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Case Report Open Access

Orbital Plasmablastic Plasmacytoma in Multiple Myeloma

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Abstract

Multiple myeloma is a malignant disease characterized by the proliferation of clonal plasma cells in bone marrow, extramedullary involvement with multiple myeloma is generally a manifestation of advanced disease. Orbital involvement by multiple myeloma is uncommon and intraocular involvement is rare. The authors present an uncommon case of orbital plasmablastic plasmacytoma.

Keywords: Orbital plasmacytoma; Plasmacytoma; Orbital plasmablastic plasmacytoma; Multiple myeloma

Introduction

Multiple myeloma is a malignant disease characterized by the proliferation of clonal plasma cells in bone marrow [1], extramedullary involvement with multiple myeloma is generally a manifestation of advanced disease [2]. Orbital involvement by multiple myeloma is uncommon and intraocular involvement is rare [3,4].

Case Report

A 73-year-old man was referred to the department of ophthalmology for swelling and ptosis of left eyelid and right lateral and left superior orbital wall masses by brain MRI. He first visited the department of neurology for headache and paresthesia of the left chin and orbit areas. The neurologist recommended to undergo brain MRI for evaluation purposes, and subsequently, multiple variably sized osteolytic lesions were observed in skull, a T1 and T2 iso signal intensity mass in clivus, expansile masses in the right lateral and left superior orbital walls involving adjacent soft tissue, and T1 low signal intensity of C2 vertebra (Figure 1). The radiologist propounded the diagnostic possibilities of multiple myeloma and bone metastasis. The patient was referred to the department of hemato-oncology, where he underwent a systemic evaluation for multiple myeloma.

Visual acuity of right and left eyes were 0.9 and 0.8, respectively. Intraocular pressure of the right eye was 11mmHg and of the left was 15mmHg by NCT. Through an eye examination, mild swelling with injection and dermatochalasis of both upper eyelids and ptosis of the left eye (Figure 2) were reported. On examination of extraocular muscle movement, no limitation of extraocular muscle movement or diplopia was observed. His Hertel exophthalmometry reading was 14 mm OD, 13 mm OS and baseline was 115 mm. MRD1 was 5 mm OD and 3 mm OS. LFT of right and left eyes were 15 and 10 mm, respectively. Subsequent, orbital CT revealed an enhancing bone destructive mass in the left superior orbital wall (Figure 3), extending into orbit and upper eyelid, and another enhancing mass in sphenoid sinus and sphenoid bone involving both vidian canals. Based on orbital CT and MRI, the diagnostic possibilities of multiple myeloma and bone metastasis were considered. Orbitotomy and excisional biopsy of the left superior orbital mass was performed and a multiseptated 2.5 × 1.0 cm mass was excised (Figure 4). Post-operative orbital CT (Figure 5) confirmed removal of the left upper eyelid portion of the mass, but a residual enhancing bony destructive mass was detected in the left orbital superior wall bulging into the extraconal area of the orbit. Histopathologic evaluation confirmed the mass as plasmablastic plasmacytoma (Figure 6), and immunohistochemical staining showed positivity in for MUM1 and a 60-70% Ki67 labeling index and negativity in for CD3, CD20, PAX5, CD138, CD30, Bcl2, and ALK. Immunostain of Bcl2 showed weak-positive in about 20~40% of cells. Immunohitochemical staining of CD79a or CD19 was not performed. Tumor cells were immunopositive for lambda light chain and negative for kappa light chain.

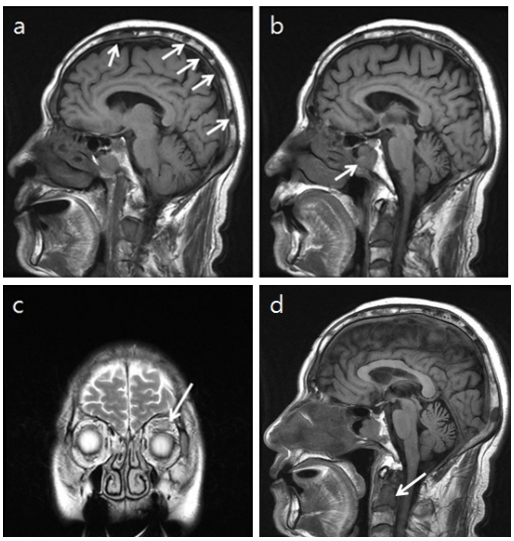

Figure 1: Brain MR images **(a)** Multiple variable sized osteolytic lesions in skull. **(b)** T1 iso signal intensity mass in clivus. **(c)** Left superior orbital wall mass involving adjacent soft tissue. **(d)** T1 low signal intensity of C2 vertebra.

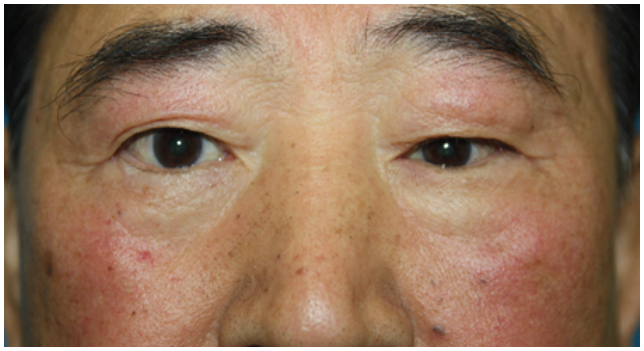

Figure 2: Eye examination showed mild swelling with injection and dermatochalasis of both upper eyelid, ptosis and hypogobus of the left eye.

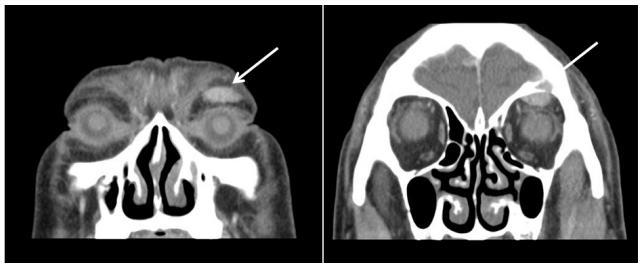

Figure 3: Pre OP orbit CT showing a well enhancing bone destructive mass in the left superior orbital wall extending into orbit and upper eyelid.

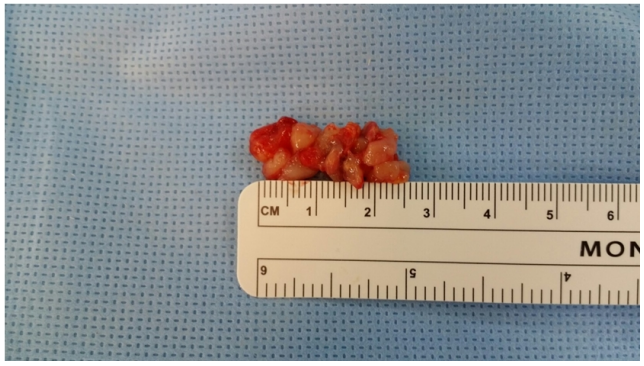

Figure 4: A multisepted 2.5 × 1.0 cm mass was excised from the left superior orbit.

Laboratory evaluation showed serum free lambda light chain (233.58 mg/L) and IgD (205.28 mg/dl) were elevated. However, other laboratory results were within the normal ranges. No remarkable findings were observed in serum protein EP or urine protein EP. Bone marrow aspiration biopsy revealed a normal distribution of hematopoietic cells and a few atypical plasma cells. The CD138 immunohistochemical stain of bone marrow presented sparsely scattered plasma cells. Chromosome analysis was normal.

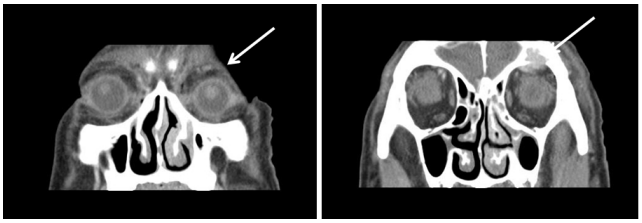

Figure 5: Postoperative orbit CT images. The left upper eyelid portion of the mass was removed, but a residual enhancing bony destructive mass was observed in the left orbital superior wall, bulging into the extraconal area of the orbit.

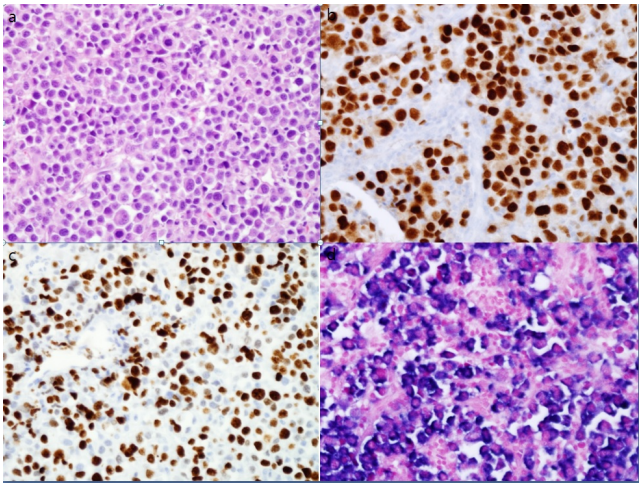

Figure 6: Histopathology **(a)** HE (400X) **(b)** Mum1(x400) **(c)** Ki67 (400X) **(d)** Lambda (in situ hybridization, 400X).

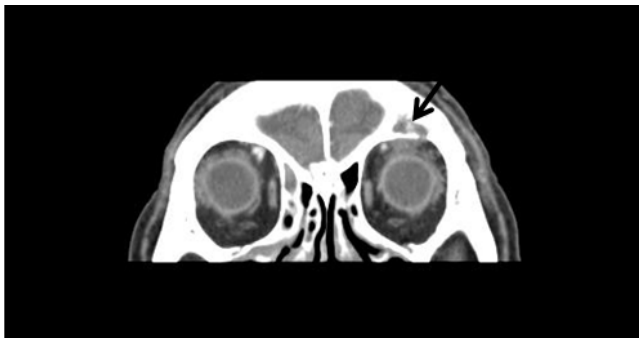

Figure 7: Orbit CT image obtained at 2 months postoperatively. The size of the enhancing bony destructive mass in left orbital superior wall (black arrow) had decreased, but a residual small, poorly enhancing, soft tissue lesion was observed in the left superior bony orbital rim and adjacent extraconal area.

The patient was diagnosed as having extramedullary plasmacytoma, and started chemotherapy (VMP: Vincreistine, Melphalan, Prednisolone) in the department of hemato-oncology and

radiotherapy for mass reduction and to treat bone destruction in the department of radio-oncology.

After first chemotherapy and radiotherapy (2 months postoperatively), orbit CT was performed (Figure 7). The size of the enhancing bony destructive mass in the left orbital superior wall was found to have decreased and a residual, small, poorly enhancing soft tissue lesion was reported in left superior bony orbital rim and adjacent extraconal area.

Discussion

According to the International Myeloma Working Group, the diagnosis of multiple myeloma should fulfill the following criteria: clonal bone marrow plasma cells ≥ 10% or biopsy-proven bony or extramedullary plasmacytoma plus any one of the following myeloma defining events: end organ damage (hypercalcemia, renal insufficiency, anemia, bone lesions), or any one or more of the following biomarkers of malignancy, a clonal bone marrow plasma cell percentage ≥ 60%, an involved:uninvolved serum free light chain ratio of ≥ 100, or >1 focal lesions by MRI [5].

In our case, extramedullary plasmacytoma was proven by biopsy and multiple bone lesions were confirmed by MRI and PET-CT (Figure 8). These findings fulfilled the International Myeloma Working Group criteria, and thus a diagnosis of multiple myeloma was made.

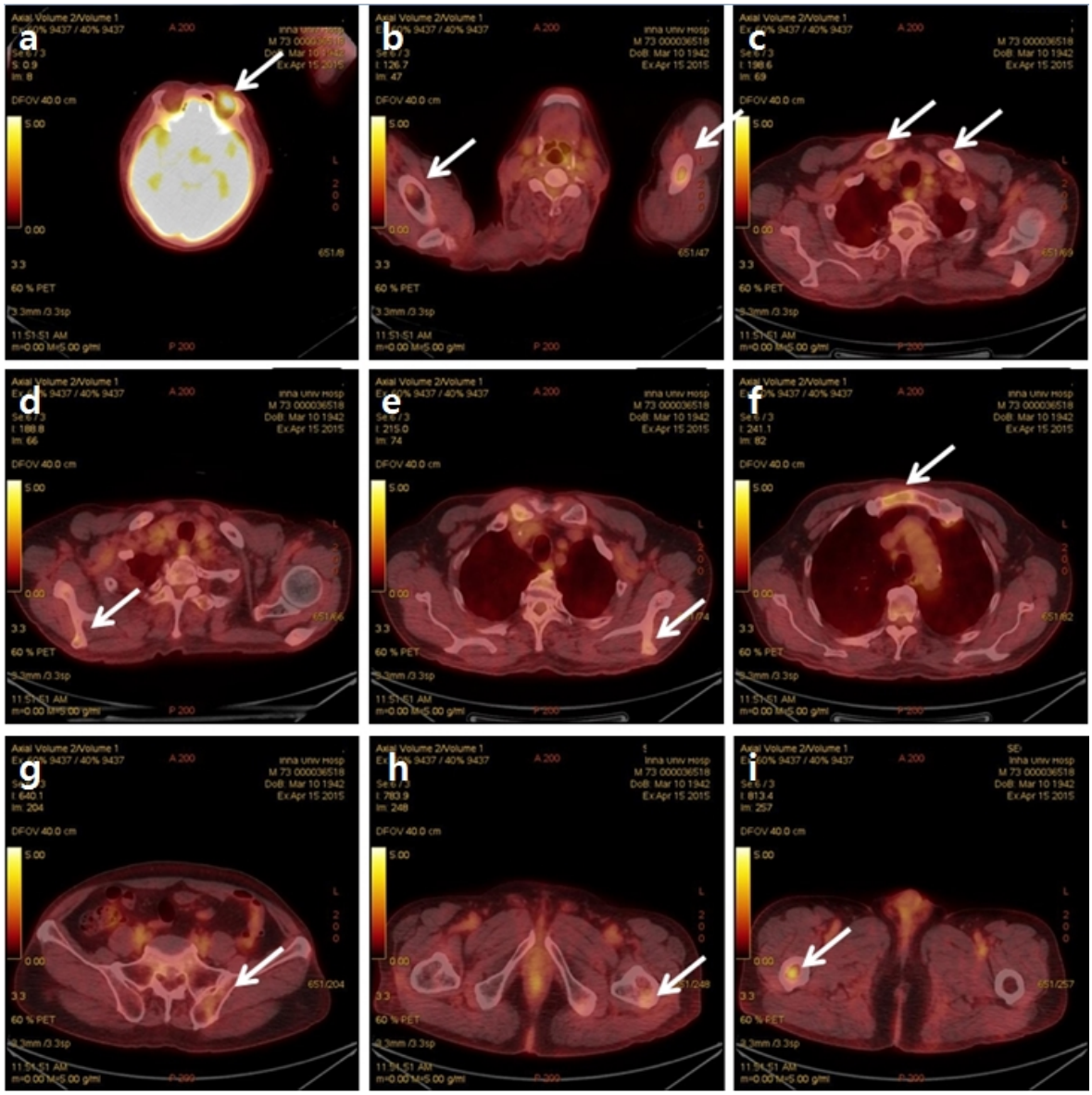

Figure 8: PET-CT images showing bone lesions on **(a)** the left orbital superior wall, **(b)** both humeri, **(c)** ribs, **(d)** and **(e)** both scapula, **(f)** sternum, **(h)** left iliac crest, and **(i)** right femur.

Orbital involvement in multiple myeloma is uncommon [3,4]. Bonavolonta et al. analyzed 2,480 spacing-occupying lesions of the orbit from 1976 to 2011 [6], and reported plasmacytoma had a prevealence of 3% patients and accounted for <1% of total orbital lesions. Because of its insidious nature and as orbital symptoms can manifest the first symptom of multiple myeloma [2-4], it is important to take multiple myeloma or plasmacytoma into consideration when an orbital spacing-occupying lesions is detected.

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