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Case Report

Multiple myeloma complicated by skull plasmacytoma discovered after head injury

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Plasmacytoma is a malignant tumor originating from the plasma cells of the bone marrow. Those discovered after a head injury is rare. We report a case of a 48-year-old female who complained of scalp mass without other symptoms after head injury. Meningioma was considered preoperatively based on imaging findings, and surgical resection was performed. Postoperatively, multiple myeloma complicated by skull plasmacytoma was diagnosed by histopathology and systematic examinations in succession. When evaluating a head mass that appeared after a head injury, plasmacytoma should be considered at times. Osteolytic changes and biconvex form on imaging are beneficial to differentiation.

Keywords

Head injury; Skull plasmacytoma; Multiple myeloma; Meningioma

1. Introduction

Plasmacytoma is a malignant tumor originating from the plasma cells of the bone marrow, mainly including multiple myeloma (MM) and solitary plasmacytoma (SP) [1]. The diagnosis for MM, defined by the International Myeloma Working Group, must meet both of the following criteria. First, clonal bone marrow plasma cells $\geq$10% or biopsy-proven bony or extramedullary plasmacytoma. Second, any of one or more myeloma-defining events, including evidence of end-organ damage (hypercalcemia, renal insufficiency, anemia, bone lesions) and biomarkers of malignancy (clonal bone marrow plasma cell percentage $\geq$60%; uninvolved serum-free light-chain ratio $\geq$100 mg/L; more than 1 focal lesion bigger than 5 mm on MRI studies) [2]. In addition, when there is a biopsy-proven solitary lesion of bone or soft tissue with evidence of clonal plasma cells and no evidence of MM, SP can be diagnosed. Solitary bone plasmacytoma has been reported to be most common in vertebrae; only 5% occur in the skull [3].

The presentation of asymptomatic scalp mass is joint in neurosurgery. However, plasmacytoma incidentally discovered after a head injury is rare. Herein, we report a case of a 48-year-old female diagnosed with MM complicated by skull plasmacytoma. Atypically, the only chief complaint was an asymptomatic scalp mass that appeared after a head injury. To our knowledge, only one similar case has been reported in English so far [4]. For neurosurgeons, a broad differential diagnosis is supposed to be made when evaluating a mass after injury.

2. Case presentation

A 48-year-old female was admitted to our hospital on 3 September 2020 for "collisional scalp mass for 4 months". Her personal and medical history were unremarkable. Four months previously, she accidentally hit her forehead against a door and initially had no complaints of pain or visible signs. Five days later, a 3 $\times$ 3-cm mass appeared suddenly at the site of the collision. The patient thought the mass was caused by injury; she paid no attention and waited for it to disappear. After four months with no change, she came to our hospital. On physical examination, she was found to have a soft mass on her left forehead, with no tenderness or mobility. Hard nodules were palpable within the mass. The Glasgow Coma Scale score was 15, and neurological signs were negative. Laboratory examinations revealed elevated urinary protein (>3 g/L), with other values being within the normal range. Computed tomography (CT) scan showed a 4.1 $\times$ 3.5-cm mass that had penetrated the skull, with a clear boundary on the left forehead (Fig. 1A,B). Magnetic resonance imaging (MRI) revealed abnormal signals on the convexity of the left frontal bone, with isointense T1 and T2 signals. The mass had a noticeable homogeneous enhancement. The frontal parenchyma was compressed and pushed inward (Fig. 1C–F).

A preoperative diagnosis of meningioma that had invaded the skull was considered because of the patient's young age, history of head injury, and imaging features. Therefore, a left craniotomy was performed. During the operation, a reddish, soft, and hemorrhagic tumor was observed. The tumor had eroded the skull and dura mater, leaving the leptomeninges and brain intact (Fig. 2A). We removed the tumor (Fig. 2B) and eroded bone completely (Fig. 2C,D), and reconstructed the skull defect. Postoperatively, the patient showed a hemoglobin decrease without signs of bleeding.

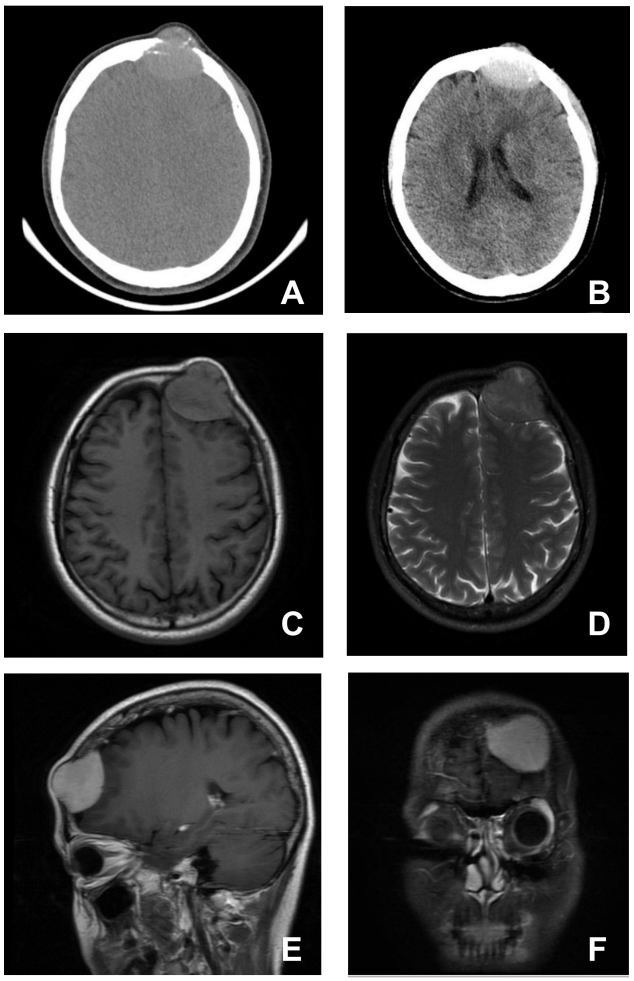

Fig. 1. (A,B) Cranial CT showed a 4.1 cm × 3.5 cm mass which had penetrated the skull with a clear boundary in the left forehead. High density is observed within the tumor. (C–F) Cranial MRI showed an iso-T1 and iso-T2 mass with a clear boundary. After Gd-DTPA injection, the mass was homogeneously enhanced. The frontal parenchyma was compressed and pushed inward.

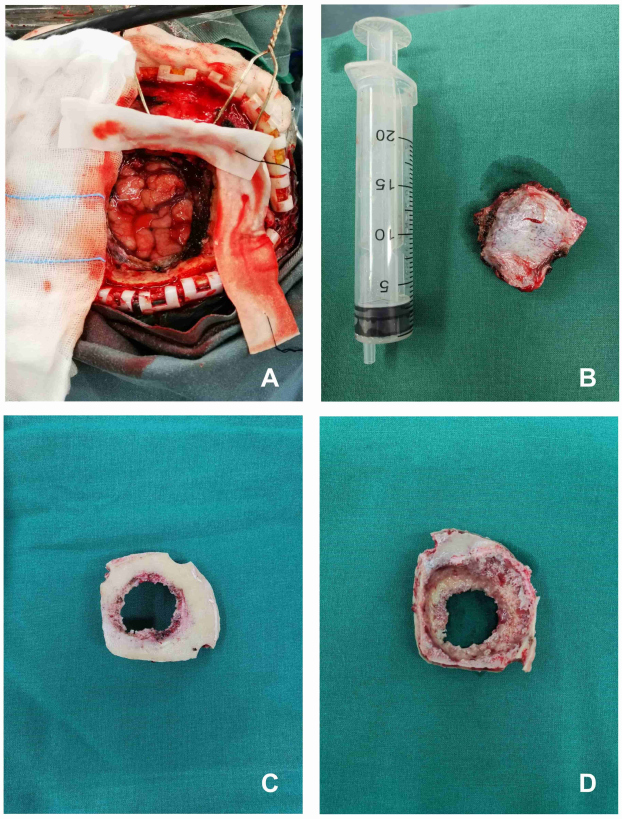

Fig. 2. (A) The intraoperative photograph showed that the pia mater and brain parenchyma were intact. (B) A reddish, soft, and hemorrhagic tumor was removed. (C,D) The tumor had eroded the inner plate of the skull obviously, remaining the outer plate thin.

Hemoglobin was 8 g/L on the first day after surgery and 5.8 g/L on the fourth day. After transfusion of 2U red blood cells, the anemia was corrected, and the patient recovered well.

Postoperative pathology examination showed plasma-cell neoplasm. Immunohistochemistry showed that the tumor cells were positive for CD138, CD38, lambda, MA and mum-1, but negative for CD20, CD79 α, SMA and Pax-5. The Ki 67 proliferation index was 30%. Therefore, the patient was preliminarily diagnosed with solitary plasmacytoma.

For further diagnosis, the patient went to the Department of Hematology two weeks after surgery. Bone-marrow aspiration and pathology examination showed active myeloid hyperplasia (granulocytes 48.5%, erythrocytes 16%, granulocytes : erythrocytes = 3.03 : 1), 23% plasma cell infiltration and erythrocyte rouleaux formation (Fig. 3). Further laboratory tests showed that 24-h urine protein was 9.85 g, M protein concentration was 3997 mg/L, total M protein was 9.1 g, and β2-microglobulin was 5.5 mg/L (0.7–1.8). A whole-body PET-CT scan showed 8 abnormal aggregations of radioactive Tc99m-MDP in the ribs and other areas (Fig. 4). Based on the above findings, the patient was finally diagnosed with MM combined with skull plasmacytoma.

For further treatment, VRd (bortezomib + lenalidomide + dexamethasone) chemotherapy was suggested. During a six-month follow-up, the patient had completed four courses of chemotherapy. She tolerated it well without nausea, vomiting, headache or other symptoms. Light-chain protein in blood turned negative, and the 24-h urine protein decreased to 0.06 g.

3. Discussion

For neurosurgeons, it is challenging to associate scalp mass, which was the only chief complaint and appeared after a head injury, with a malignant-tumor-like plasmacytoma. On the one hand, although scalp masses can be the initial presentation for MM, patients typically present with additional symptoms, which include acute weakness, bone pain, and confusion [5]. In addition, not all skull plasmacytoma can present with scalp mass (e.g., lesion in skull base) [1]. On the other hand, plasmacytoma incidentally discovered induced by injury is rare. To our knowledge, only one similar case has been reported in English so far [4], and we reported a case

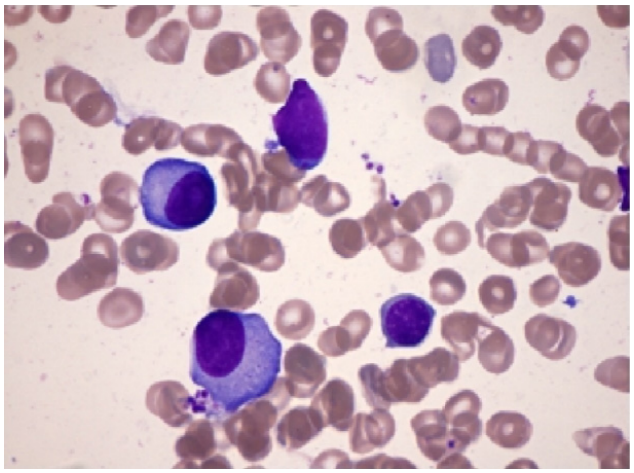

Fig. 3. Bone marrow aspiration and pathological examination showed that granulocytes accounted for 48.5%, erythrocytes accounted for 16%, granulocytes : erythrocytes = 3.03 : 1, plasma cell infiltrations was 23%. In addition, there was erythrocyte rouleaux formation (arrow).

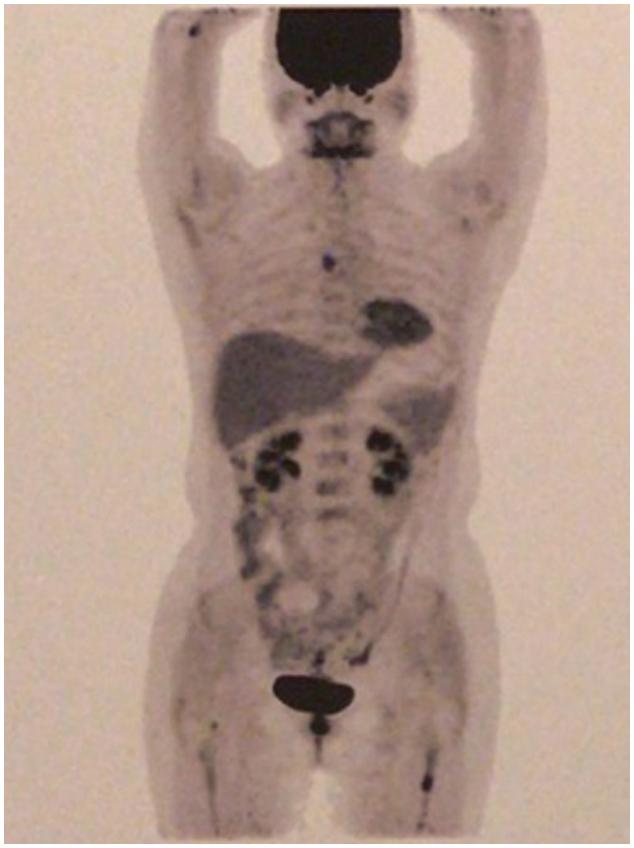

Fig. 4. PET-CT showed abnormal aggregations of radioactive Tc99m-MDP at 7th cervical spine, 10th thoracic vertebrae, 11th and 12th lumbar vertebrae, sternum, left rd anterior rib and bilateral femurs.

under the age of 50 for the first time.

Plasmacytoma incidentally discovered after a head injury is rare. Some head tumors are related to injuries, such as eosinophilic granuloma [6, 7] and meningioma [8]. The skull-bone cells may undergo genetic mutation during the process of wound repair, leading to tumors over a long period of time. The patient paid no attention in the present case and did not come to the hospital until four months later. However, in this case, the mass had appeared much earlier than four months. The injury might have led to a pathological fracture of the eroded skull, accelerating the extension of cranial plasmacytoma toward the scalp and forming a lump as a result. We believe that was why the mass appeared suddenly and expanded rapidly on the fifth day after the injury, as shown in this case. The high density shown on preoperative CT might be the skull fragment caused by a pathological fracture. During surgery, the outer plate of the eroded cranium was found to be very thin.

Multiple myeloma with scalp mass should be differentiated from other intracranial occupying lesions, including epidural hematoma, meningioma, eosinophilic granuloma, metastasis, chordoma, lymphoma, etc. However, it is most likely to be misdiagnosed as meningioma [9]. Therefore, it is essential to compare the features of meningiomas and skull plasmacytoma on imaging. Both of them can appear as a well-defined and homogeneously enhanced mass on CT. In addition, both the calcification of the meningioma and the bone fragments of skull plasmacytoma can show high density. However, there are some differences. First, on CT, meningioma usually causes hyperplasia and sclerosis of the adjacent bone [9], whereas SP shows osteolytic changes and lacks osteosclerosis and obvious periosteal reaction (Fig. 1A). Second, meningioma originates from the dura and can invade the skull, with the main body of the tumor usually on the inside of the skull [9]. In contrast, skull plasmacytoma originates from the skull, with the main body extending outside and presenting as a biconvex mass centered on the penetrated skull [10].

In reviewing the patients of MM combined with skull plasmacytoma who presented with scalp mass, we found that besides the typical complaint of bone pain, there were various symptoms due to brain parenchyma compressions, such as mood disorders and hemiplegia [6]. One patient had acute severe fatigue and insanity, deteriorated rapidly, and died [10]. The sensitivity of specific laboratory examinations, such as for M protein, is low. Some cases showed no M protein either in serum or urine [6]. A review of 1027 patients with MM showed that 80% had no M protein in their urine, and 3% of them had no M protein either in their urine or blood [11].

Surgical resection and radiotherapy (RT) are adequate for most SP [6]. According to NCCN guidelines, primary RT (40–50 Gy in 1.8–2.0 Gy/fraction) to the dynamic field is recommended and can provide excellent local control [1]. Better outcomes could reportedly be achieved for lesions smaller than 5 cm, although other studies did not find tumor size a factor [7]. The most extensive retrospective study showed no correlation between local failure and radiation dose for large tumors [3]. If progression to MM occurs, systemic therapy

like high-dose chemotherapy and autologous hematopoietic-cell transplant may be administered [1]. However, patients may die during conservative treatments like radiotherapy and chemotherapy [8, 10, 12]. If the lesion causes structural instability or neurologic compromise, surgery may be performed [1]. Surgery can relieve compression of brain parenchyma [6] and reduce the tumor burden. However, there is a risk of bleeding due to the rich blood supply of skull plasmacytoma [4–6]. In addition, the tumor may be stimulated by surgery. In that case, anemia after surgery might result from surgery-related hemorrhage, tumor activation, and temporary inhibition of bone marrow hematopoiesis. Therefore, the indication and opportunity for surgery should be further evaluated for risks and benefits.

4. Conclusions

We herein report a case of a 48-year-old female diagnosed with MM, complicated by skull plasmacytoma, presenting with scalp mass that appeared after head injury without other symptoms. Different intracranial lesions like meningiomas, osteolytic changes and biconvex form on imaging should be emphasized. For neurosurgeons, when evaluating a head mass that appeared after a head injury, a broad differential diagnosis should be considered. Systemic therapies are needed for these patients.

Abbreviations

MM, multiple myeloma; SP, solid plasmacytoma; sSP, skull solid plasmacytoma; CT, computed tomography; MRI, magnetic resonance imaging; RT, radiotherapy.

Author contributions

QY and XMM had equal contributions to the essay. QY, MF and XMM designed the research. QY collected clinical data of the patient. QY and XMM consulted the literature and wrote the manuscript. MF, YY, XJB, KD and RZW provided help on the essay writing and reviewed the manuscript. All authors contributed to editorial changes in the manuscript. Finally, all authors read and approved the final manuscript.

Ethics approval and consent to participate

The Research Ethics Committee of Peking Union Medical College (Beijing, China). All the procedures were performed according to ethical standards and practices. In addition, written informed consent for publication of case details and pictures was obtained from the patient and her husband.

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Conflict of interest

The authors declare no conflict of interest.

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