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Giant cranial plasmacytoma: case report and discussion of a potential relationship with sex hormones

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To the Editors

Cranial solitary plasmacytoma is a very rare clinical manifestation of multiple myeloma (MM) [1–3]. Moreover, cranial plasmacytomas presenting as visibly prominent swellings on the head are extremely rare, and their management poses a challenge for neurosurgeons. It remains unclear which patients are most likely to develop this type of large, solitary, protruding cranial plasmacytoma. Age and gender are the two main non-modifiable factors in an individual's risk of being diagnosed with cancer. [4] Various biological and behavioural factors have been proposed to explain how age and gender significantly modulate the incidence of cancer at the population level. Exploring common features of such cranial plasmacytomas may shed light on their pathophysiology.

A 90-year-old female patient was admitted to our hospital complaining of a soft, painless, rapidly swelling mass that had developed within a month on the left frontal region of her scalp. The skin overlying the lesion was hyperaemic because of enlarged vessels. The patient did not have any complaints about the bump, such as a headache, other than its cosmetic appearance, and no neurological deficit was detected during the examination. Her medical history revealed that not only had she been diagnosed with MM eight years before (which was treated with chemotherapy), but also that she had been diagnosed with basal cell carcinoma in the sacral region four years before presenting on this occasion. The patient was in remission, but had disrupted her treatment for the preceding two years because of the COVID-19 pandemic. The patient's blood and urine analyses were consistent with the diagnosis of MM, including increased monoclonal immunoglobulins, anaemia, and hypercalcaemia.

Cranial magnetic resonance imaging revealed a giant left frontal extra-axial lesion adjacent to the superior sagittal sinus (Fig. 1a). The lesion had compressed the cerebral tissues, causing a left ventricular collapse and subfalcine herniation. The lesion was isointense and showed homogeneous contrast enhancement on the T1-weighted sequences. Dural boundaries were indistinguishable. Magnetic resonance angiography showed that the mass was being fed from the distal branches of the external carotid artery (Fig. 1b). It was observed that the tumour had destroyed both the inner and outer tables of the skull in an area approximately 3 cm in diameter.

The patient was operated on with a bifrontal incision under general anaesthesia (Fig. 1c). There was a cleavage line between the skin and the tumour. The galea was taut but intact. Although the tumour was vascular in nature, no excessive bleeding was observed. The tumour had completely eroded the

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calvarial bone in a small area. The parts of the tumour extending outward from the bone surface were completely removed up to the bone surface. The grey-brown and slightly hard intracranial component of the tumour was also completely resected using a cleavage line plane between the tumour and the brain (Fig. 1d). The dura was closed using a galeal graft. In this elderly patient, cranioplasty was not performed, not only because of the small calvarial defect, but also to shorten the operation time and prevent the risk of infection. The patient woke from anaesthesia with a Glasgow Coma Scale (GCS) score of 15 without any problems and was extubated in the operating room. She was then taken as routine to the intensive care unit and transferred to a ward bed a day later. Diffuse plasma cell infiltration was observed histologically; the results were positive for kappa light chain and negative for lambda light chain (Fig. 1e). Plasma cells, which were kappa monoclonal, displayed strong and diffuse expressions of CD138, CD38 and CD56.

The patient did well postoperatively. She was transferred to the haematology ward on the seventh postoperative day, and after three weeks of chemotherapy, she was discharged. A postoperative MRI scan showed total removal of the tumour (Fig. 1f). Four months later, the patient developed sudden mild hemiparesis due to a cerebrovascular ischaemic event, but recovered after treatment.

In this article, we present a case of solitary cranial plasmacytoma presenting as a giant swelling on the head of a 90-year-old female patient with a history of sacral basal cell carcinoma and an eight-year history of MM in remission. Although MM is more common in men, cranial plasmacytomas tend to be more common in women. Similarly, basal cell carcinomas are more common in women under the age of 40 than in men [4]. To the best of our knowledge, the potential relationship between sex hormones and cranial plasmacytomas has not been explicitly researched.

Osteoclasts are important cells involved in the bone lysis of MM. Osteoclast formation during bone remodelling is affected by a protein from the tumour necrosis factor family called the receptor activator of nuclear factor-κB ligand (RANKL) [5]. RANKL increases osteoclast activation, which results in augmented bone resorption, leading to the release of IGF1 and TGF-β. This process then leads to immune suppression and the further proliferation of MM cells. An increase in serous RANKL has been found to increase the risk of sex hormone-induced breast cancer associated with a familial BRCA1 mutation. Females with BRCA1 mutations have an increased risk of developing breast and ovarian cancers. Although no laboratory examination for either RANKL or BRCA1 mutations could be performed on our patient, increased RANKL activation and/or a BRCA1 mutation may underlie the effects of aggressive bone lysis in females with giant, rapidly growing, bone-destroying plasmacytomas. With the excessive shift of the osteoclastic-osteoblastic balance in favour of osteoclastic activity, rapid destruction of the bone and rapid growth of the

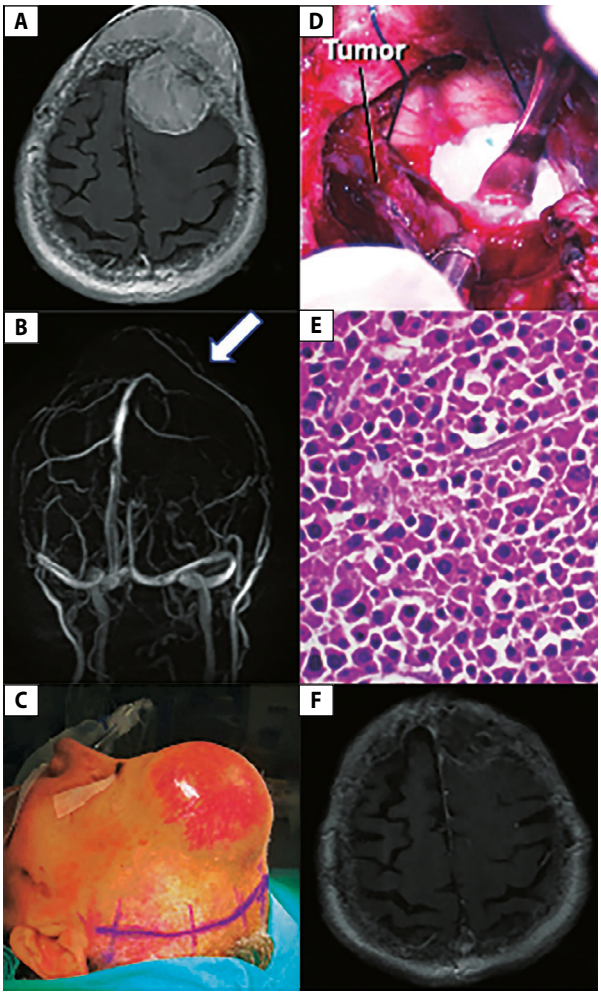

Figure 1. A. Cranial magnetic resonance image showing giant left frontal extra-axial lesion adjacent to superior sagittal sinus; **B.** Magnetic resonance angiographic scan showing that mass was being fed from distal branches of external carotid artery (arrow); **C.** Patient was operated on with a bifrontal incision under general anaesthesia. Skin overlying the lesion was hyperaemic because of enlarged vessels; **D.** Grey-brown and slightly hard intracranial component of tumour was also completely resected using a cleavage line plane between tumour and brain; **E.** Diffuse plasma cell infiltration was observed histologically; **F.** Postoperative MRI scan showing total removal of tumour

plasmacytoma from the bone surface towards a weak barrier, such as the skin, may be possible. Under the influence of sex hormones, excessive osteoclastic activity may cause the appearance of giant cranial solitary plasmacytomas in women more than in men. Another observation that supports this hypothesis is that Paget's disease, which is known to be more common in women, can be seen frequently with MM.

Although preoperative superselective tumour embolisation was recommended to prevent excessive blood loss during the operation, embolisation was not attempted in this case in order not to cause additional morbidity for the patient with advanced age and comorbidities. There was no uncontrollable

bleeding in the surgery. The primary surgical goal should be radical tumour resection, but radiotherapy may be preferred for small tumours or when resection is not possible because the tumour is extremely radiosensitive. Since the tumour was completely resected in the presented case, radiotherapy was planned to be considered only in case of recurrence.

Since cranial plasmacytomas can present in many different forms, many pathological lesions, such as glioblastomas, subdural haematomas, metastases, meningiomas, sarcomas, and giant cell tumours, should be considered during differential diagnosis [1, 2].

This case demonstrates the complexity of the pathophysiology of giant cranial solitary plasmacytomas. A potential relationship between sex hormones and giant cranial plasmacytoma may exist. Notably, this case featured the oldest patient to have been successfully surgically treated and documented in the medical literature.

Ethical issues: *The ethical issues for this study have been carefully considered in line with the Declaration of Helsinki and its amendments. Written informed consent and permission to reproduce copyrighted materials were obtained from the patient.*

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