Received: 18.11.2009 / **Accepted:** 03.08.2010
DOI: 10.5137/1019-5149.JTN.2788-09.2

Plasmocytoma of the Skull Vault

Kranial Kemiklerin Plazmositomu

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ABSTRACT

AIM: Plasma cell tumours include solitary plasmocytoma, multiple plasmocytoma and multiple myeloma. Herein we report a case of plasmocytoma of the skull.

CASE HISTORY: A 49 year-old-man suffered from amnesia, irritability and a rubbery swelling which was gradually increased approximately to 4-5 cm in diameter at the frontal vertex over a year. His neurological examination was normal. The skull direct X-ray revealed patchy destruction of the left frontal bone 10 cm in diameter. CT with bone windows showed relatively preserved but severely thinned inner table and irregularly destructed outer table. MR scan showed a hypervascular, mainly solid mass measuring 9x5.5x8.5 cms. Radical surgery consisting of total tumour extirpation was undertaken. Histopathological examination confirmed a plasmocytic proliferation. The postoperative period was uneventful and the patient was discharged on the 6th day postoperatively. Bone marrow sampling was scheduled to rule out multiple myeloma, but the patient committed suicide a week later for reasons not known to us.

CONCLUSION: Although the prognosis of a plasmocytoma is relatively good, this study demonstrated that patients with ventral frontal cortex tumour may have significantly worse mood pre- or postoperatively. Clinicians should therefore be careful in the follow-up period.

KEYWORDS: Frontal, Plasmacytoma, Skull vault

ÖZ

AMAÇ: Plazma hücreli tümörler başlığı altında soliter plazmasitoma, multiple plazmasitoma ve multiple miyeloma yer alır. Bu yazıda frontal kranial kemik yerleşimli plazmasitomu olan bir hasta rapor edilmiştir.

YÖNTEM ve GEREÇ: 49 yaşındaki erkek hasta amnezi, sinirlilik ve frontal bölge cildinde sert kıvamlı ve 1 yıl içinde 4-5 cm çapa ulaşan şişlik şikayeti ile başvurdu. Nörolojik muayene bulguları normal saptanan hastaya direkt kafa grafisi çekildi ve frontalde yaklaşık 10 cm çapında kemik defekti saptanarak kranial MRG yapıldı. MRG de frontal bölgede hipervasküler görünümlü, solid-kistik komponenti olan ve 9x5, 5x8,5 cm çapında ekstradural yerleşimli kitle tespit edilerek gros total kitle eksizyonu ameliyatı yapıldı. Operasyon sırasında tümörün kemik iliğinden kaynaklandığı ve eksternal ve internal kemik tabulaları arasında yer alıp kemik yapıların oldukça incelmesine neden olduğu gözlendi. Histopatolojik inceleme sonucu plazmositik proliferasyon olarak rapor edildi. Ameliyat sonrası takibinde problem olmayan hasta altıncı günde taburcu edildi. İzleminde multiple miyelomaya yönelik kemik iliği örneklemesi planlanan hastanın taburculuğundan bir hafta sonra intihar ettiği öğrenildi.

SONUÇ: Her ne kadar plazmositomanın prognozu relatif olarak iyi olsa da, özellikle ventral frontal yerleşimli tümörlerin ameliyat öncesi ve/veya sonrası ciddi davranışsal problemlerin oluşmasına zemin hazırlayabileceği ve bu nedenle de bu tarz hastaların ameliyat sonrası dikkatli takibinin gerekliliği akılda tutulmalıdır.

ANAHTAR SÖZCÜKLER: Frontal, Kafatası, Plazmositoma

INTRODUCTION

Plasma cell tumours include solitary plasmocytoma, multiple plasmocytoma and multiple myeloma. Plasmocytomas may associate with or progress to multiple myeloma in 50% of patients (2). Solitary extramedullary plasmocytoma is defined as an extraosseous proliferation of neoplastic plasma cells without bone marrow or systemic involvement and they usually occur in the upper aerodigestive tract (1). Solitary bone plasmocytoma appears in about 5% of patients with plasmocytoma and most frequently localizes in the vertebral column. Primary craniocerebral plasmocytoma is very uncommon and may be either plasmocytoma of the skull (osseous form) or dural plasmocytoma (non-osseous form) (3). Herein we report a case of plasmocytoma of the skull.

CASE REPORT

A 49 year-old-man suffered from amnesia, irritability and a rubbery swelling which was gradually increased to approximately 4-5 cm in diameter at the frontal vertex over a year. His neurological examination was normal. The skull direct X-ray revealed patchy destruction of the left frontal bone 10 cm in diameter. Computed tomography (CT) showed a large extradural mass with homogeneous enhancement after intravenous contrast administration. CT with bone windows revealed a solitary osteolytic lesion changing the external as well as internal tables into a thin layer (Figure 1A,B). Magnetic resonance (MR) scans revealed a hypervascular, mainly solid mass measuring 9x5.5x8.5 cm. It enhanced diffusely at the early arterial phases and it did not

have a dura tail. MR-angiography images showed a massive vascular blush fed by vessels originating from the right and left superficial temporal arteries; and it revealed that the anterior part of the superior sagittal sinus was occluded by the tumour. However, other dural sinuses and deep cerebral veins were patent. The MR scan also disclosed two other tumoural masses: the first a lobulated and hypervascular tumour located in the nasopharyngeal fossa; and the second an epidermoid tumour located in the prepontine cistern (Figure 2A-D). The feeding arteries, especially those coming from the left superficial temporal artery, were embolised by an interventional radiologist before extensive radical surgery

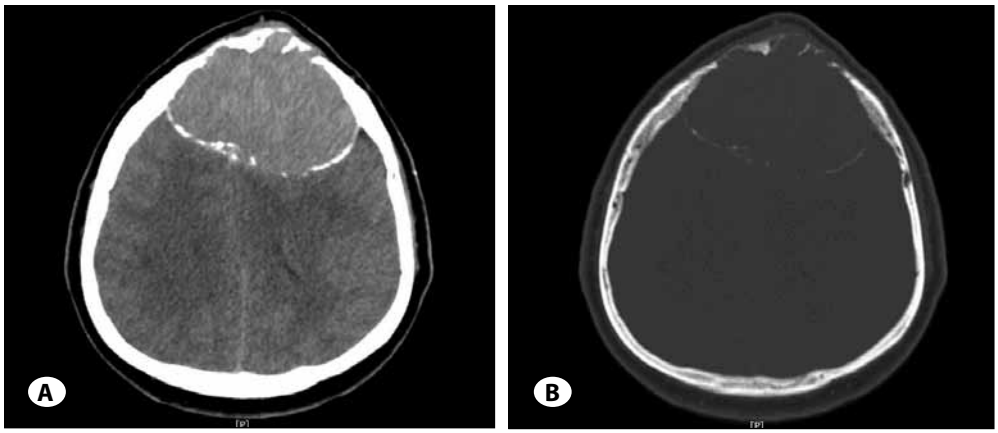

Figure 1: A) Computed tomography (CT) showed a large hyperdense extradural mass with homogeneous enhancement after intravenous contrast administration.
B) CT with bone windows revealed a solitary osteolytic lesion changing the external as well as internal tables into a thin layer.

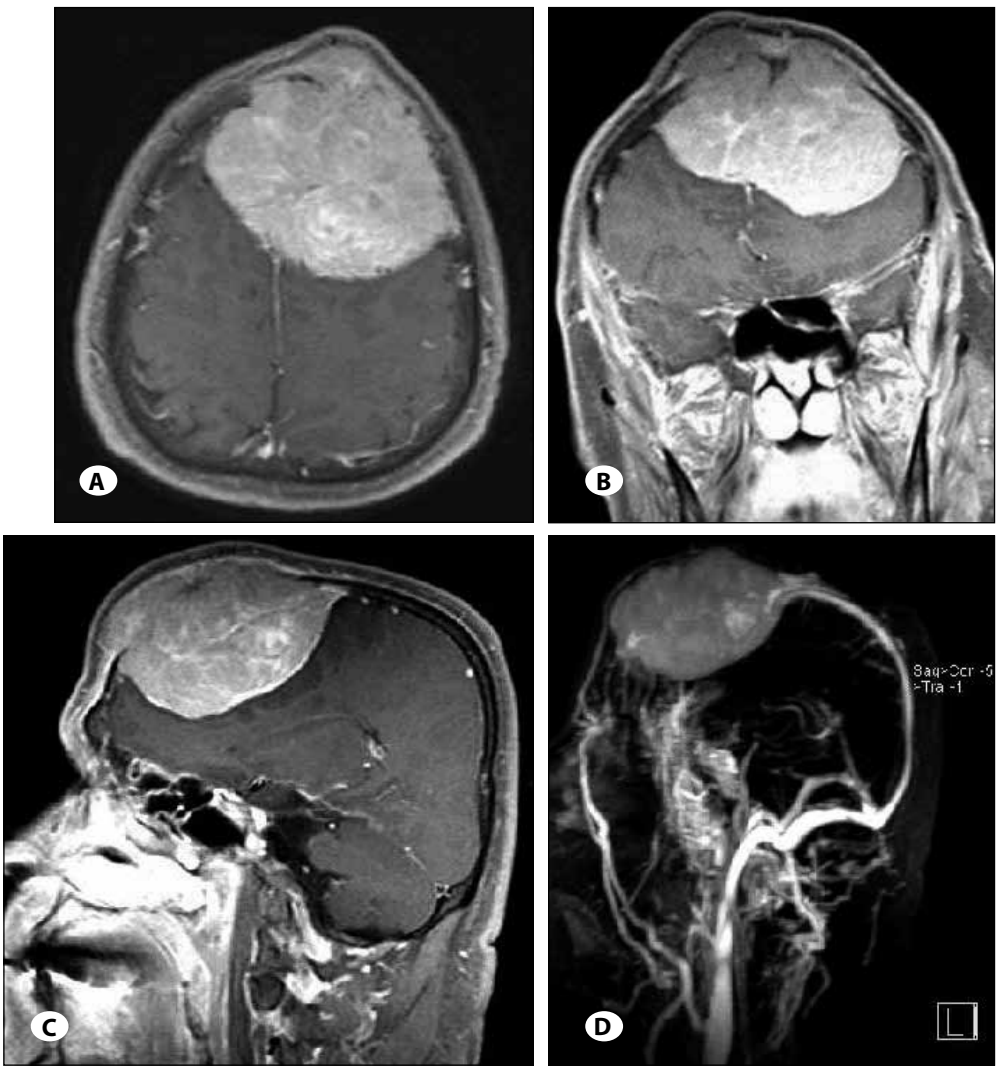

Figure 2: A-C) MR images with gadolinium show a hypervascular, mainly solid mass which destructed the frontal and left parietal bones and extended to the subgaleal area of the left frontal region; and
D) enhanced diffusely at the early arterial phases of MR angiography.

could be undertaken. At surgery, it was seen that the mass originated from bone marrow and it had changed the external and internal tables into a thin layer. The mass was delicately removed with the bone lamellas which formed the internal tabula over the dura mater; and the resulting bone defect was repaired with a graft taken from the ipsilateral temporal bone (Figure 3A,B). Histological examination confirmed a plasmocytic proliferation with CD-38 and kappa-chain protein positive cells (Figure 4). Immunoelectrophoresis of serum, urine analysis for Bence-Jones protein, lambda and kappa light chain proteins, serum level of beta-2 microglobulin, all immunoglobulins, and creatinine, calcium, blood count were found to be normal. However, the urine beta-2 microglobulin was slightly increased. The postoperative period was uneventful and the patient was discharged on the 6th day postoperatively.

To distinguish multiple myeloma, bone marrow sampling was scheduled during the early follow-up period but he jumped off the fifth floor a week later and this suicide attempt ended with the patient's death. A postmortem examination was not possible.

DISCUSSION

Multiple myeloma (MM) is a disseminated malignancy of plasma cells and its diagnosis is based on (a) histological evidence of plasmocytoma or plasmocytosis of bone marrow, (b) clinical evidence of disease such as anemia or renal failure, (c) serum or urine monoclonal gammopathy or osteolytic lesions on X-rays. Solitary plasmocytomas are solitary lesions without clinical, histologic, or radiological evidence of MM. True solitary plasmocytoma of the skull vault is very rare. Plasmocytoma involving the skull base originating from the nasopharynx, oropharynx, or paranasal sinuses is defined as "extramedullary plasmocytoma". These secondary lesions carry a high risk of progression to multiple myeloma (7). Diagnostic criteria of a solitary plasmocytoma include a radiologically solitary lesion, histopathological confirmation, negative bone marrow examination, negative immunoelectrophoresis, negative urine test for Bence Jones protein, no evidence of hypergammaglobulinemia, and absence of anemia (6, 9). Our case meets all these criteria except additional nasopharyngeal lesion and absent bone marrow examination.

Plasmocytomas generally appear as a bone or soft tissue tumour with mass effect, pain, weakness, fatigue, fever, infection and infiltrative behaviour. In patients with primary brain tumours, the lesion itself generates direct focal cerebral dysfunction depending on tumour location, and may therefore cause somatic symptoms such as fatigue, sleep disturbances, anorexia, etc. Furthermore patients with a tumour located anteriorly (frontal region) in a hemisphere may have emotional distress and depressive symptoms such as sadness and lack of motivation. However, these symptoms may decrease after the surgery. Irle et al (1994) underlined the fact that patients with ventral frontal cortex tumour have significantly worse mood postoperatively than those with

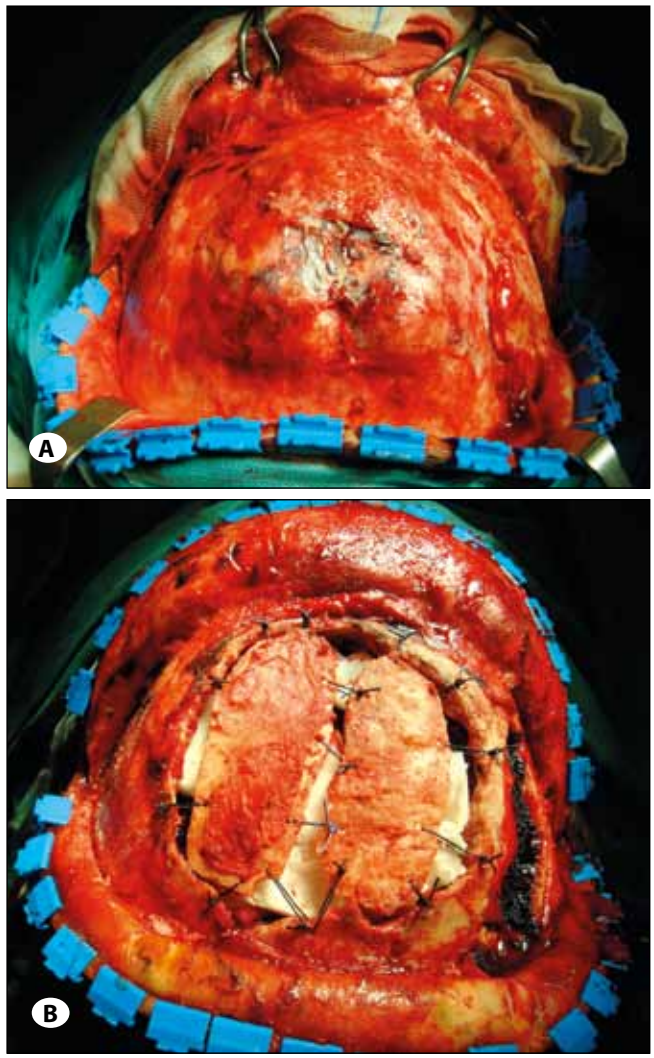

Figure 3: A) mass originating from bone marrow has changed the tabula externa into a thin layer. **B)** and the resulting bone defect was repaired with a graft taken from the ipsilateral temporal bone.

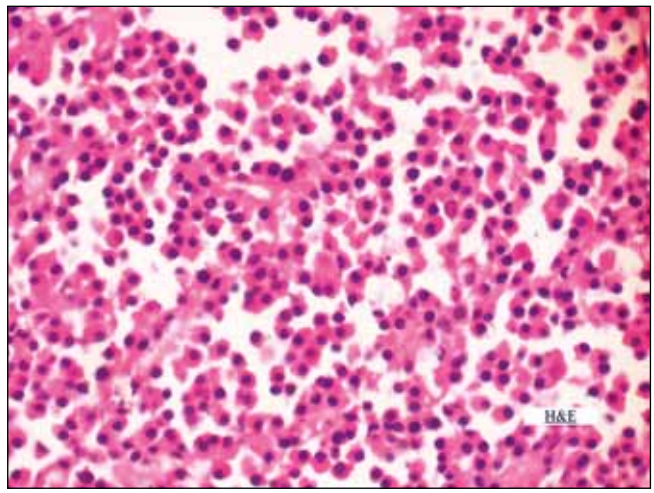

Figure 4: Histopathological consideration is solitary plasmacytoma (H&E, x200).

lesion in other regions of brain (4). The occurrence of these symptoms most probably originates from frontal and/or limbic release or disinhibition (5). On the other hand Pringle et al (1999) showed that patients with a meningioma had higher levels of anxiety and depression than those with any other tumour types (8). Although the mass was located extra-axially in our patient, this frontal lesion may have caused amnesia, and irritability by its mass effect preoperatively; and furthermore in the postoperative period these symptoms might have been aggravated by frontal and/ or limbic release or by increased frontal blood flow due to metabolic processes in this area; and finally he had worse mood than before which could have caused his suicide. Nevertheless, the exact reason will remain unknown.

Plasmocytoma may be confused with meningioma, metastasis, sarcoma, chordoma, and giant cell tumour. Meningioma may cause a similar high density peripheral lesion with homogenous enhancement and they have a "dura tail". Furthermore, except for rare intraosseous forms, meningiomas rarely cause lytic bony changes and their blood supply usually comes from scalp arteries. However, the blood supply of a plasmocytoma derives from the external carotid artery (superficial temporal artery, external occipital artery, and middle meningeal artery) and amuscular branch of the vertebral artery (3). In our patient, the lesion enhanced diffusely at the early arterial phases and it did not have dura tail.

The treatment regimen for a solitary plasmocytoma is total tumour extirpation and radiosurgery. Survival without recurrence or a systemic involvement of the disease after total tumour removal has been reported. Some authors state that these tumours usually disseminate and that the prognosis is very poor. Radiosurgery has been suggested if there is a residual mass after surgery (6, 9).

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