of systemic myelomatosis. Some authors say that postoperative radiotherapy is not required when the tumor is completely resected, although it is given in most cases.

We describe a patient with a solitary plasmacytoma of the skull treated by surgery.

Case Report
A 55-year-old female first noted a painless, soft swelling in the right temporal region in October 1992. The swelling progressively grew and became slightly tender in June 1993. A skull tumor was diagnosed at a local hospital and she was transferred to our clinic in July 1993.

Physical examination found a soft swelling, 7 x 5 cm, in the right temporal region extending to the lateral supraorbital region, with normal overlying skin. Neurological examination found no abnormalities. Computed tomography (CT) showed a large extradural mass in the right temporal region, with homogeneous enhancement except for a small portion in the center after intravenous administration of contrast material (Fig. 1). Bone window CT disclosed a lytic lesion involving the whole layer of the skull. The inner and outer tables were partly pushed apart at the edge of the defect (Fig. 2). Magnetic resonance (MR) imaging showed that the mass was mostly isointense with the brain parenchyma on both T1- and T2-weighted images and homogeneously enhanced by gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA), except for a small area in the center of the mass which was hypointense on both
Fig. 1 CT scans showing an extradural high-dense mass in the right frontotemporal region destroying the skull (left column), and homogeneous enhancement of the tumor (right column).

Fig. 2 Bone window CT scans (W 4000, L 200) showing destruction of the whole layer of the skull.

T1- and T2-weighted images. There was a line of low signal intensity between the tumor and the brain on the T2-weighted image, indicating that the dura had prevented tumor invasion of the brain (Fig. 3). Chest radiographs revealed no pathological findings. Neither bone scintigraphy using technetium-99m-hydroxy methylene diphosphonate nor gallium-67-citrate scintigraphy showed any hot spots throughout the body. Cerebral angiography showed a marked tumor stain which was fed by the middle meningeal and accessory meningeal arteries (Fig. 4 left).

Laboratory examinations found red blood cell count of $4.38 \times 10^6$/mm$^3$, hemoglobin 13.5 g/dl, white blood cell count 3700/mm$^3$, platelet 203,000/mm$^3$, total serum protein 7.0 g/dl, globulin 2.91 g/dl, gamma globulin 13.5%, serum calcium 9.3 mg/dl, and serum phosphorus 4.0 mg/dl, which were all within the normal range. Other serum electrolytes were also within the normal range. Only alkaline phosphatase, 178 IU/l, was above the normal limit of 125 IU/l. The immunoelectrophoresis of serum proteins showed normal levels of immunoglobulins (Igs) with IgG 1350 mg/dl, IgA 161 mg/dl, and IgM 230 mg/dl. A urine test for Bence Jones protein was negative. Bone marrow aspiration revealed no evidence of systemic myelomatosis. Tumor markers such as carcinoembryonic antigen, alpha fetoprotein, and carbohydrate antigens 19-9 and 125, and squamous cell carcinoma-related antigen were all within the normal range.

The feeding arteries were successfully embolized using polyvinyl alcohol and Hilal Microcoil\textsuperscript{TM} (COOK Inc., Bloomington, Ind., U.S.A.) on August 11. The tumor stain disappeared almost completely (Fig. 4 right). She underwent a frontoparietotemporal craniotomy on August 18. The tumor was encapsulated, and there was no adhesion to the overlying skin and temporal muscle. The yellowish tumor was soft and partly elastic, and easily separated from the dural surface. The dura was left intact. Bleeding from the tumor was minimal. The frontal sinus was destroyed in the right upper corner, but the mucous membrane inside was intact. The lateral part of the orbital roof was also destroyed, but the periorbital membrane remained intact. The skull defect was sharply demarcated and there was no osteoplastic reaction along the edge. The bone defect was widened to ensure the complete removal of the tumor. The skull defect was reconstructed using the inner table separated from the parietal bone flap. The outer table was returned to the parietal defect. Histological examination of the specimen showed plasmacytoma. The cells were stained for Ig kappa light chain, but not for lambda light chain (Fig. 5).

She received a postoperative radiotherapy of 50 Gy and was discharged without neurological deficit. At the follow-up examination 2 years after the operation, she was doing well. MR imaging showed no sign of local recurrence. The systemic bone scintigram was negative. She had no anemia, and im-
Immunoelectrophoresis of serum proteins showed a normal pattern. Serum electrolytes were within normal limits, with normalization of alkaline phosphatase. The urine test for Bence Jones protein was negative.

Discussion

Previous cases of solitary plasmacytomas of the bone have arisen from within the diploic space and expanded in both directions. Typically patients presented with a painless growing tumor, and normal skin overlying the mass. The tumor may extend subperiosteally, not involving the galea. Diagnostic criteria include: a radiologically solitary lesion, histological confirmation, negative bone marrow examination, negative immunoelectrophoresis, negative urine test for Bence Jones protein, no evidence of hyperglobulinemia, and absence of anemia. Furthermore, there should be no laboratory or radiological evidence of systemic dissemination for 2–3 years following the initial diagnosis. The present case meets all these criteria. The diagnosis of solitary plasmacytoma can never be absolutely certain on the basis of examination of bone marrow. Bone marrow involvement in multiple myeloma may be patchy, and the specimen may be taken from a section not involved by the disease.
Patients with an apparently solitary lesion might have occult multiple myeloma. Solitary plasmacytoma may be a stage of multiple myeloma or a variant of the same disease, a well-established entity unrelated to multiple myeloma, or a separate clinical entity with a different natural history. Multiple myeloma may present as a solitary mass of the skull. The marked similarity between multiple myeloma and the disseminated stage of solitary plasmacytoma of the bone in appearance, distribution, and multiplicity of the bone lesions suggests that solitary plasmacytoma of bone represents the same pathological process as multiple myeloma. Solitary plasmacytoma of the bone may represent an early stage of multiple myeloma which will eventually become multiple myeloma. On the other hand, the occasional cases of solitary plasmacytoma of the bone that remain solitary for the patient's life time imply a distinct entity. Multiple myelomas are invariably fatal in a few years, but solitary myelomas are apparently quite unpredictable in behavior. Some patients may have a benign course for several years with an apparent cure, but others may die due to dissemination within months.

The differential diagnosis includes meningioma, metastatic tumor, and multiple myeloma presenting as a solitary mass lesion of the skull. Histologically, plasmacytomas must be differentiated from plasma cell granulomas, inflammatory reaction, and meningiomas with plasma cell infiltrations. CT shows a tumor as a well-demarcated, high density, extradural mass which is homogeneously enhanced by intravenous administration of contrast material. Bone window CT shows a lytic lesion of the whole layer of the skull. Carotid angiography shows a tumor stain supplied by branches of external carotid artery, such as the superficial temporal, occipital, and middle meningeal arteries. The MR appearance of solitary plasmacytoma of the skull vault is not yet clear, but is supposed to be almost the same as that of a solitary lesion in multiple myeloma, i.e. isointense, hyperintense, or heterogeneous intensity compared with the brain parenchyma on T1-weighted images, and homogeneously enhanced by intravenous administration of Gd-DTPA. The present case showed all these neuroradiological features.

Surgical removal followed by postoperative irradiation is the treatment of choice. The tumor is very radiosensitive, and can be cured by biopsy or subtotal resection and radiation therapy. However, solitary plasmacytoma of the cranial vault has a good prognosis and radiotherapy may not be required if radical removal is achieved. Irradiation should be given only in cases with local recurrence.

Fig. 5 Photomicrograph of the tumor specimen showing oval tumor cells with diffuse proliferation of eccentric nuclei (upper: HE stain, ×200). Tumor cells are diffusely positive for Ig kappa chain (middle: ×200), and partly positive for leukocyte common antigen (lower: ×200).

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or in which the tumor was not completely re-
removed. On the other hand, complete resection of
solitary plasmacytoma of the skull without adju-
vant radiotherapy has lead to multiple bone metas-
tasis 3 years later. Although the tumor was totally
removed, our patient received 50 Gy irradiation to
be on the safe side. Since patients receiving 30 Gy or
less have suffered local recurrence, a dose of 40–60
Gy is recommended, but local recurrences or meta-
tasis have occurred even with higher doses. The
efficacy of chemotherapy for this tumor is not
certain. Chemotherapy with melphalan, cyclophos-
phamide, vincristine, vindesine, adriamycin, or
prednisolone alone or in combination has failed to
provide any benefit to patients. Multiple bone
involvement requires systemic chemotherapy, but
protracted courses of chemotherapy should be
avoided in patients with solitary plasmacytoma of
bone who have the potential for long-term survival
because of the high incidence of therapy-related
acute leukemia.

Solitary plasmacytomas of the cranial vault are
characterized by a good prognosis, if there is no evi-
dence of systemic involvement. Nevertheless,
there is a possibility of dissemination years later, so
regular follow-up examinations for myelomatosis
are required.

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