

## Case Report

# Solitary plasmacytoma of the skull: case report

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**Abstract:** Solitary plasmacytoma of the skull is very rare and only a few cases have been reported in the English literature. Skull solitary plasmacytoma tends to disseminate or progress to multiple myeloma (MM) during follow-up. If the skull lesion is isolated, couple with tenderness and marked swelling in the area, plasmacytoma should be considered, especially for the lesion showing isointense with the brain parenchyma on both T1- and T2-weighted magnetic resonance imagings. A 42-year-old female patient was diagnosed with skull solitary plasmacytoma after surgery because of preoperative imaging and physical examination suggesting isolated mass. However, the patient received further examinations after the diagnosis of MM being made according to bone marrow aspiration to predict the clinical course. The patients received postoperative chemotherapy. Survival time following surgery was 19-month.

**Keywords:** Plasmacytoma, skull, treatment

### Introduction

Plasma cell neoplasms arise from the proliferation of a single clone of B-cell lymphocytes, 3% of which present as solitary lesions [1]. It produces a wide spectrum of disorders, ranging from benign solitary plasmacytoma to malignant multiple myeloma (MM). Whether MM is the outcome of solitary plasmacytoma is still a debatable issue, however, the diagnosis criteria of solitary plasmacytoma has been built due to the better prognosis compared to MM. We are reporting a case of a solitary plasmacytoma of the skull, which may present an initial manifestation of MM with postoperative palpable clinical presentations compared to preoperative asymptomatic solitary lesion.

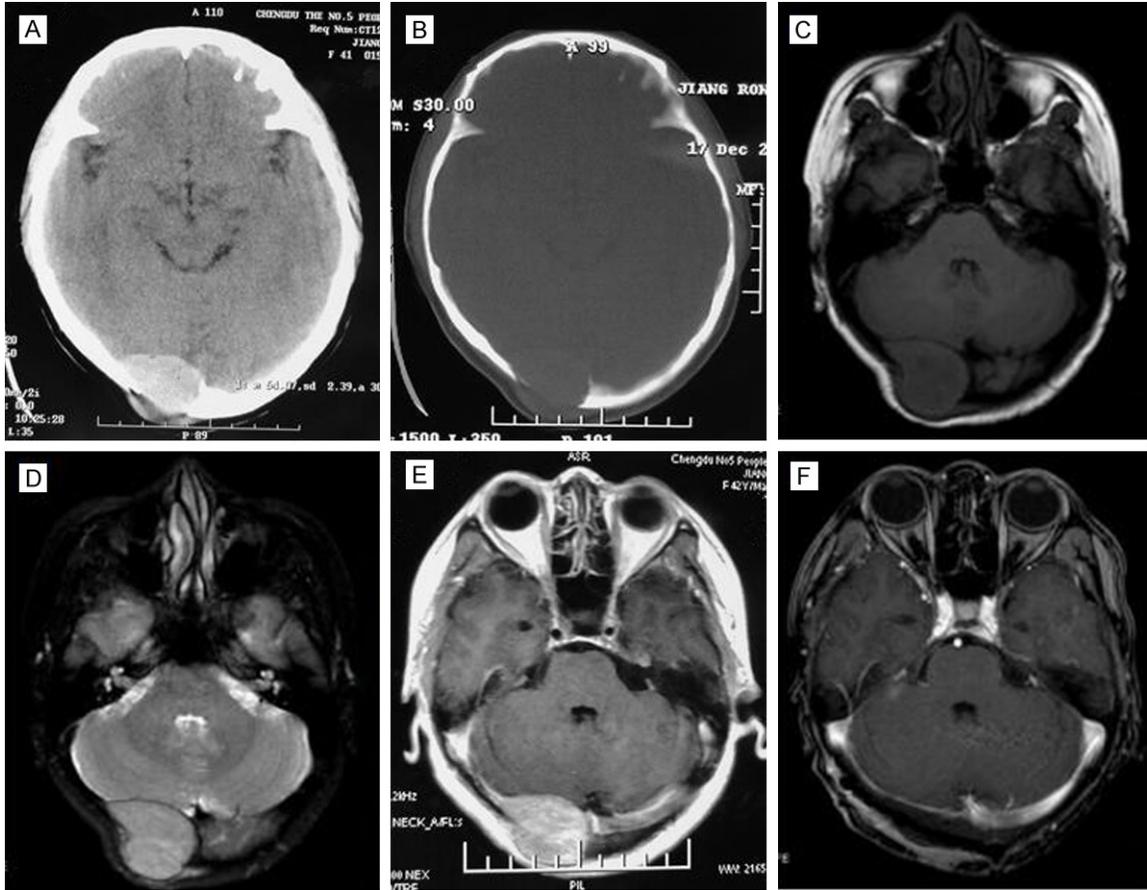
### Case presentation

A 42-year-old female first noted a soft mass which measured 4.5 × 3 cm, in the occipital region in Dec 2012. Neurological examination revealed nothing. No lesions were detected in other parts of the body. Computed tomography (CT) showed a large extradural mass, and bone CT revealed a solitary osteolytic lesion involving the whole layer of the skull on Dec 17, 2012 (**Figure 1A, 1B**). Magnetic resonance imaging

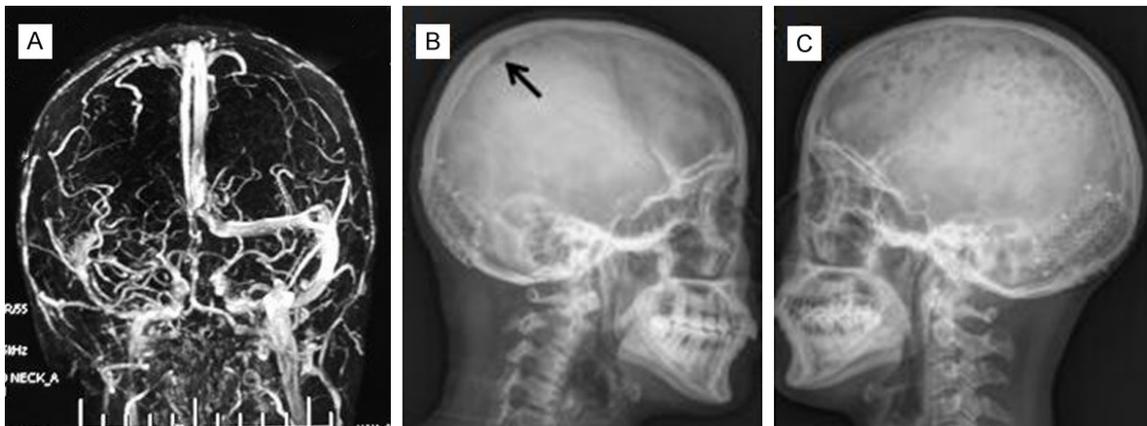
(MRI) scan revealed that the occupying lesion in the right occipital region across the transverse sinus was mostly isointense with the brain parenchyma on both T1- and T2-weighted images and was homogeneously enhanced (**Figure 1C, 1E**). Magnetic Resonance venography (MRV) displayed the right transverse sinus was pressed to full-closed position (**Figure 2A**). Preoperative hematological parameters were within the normal range.

The patient underwent a craniectomy on Dec 27, 2012. The tumor extended to the subcutaneous and the space between the dura mater and skull through skull defects. The tumor was a completely epidural mass without transverse sinus involvement. It had a rich blood supply and was easily separated from the skull. Expanded range of bone window was designed to control intraoperative bleeding and ensure completely resection (**Figure 1F**). The tumor was 5 × 6 × 3 cm in size. Pathological diagnosis of the tumor was plasmacytoma (**Figure 3**). Immunohistochemistry showed CD138(+), Igκ(+) and Ki-67 (about 60%). Bone marrow aspiration revealed MM on Jan 17, 2013. X-ray revealed several small lesions in the parietal bone and there was no exception in other skeletal sys-

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**Figure 1.** Mass destruction and skull defect were shown in the right occipital bone by computed tomography (A, B). Axial Magnetic resonance images showing the lesion isointense with the brain parenchyma on both T1- and T2-weighted images and homogeneously enhanced (C-E). Complete resection was achieved after surgery on contrast-enhanced MRI (F).

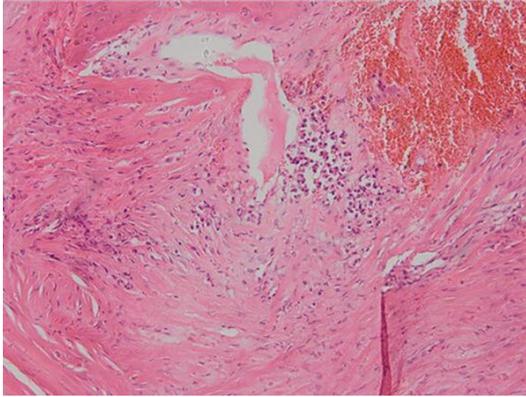


**Figure 2.** Magnetic Resonance venography displayed the right transverse sinus was pressed to full-closed position (A). X-ray revealed several small lesions in the parietal bone on Jan 17, 2013 (B, black arrow) and on June 4, 2014 (C).

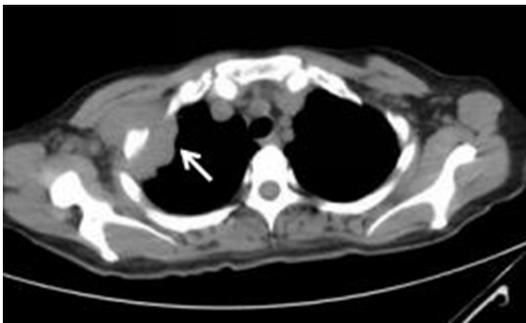
tems (**Figure 2B**). Repeated laboratory examinations showed HGB, 86 g/L; globulin, 55.3

g/L. Serum protein electrophoresis (SPE) showed M-protein levels > 36.9%.

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**Figure 3.** Plasma cells which have dispersed chromatin, high nuclear and cytoplasmic ratio and prominent nucleoli form the infiltration ( $\times 20$ , H&E).



**Figure 4.** Chest CT scan showed solid tissue infiltrating both the ribs and the pleura (white arrow).

The patient refused the treatment of bortezomib and autologous hematopoietic stem cell transplantation (ASCT), so the chemotherapy planning of VPDT (Vindesine +Pharmorubicin +Dexamethasone +Thalidomide) was given to the patient seven times. However, the patient began to have a chest pain on May 24, 2014. Systemic evaluation included HGB, 62 g/L; globulin, 92.9 g/L; Cr, 166  $\mu\text{mol/L}$ ; UA, 864  $\mu\text{mol/L}$ ; Ca, 4.11 mmol/L; M-protein, 57.2%;  $\beta$ -2 microglobulin, 8.53 mg/L on June 4, 2014. Chest CT revealed the right second rib destruction association with tumor formation (**Figure 4**) and X-ray showed multiple lesions on skeletal survey (**Figure 2C**). The patient received the treatment of bortezomib, but she had a poor prognosis and died on July 20, 2014 because of the rapid progression of the disease.

### Discussion

MM is a malignant plasma cell disease of B lymphocytes with poor outcome. The prognosis

for solitary plasmacytoma of the skull appears to be good when it is diagnosed on strict criteria. So, making the appropriate diagnosis of solitary plasmacytoma is critical, which contains biopsy proven plasmacytoma, no evidence of systemic disease on skeletal survey, and fewer than 10% plasma cells on bone marrow biopsy [2]. It remains controversial that whether solitary plasmacytoma is independent existence. After all, some learners hold that solitary plasmacytoma is the early stage of MM [3]. They think that as long as enough follow-up time is given, solitary plasmacytoma will develop into MM [4]. In our case, solitary plasmacytoma may be an initial manifestation of MM. Indeed, preoperative imagings and physical examination revealed isolated mass, no more evidence can be used to support MM. However, the disease progressed rapidly after the accurate diagnosis of MM by bone marrow aspiration. Therefore, we also maintain that there are differences between solitary plasmacytoma and MM. Additionally, the clinical characteristics and outcomes are distinct different between the initial diagnosis MM and the secondary MM resulting from solitary plasmacytoma.

Once plasmacytoma is proved by the biopsy, bone marrow aspiration is extremely necessary to determine MM. Moreover, it is critical for the patient to receive a full systemic work-up to evaluate if the patient has MM, which includes serum protein electrophoresis, serum immunoglobulins, blood count, urine Bence Jones protein and kidney function [5]. Only blood count and biochemical markers are routinely checked preoperatively in our institution, considering the diversity of the skull mass and the lower incidence of skull solitary plasmacytoma, it is very difficult to make correct diagnosis before surgery. Actually, intraoperative hemorrhage in our case was not much ( $< 150$  ML) though the tumor had rich blood supply. However, the parameter of HGB decreased obviously from normal to 86 g/L, association with the index of globulin (55.3 g/L), this may indicate that the rapid progression of MM with postoperative obvious clinical presentations compared to preoperative asymptomatic solitary lesion. End-stage disease with severe anemia, poor renal function and hypercalcemia meant a poor prognosis according to systemic evaluation.

Compared to CT scans, X-ray has its own advantages to detect multiple lesions on skeletal sur-

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vey. Preoperative CT did not show any more skull lesions except the isolated occipital deficit. While, X-ray revealed several small lesions in the parietal bone after the diagnosis of MM. Of course, we could not rule out the rapid progression of disease involving the other parts of the skull within one month interval (preoperative CT on Dec 17, 2012; postoperative X-ray on Jan 17, 2013). Besides, the presentations of the occupying lesion showing mostly isointense with the brain parenchyma on both T1- and T2-weighted MRI scans have a unique imaging characteristics unlike other skull lesions [5]. Plasmacytoma should be considered if an isolated skull lesion displays isointense with the brain parenchyma on both T1- and T2-weighted MRI scans. Although the patient received seven times of chemotherapeutic treatment and showed stable before May 24, 2014, she suffered from treatment failure ultimately. Clinical benefit of the proteasome inhibitor bortezomib for MM patients remains unchallenged [6]. Although she received the treatment of bortezomib after failure, the prognosis was still poor. The patient may have a better outcome if she received bortezomib at earlier stage.

### Disclosure of conflict of interest

None.

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### References

- [1] Gill MK, Makkar M, Bains SP. Solitary plasmacytoma of skull: a rare cytological diagnosis. *J Clin Diagn Res* 2013; 7: 1702.
- [2] Warsame R, Gertz MA, Lacy MQ, Kyle A, Buadi F, Dingli D, Philip RG, Suzanne RH, Shaji KK, John AL, Stephen JR, Thomas EW, Joseph M, Nelson L, Steven RZ, Vincent R, Angela D. Trends and outcomes of modern staging of solitary plasmacytoma of bone. *Am J Hematol* 2012; 87: 647-651.
- [3] Shah BK, Saifuddin A, Price GJ. Magnetic resonance imaging of spinal plasmacytoma. *Clin Radiol* 2000; 55: 439-445.
- [4] Hawkesford MP, Bowey AJ, Rao J, Meara NJ. Synchronous presentation of Gaucher disease and solitary plasmacytoma with progression to multiple myeloma. *Scott Med J* 2011; 56: 236.
- [5] Dong L, Zhang X, Zhang H, Song R, Gu X, He L. Solitary plasmacytoma of the skull: two case reports. *Oncol Lett* 2013; 5: 479-482.
- [6] Malek E, Abdel-Malek MA, Jagannathan S, Vad N, Karns R, Jegga AG, Broyl A, Van Duin M, Sonneveld P, Cottini F, Anderson KC, Driscoll JJ. Pharmacogenomics and chemical library screens reveal a novel SCFSKP2 inhibitor that overcomes Bortezomib resistance in multiple myeloma. *Leukemia* 2017; 31: 645-653.