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Solitary osseous plasmacytoma of the orbit with multiple myeloma: A case report

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Abstract

Solitary osseous plasmacytoma of the orbit is very rare. It can be part of generalized disease (multiple myeloma) or as localized disease, presenting as orbital tumor. We report a case of solitary osseous plasmacytoma of the orbit with multiple myeloma. This 43-year old male presented with 6 months history of proptosis and decreased visual acuity. CT scan showed an osteolytic, well demarcated lesion with homogenous contrast enhancement in the superolateral part of the right orbit. The tumor including the osteolytic lesion was totally removed. The pathological report revealed a plasmacytoma. Further investigation showed that the plasmacytoma was part of generalized multiple myeloma. The patient was treated with radiation and combination chemotherapy postoperatively. Twenty months after surgery, the local tumor remained well controlled. However, there was systemic progression of the disease, with multiple cutaneous mass and pleural effusion. These resulted in the patient's deterioration and final demise.

INTRODUCTION

Solitary plasmacytoma of the orbit can originate either from the bone (solitary plasmacytoma of the bone) or from the orbital soft tissue (solitary extramedullary plasmacytoma).[1,2] Solitary osseous plasmacytoma of the orbit is a plasmacytoma of the skull base, which is the rarest type of solitary plasmacytoma of the bone. Orbital osseous plasmacytoma can either be part of generalized disease or confined as localized disease.[2,3]

Plasmacytoma of the skull base manifests in variety of signs and symptoms depending on the site of origin.[4,5] It can present as multiple cranial nerve palsies or orbital mass.[2-5] Plasmacytoma of the skull base is more often part of a disseminated disease, rather than a local disease.[3-6] We report a case of a solitary osseous plasmacytoma of the orbit with systemic multiple myeloma.

CASE REPORT

A 43 year old male was referred from The Department of Ophthalmology with severe proptosis and visual impairment. The patient complained of 6 months history of progressive proptosis associated with decreased visual acuity of the right eye. Physical examination showed proptosis, chemosis, limited ocular movement, swelling of the upper eyelid and opacity of the sclera right eye. The visual acuity was hand movement only. Orbital CT scan showed an osteolytic, well demarcated lesion with homogenous contrast enhancement in superolateral part of the orbit (Figure 1). Small part of the tumor had extended into intracranial space without evidence of brain infiltration. The patient was initially diagnosed as meningioma of the orbit. Frontotemporal craniotomy and tumor resection was performed. The tumor originated from orbital roof and lateral part of the orbit. It was highly vascular, soft and easily separated from the surrounding structures. The bone defect was sharply demarcated with no osteoplastic reaction seen along the margin. The tumor was completely resected both in the intraorbital and intracranial space. The involved bone was also resected with margin. Hematoxylin-eosin preparation of the surgical specimen showed diffuse plasma cells infiltration, from almost 80 % of mature to 20 % of immature morphology. The mature plasma cells were ovoid with round eccentric nuclei, clock-face chromatin pattern, and abundant cytoplasm and marked perinuclear "hof". The immature plasma cells showed dispersed nuclear chromatin, high

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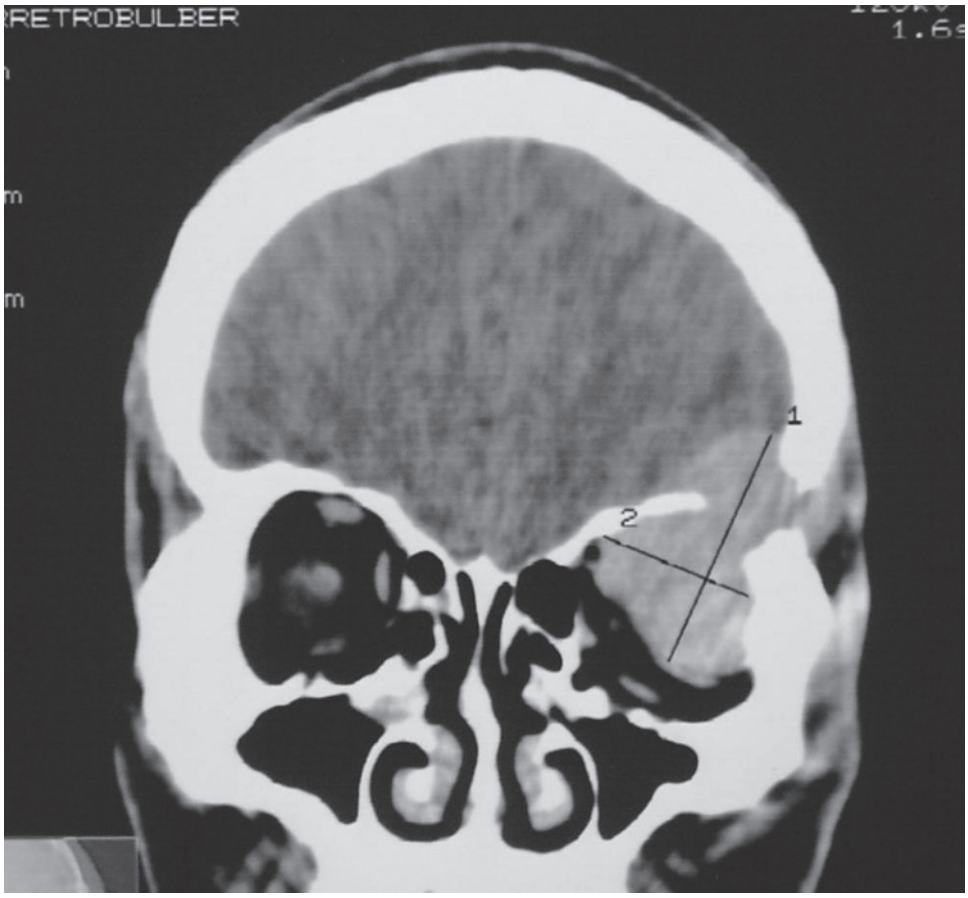

Figure 1. Coronal CT scan showing an osteolytic, well demarcated tumor with homogenous contrast enhancement in superolateral part of the right orbit

nuclear cytoplasmic ratio, and prominent nucleoli. These morphologic features were consistent with plasma cell myeloma according to WHO classification.[7]

Immunohistochemical staining revealed that majority of the plasma cells expressed CD38 and were negative for CD20. Kappa light chain was nearly not expressed (less than 10%) and lambda light chain was negative in more than 80%. Skeletal survey demonstrated multiple lytic lesions of the ribs. Bone marrow biopsy revealed extensive infiltration with myelomatous cells (more than 30%) confirming the diagnosis of multiple myeloma. Serum protein electrophoresis revealed a sharp-spiked in gamma globulin. Based on these findings, the solitary osseous plasmacytoma was part of disseminated disease from multiple myeloma. The patient received radiation (45 Gy) for the local tumor followed by systemic vincristine, adriamycine and dexamethasone (VAD) combination chemotherapy. Twelve months after surgery the patient remained well with no symptoms and signs indicative of local recurrence or relapse of the multiple myeloma. The surgical outcome was considered good. Twenty months after surgery however, the patient was readmitted to our hospital because of progressive shortness of breath over 10 days. X-ray of the thorax showed pleural effusion. Physical examination showed a cutaneous mass on the right temporal area without signs and symptoms of recurrent orbital tumor. Similar masses/nodules were also observed on the forehead, chest, arms and back. Fine needle aspiration of the nodules revealed plasmacytoma. Protein electrophoresis of pleural fluid revealed a sharp-spiked in gamma globulin. Second bone marrow aspiration showed massive infiltration of myelomatous cells (more than 60%). The patient underwent thoracic tube insertion, drainage of pleural fluid, and another VAD combination chemotherapy. However, the patient's condition deteriorated and the patient died 4 weeks after admission.

DISCUSSION

Solitary osseous plasmacytoma is characterized by osteolytic lesion and the presence of plasma cells in the biopsy.[2,3,8] Solitary osseous plasmacytoma of the orbit, either as part of a disseminated myeloma or as localized disease, is very rare.[2,3] Extramedullary plasmacytoma of the orbit very rarely presents with bony erosion.[1] Progression of plasmacytoma of the bone, especially skull base, to develop multiple myeloma is much more common than extramedullary plasmacytoma.[1,3,4-6] Therefore, determining whether a solitary plasmacytoma is an osseous or extramedullary origin is important in determining prognosis.[1]

Our case of plasmacytoma of the skull base manifested as orbital mass. Systemic studies confirmed that our patient's orbital tumor was part of a disseminated disease. Solitary plasmacytoma of the skull base very rarely presents as locally confined disease. It is usually part of the systemic disease.[3-6] Wein *et al* divided the primary site of plasmacytoma of skull base into anterior (nasopharyngeal) group and central (sphenoid, clivus, petrous apex) group.[3] In central group, the rate of progression to multiple myeloma is 63.6 %, much higher than that in anterior group (9.5 %). Thus, Bindal *et al* reported 8 cases of intracranial plasmacytoma, 2 of them originated from the central skull base (clivus in one and petrous and mastoid in the other), which were associated with multiple myeloma.[6] Pancholi *et al* reported plasmacytoma of central skull base involving the left petrous apex, clivus and sphenoid bone also associated with multiple myeloma.[5] Similarly, Ustuner *et al* reported a case of plasmacytoma of central skull base involving ethmoid sphenoid, clivus and petroclival junction in a 39 year old male, who was associated with multiple myeloma.[4] However, our patient with anterior skull base disease also had multiple myeloma.

On CT scan, plasmacytoma of the bone shows an osteolytic, well demarcated lesion with a fairly homogenous contrast enhancement that mimics meningioma.[5,6,8] An important differential diagnosis of skull base plasmacytoma is adenoid cystic carcinoma, which usually presents with bone destruction.[5]

Management of plasmacytoma of the skull base is complete removal of the tumor whenever possible, followed by radiation therapy.[6] This combination treatment give rise to high rate of local control.[6] If the tumor is not resectable, radiation therapy alone can also control the tumor locally.[4,6,9] When solitary plasmacytoma with multiple myeloma is diagnosed, local treatment should be followed by systemic combination chemotherapy.[3-6]

Involvement of the skin and the development of myelomatous pleural effusion are late manifestations of multiple myeloma, which are expression of aggressive disease.[10-12] Most patients with pleural effusion die within weeks.[10,12,13] The pathogenesis of myelomatous pleural effusion is unknown. The possible mechanisms are invasion from adjacent skeletal lesion, direct pleural involvement by myeloma, and extension from the chest wall with plasmacytoma.[12] Skeletal survey in our patient revealed multiple osteolytic lesions of the ribs. This might support the first mechanism of pleural effusion described above.

The prognosis of solitary plasmacytoma is dependent on the presence of disseminated disease.[5] Multiple myeloma is a systemic disease, having no potential for complete cure.[5] The prognosis of plasmacytoma with multiple myeloma is therefore very poor, with most patients die within 2 years after diagnosis[5], and the 3 year survival rate of around 10%.[5] In conclusion, we report a case of solitary osseous orbital plasmacytoma of the skull base with generalized myeloma. The local tumor was well controlled. But the patient died from aggressive generalized disease.

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