Clinical Case Report

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# Multiple myeloma with pathologically proven skull plasmacytoma after a mild head injury

## Case report

Wenzhen Yang, MD[a], Jing Zheng, MD[b], Ruihao Li, MD[a], Haijun Ren, MD[a], Boru Hou, MD[a], Zhiyong Zhao, MD[a], Dengfeng Wang, MD[a], Gang Wang, MD[a], Jixing Liu, MD[a], Guizhong Yan, MD[a], Dong Wang, MD[a], Xinding Zhang, MD[a,*]

**Abstract**

**Rationale:** MM is a malignant tumor originating from the plasma cells of the bone marrow. Central nervous system myelomatosis is very rare and may be a complication of MM.

**Patient concerns:** A 60-year-old man presented with a slowly growing soft mass at his right frontal scalp after a mild head injury 6 months ago.

**Diagnoses:** Neuroradiological examinations revealed a solid intracranial-extracranial mass with an osteolytic lesion in the skull. Histopathological examination showed skull plasmacytoma, and postoperative examinations revealed multiple myeloma.

**Interventions:** The tumor was completely removed and the skull defect repaired with the titanium mesh. Then, chemotherapy was initiated after surgery with bortezomib and dexamethasone.

**Outcomes:** The patient received eight chemotherapies within one year after surgery.

**Lessons:** Despite a history of head injury, a differential diagnosis should be kept in mind during the diagnosis of solid intracranial-extracranial masses, especially in the presence of osteolytic skull at the lesioned site.

**Abbreviation:** MM = multiple myeloma.

**Keywords:** head injury, multiple myeloma, neurosurgery, skull tumor

### 1. Introduction

Scalp masses are relatively common in the neurosurgical practice. After a mild head injury, a scalp hematoma is usually suspected.[1] However, other diseases, such as malignant tumors, cannot be excluded despite a history of a mild head injury. Herein, we report a case of multiple myeloma (MM) in a patient with radiologically suspected hemangiopericytoma and a history of mild head injury.

### 2. Case presentation

A 60-year-old man was admitted due to a frontal scalp mass on October 20, 2016. He complained that the scalp mass was present 6 months ago after his head slightly hit a car door. The mass was small soon after head injury, and there was no significant change during the first 5 months. There were no pain and no abnormal skin manifestations, and thus he paid no attention to the skull mass. In the prior month, the mass became larger gradually without headache and any history of loss of consciousness or seizure.

The physical examination and neurological examinations showed normal. His Glasgow Coma Scale Score was 15. Serologic tests for hepatitis, syphilis, and HIV infection showed negative. A CT scan revealed a large epidural mass with isointensity to the brain. And the skull at the site of mass was destroyed and became osteolytic (Fig. 1A). MRI revealed that a half of the right frontal mass was in the brain, and the remaining was out of the brain and invaded the skull. There was an obvious mass effect on the adjacent brain tissues. On T1-weighted and T2-weighted images, the mass showed isointensity to the brain, and had a homogeneous enhancement after Gd-DTPA injection (Fig. 1 B–H). The preoperative radiological diagnosis of the mass was hemangiopericytoma which should be differentiated from meningioma.

The patient underwent right frontoparietal craniotomy and removal of the mass due to the patient's requirement and the obvious mass effect on CT and MRI. After craniotomy, a reddish, rubbery, soft, and bloody tumor was observed; the tumor destroyed the skull and dura mater, but the brain tissues remained intact. Then, the tumor and the lesioned bone and dura mater were completely removed, and the skull defect was repaired with the titanium mesh (Fig. 2).

Post-operative pathological examination showed the skull plasmacytoma (Fig. 3). On pathological examination, the tumor cells consisted of plasma cells with different degrees of differentiation, showed diffused distribution, and had consistent size. The tumor cells were rich in cytoplasm which was stained

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*This study has been approved by the Ethics Committee of the 2nd Hospital of Lanzhou University. Patient gave informed consent for the case presentation in accordance with the Declaration of Helsinki.*

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*[a] Department of Neurosurgery, the 2nd Hospital of Lanzhou University, [b] Gynecology Department, Maternity and Child Care Hospital of Gansu Province, Lanzhou, Gansu Province, China.*

*[*] Correspondence: Xinding Zhang, Department of Neurosurgery, the 2nd Hospital of Lanzhou University, Lanzhou, Gansu Province 730030, China (e-mail: zhangxinding@126.com).*



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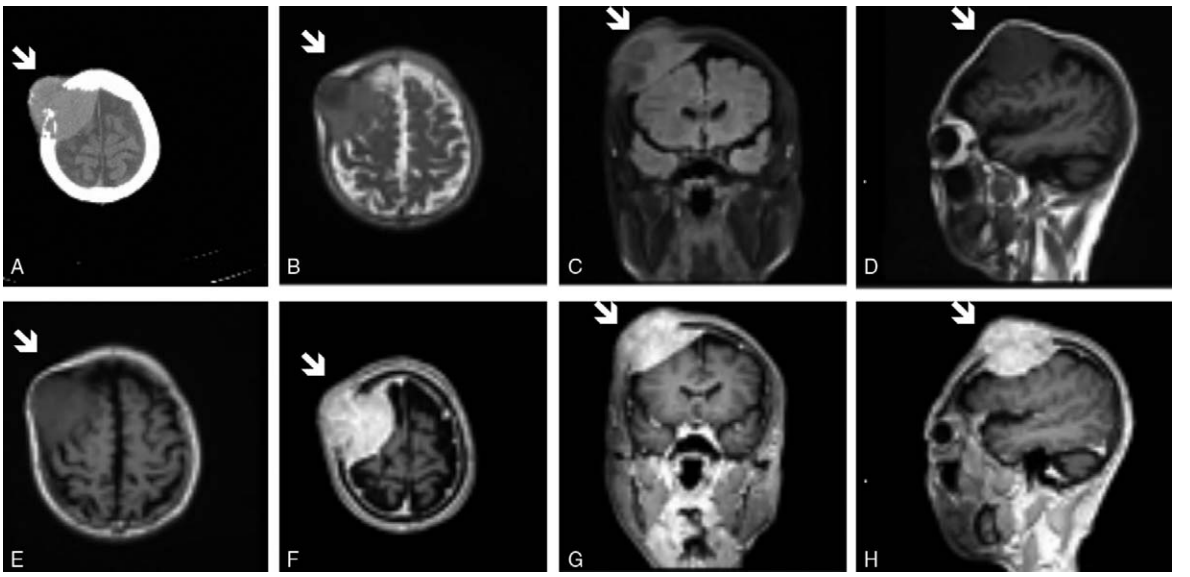


**Figure 1.** (A) CT showed a large epidural mass with isointensity to the brain. The skull at the mass was destroyed and became osteolytic. (B-H) Cranial MRI showed a half of the right frontal mass was in the cranial, and the remaining was outside the skull with skull involvement. There was an obvious mass effect on the adjacent brain. On T1-weighted and T2-weighted MR images, the mass showed isointensity to the brain, and homogeneous enhancement after Gd-DTPA injection.

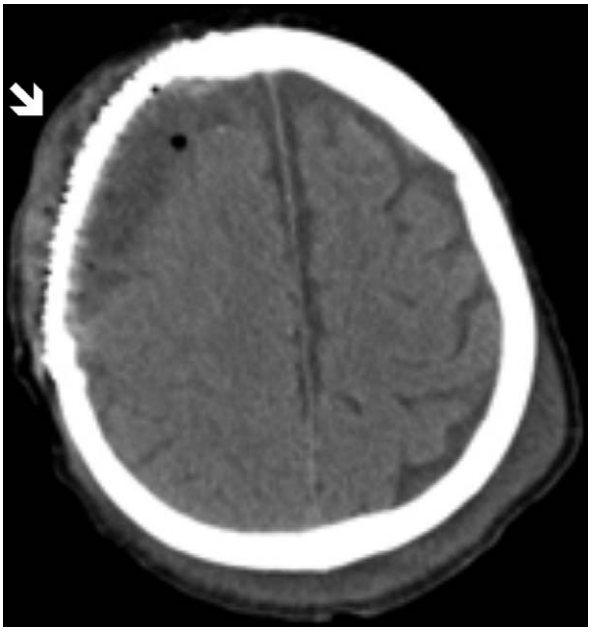

**Figure 2.** The bone and dura mater invaded by the mass were completely removed and the skull defect was repaired with the titanium mesh.

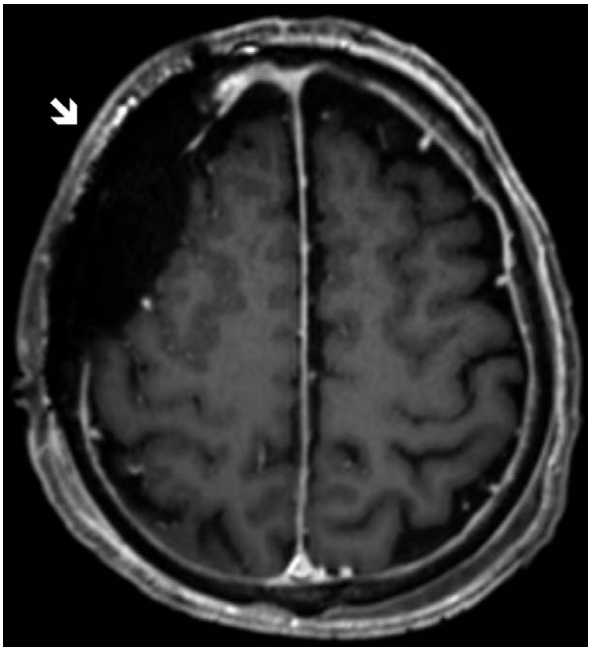

**Figure 4.** Post-operative MRI showed normal.

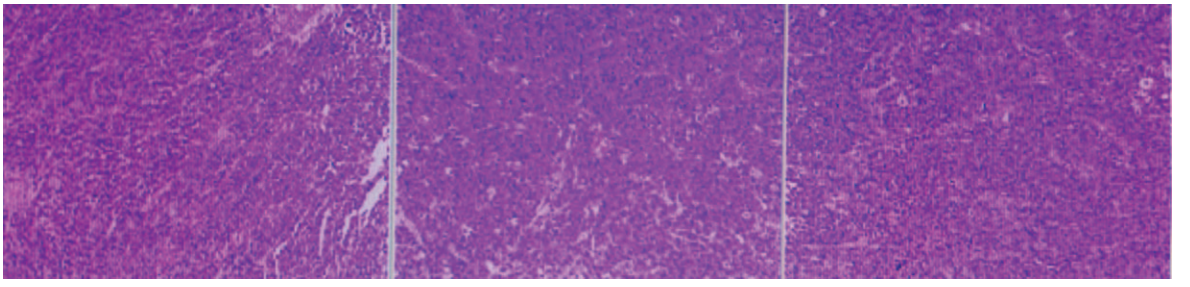

**Figure 3.** Pathological examination showed the skull plasmacytoma.

red; the nuclei were round and partially and slightly enlarged; the caryokinesis was common.

Immunohistochemistry showed the tumor cells were positive for Vim, D38, CD138, EBER, and K, but negative for λ, CD21, CD20, CD3, and CKp. The Ki 67 proliferation index was 60%.

The post-operative recovery was uneventful and abnormalities were not observed in post-operative MRI (Fig. 4). The post-operative whole body bone scan revealed that abnormal aggregation of radioactive Tc99m-MDP in the seventh thoracic vertebrae and the left tenth rib (Fig. 5). The post-operative bone marrow aspiration 2 weeks after operation (Fig. 6) and pathological examination showed active proliferation of bone marrow cells (G=62.00%, E=1800%, and G/E=3.4:1), granulocytosis, erythroid hyperplasia mostly with immature erythrocytes of metaphase and late phase, and mature erythrocytes of different sizes with good hemoglobin filling, and normal lymphocytes. There were totally 37 megakaryocytes and scattered platelets. Myelogram indicated proliferation of 3 lines. The post-operative bone marrow aspiration 3 months after operation (Fig. 7) and pathological examination showed active

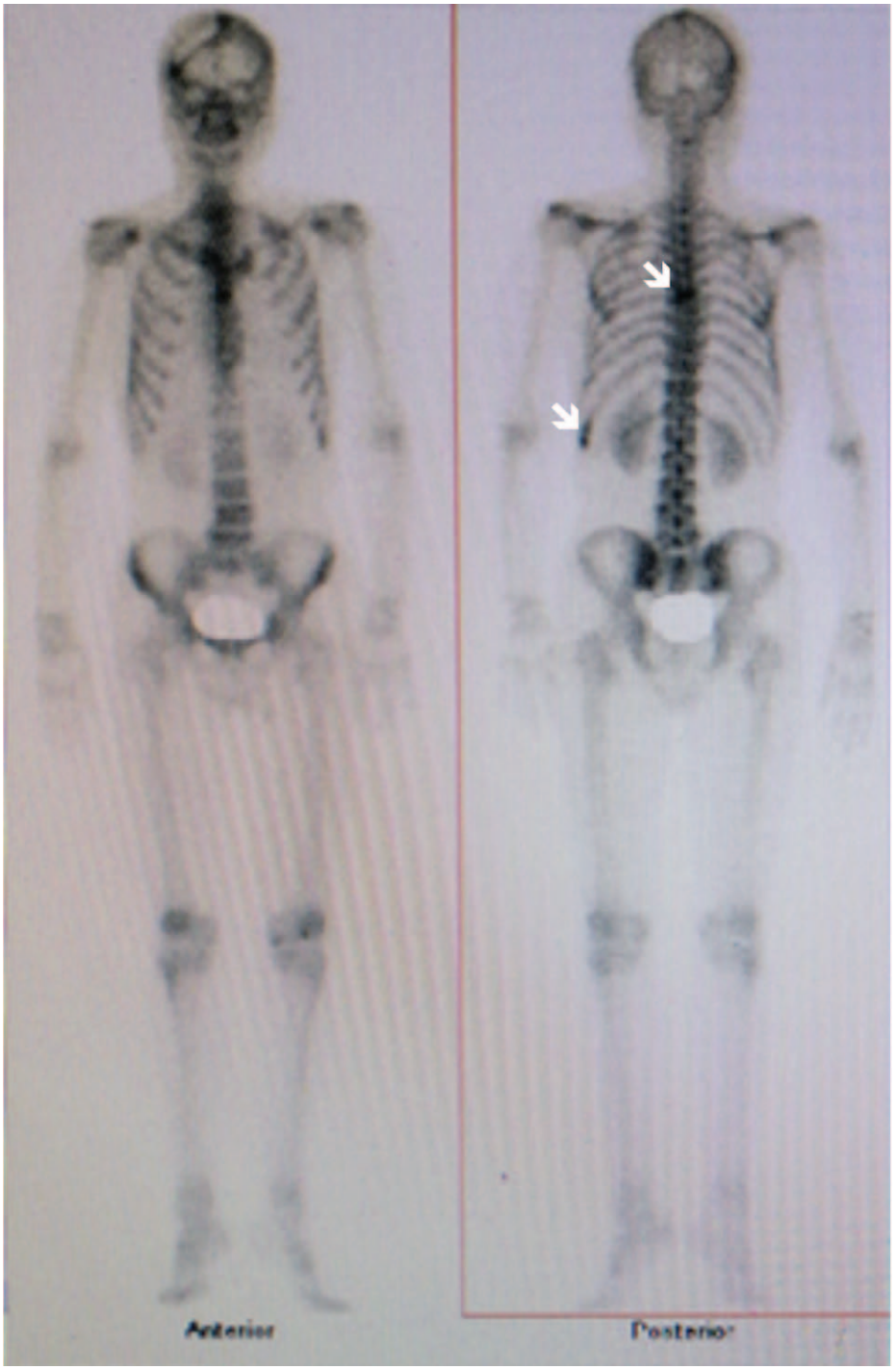


**Figure 5.** The post-operative whole body bone scan 10 days after surgery revealed that abnormal aggregation of radioactive Tc99m-MDP in the seventh thoracic vertebrae and the left tenth rib.

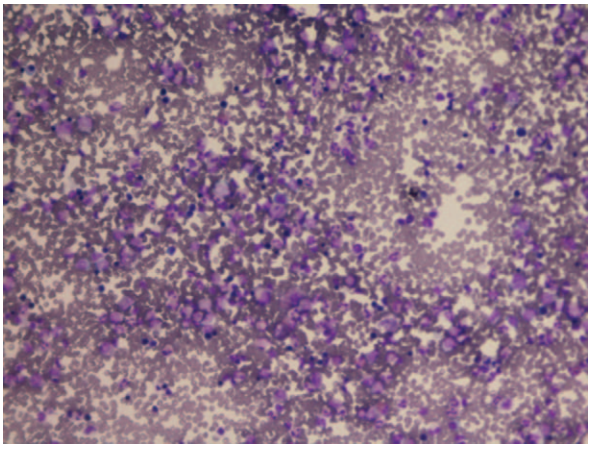

**Figure 6.** The post-operative bone marrow aspiration 2 weeks after operation and pathological examination showed active proliferation of bone marrow cells (G=62.00%, E=1800%, and G/E=3.4:1), granulocytosis, erythroid hyperplasia mostly with immature erythrocytes of metaphase and late phase, and mature erythrocytes of different sizes with good hemoglobin filling, and normal lymphocytes. There were totally 37 megakaryocytes and scattered platelets. Myelogram indicated proliferation of three lines.

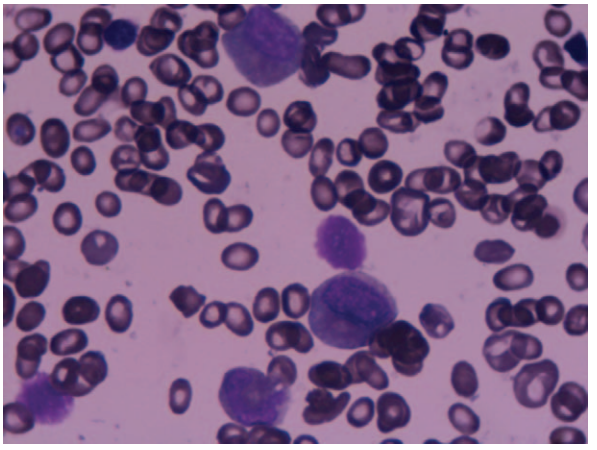

**Figure 7.** The post-operative bone marrow aspiration 3 months after operation and pathological examination showed active proliferation of bone marrow cells (G=51.00%, E=35.00%, and G/E=1.5:1), granulocytosis with normal proportion and morphology, erythroid hyperplasia mostly with immature erythrocytes of metaphase and late phase, and mature erythrocytes of different sizes with good hemoglobin filling, and relatively decreased lymphocytes. There were totally 72 megakaryocytes and scattered platelets. Myelogram indicated MM. MM=multiple myeloma.

proliferation of bone marrow cells (G=51.00%, and E=35.00%, G/E=1.5:1), granulocytosis with normal proportion and morphology, erythroid hyperplasia mostly with immature erythrocytes of metaphase and late phase, and mature erythrocytes of different sizes with good hemoglobin filling, and relatively decreased lymphocytes. There were totally 72 megakaryocytes and scattered platelets. Myelogram indicated MM. On the available findings from clinical examinations, the patient was diagnosed with MM. Then, the patient was treated with bortezomib and dexamethasone in the Department of Hematology. The patient received 8 chemotherapies within 1 year after surgery, who refused further chemotherapies later. There was no nausea, vomiting, headache, dizziness, or dysneuria during chemotherapies. There was persistent pain at right thigh after eighth chemotherapy, which did not affect walking. The patients refused re-exanimation at hospital, thus we speculated that the pain might be due to bone invasion of MM.

This patient was diagnosed with an isolate cranioaural lesion at admission and before surgery, but he was diagnosed with MM through pathology and bone marrow aspiration after surgery. Thus, this patient was diagnosed with systemic MM complicated with solitary skull plasmacytoma.

## 3. Discussion

MM is a malignant tumor originating from the plasma cells of the bone marrow. Central nervous system myelomatosis is very rare and may be a complication of MM. The central nervous system myelomatosis accounts for approximately 1% of plasma cell neoplasms,[2–5] and intracranial myelomatosis is less reported. Few studies have reported intracranial–extracranial mass involving the dura and calvarium.[6–8] As reported in the available studies, the prognosis of central intracranial myelomatosis is very poor, with the median survival time of $\leq$1 year.[9] The clinical diagnosis of intracranial myelomatosis is quite a challenge for not only neurosurgeons and radiologists, but also pathologists. In the diagnosis of intracranial myelomatosis, pathological examination is the gold standard. Radiological manifestations and pathological findings after bone marrow aspiration are helpful for its diagnosis. The confirmed diagnosis of solitary intracranial myelomatosis can only be made by post-operative pathological examination. As reported in studies, intracranial plasmocytomas generally appear as a skull or intracranial tumor exerting mass effect. Generally, the osteolytic skull can be identified in radiological examinations.[6–8]

No standard treatment exists for the intracranial myelomatosis partially due to its rarity. Studies have shown the surgical removal of lesioned skull and the tumor is required for the treatment of solitary plasmocytoma, and radiotherapy and chemotherapy may be initiated postoperatively.[5,7,9–11] In our case, the tumor and the lesioned skull were completely removed and thereafter patient was transferred to the Department of Hematology and received chemotherapy with bortezomib and dexamethasone.

A recent case series reported that a combination of intrathecal chemotherapy, radiotherapy, and immunomodulatory therapy could prolong the survival time of patients with central nervous system myeloma.[12] In another report, treatment with novel drugs (such as bortezomib, thalidomide, and lenalidomide) was also found to improve the overall survival significantly.[10] The main goals of chemotherapy for intracranial myelomatosis are the treatments of intracranial disease and systemic myelomatosis. Although the extent of novel drugs crossing the blood brain barrier is not clear, bortezomib has the promise to improve the clinical outcomes.[10,11,13]

Taken together, intracranial myelomatosis, as a complication of MM, is rare. Despite a history of head injury, this as a differential diagnosis should be kept in mind in the diagnosis of solid intracranial–extracranial masses, especially in the presence of osteolytic skull at the lesioned site shown in radiological examinations.

## Author contributions

**Conceptualization:** Wenzhen Yang.
**Data curation:** Wenzhen Yang.
**Formal analysis:** Wenzhen Yang.

**Investigation:** Wenzhen Yang, Jing Zheng, Ruihao Li, Haijun Ren, Boru Hou, Zhiyong Zhao, Dengfeng Wang, Gang Wang, Jixing Liu, Guizhong Yan, Dong Wang.
**Methodology:** Wenzhen Yang.
**Resources:** Wenzhen Yang.
**Supervision:** Xinding Zhang.
**Writing – original draft:** Wenzhen Yang.
**Writing – review & editing:** Xinding Zhang.

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