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Case Report Open Access

Multiple Myeloma Presenting as Unilateral Proptosis: A Case Report

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Abstract

Purpose: To report on the clinical, imaging and histopathological characteristics of an orbital mass in a patient presenting with unilateral exophthalmos.

Methods: Imaging studies revealed a lobulated orbital mass situated in the superlateral quadrant of the orbit with adjacent bony erosion of the sphenoid bone. Fine needle aspiration cytology form the orbit suggested lymphoproliferative disorder. The bone marrow showed increased plasma cells and was positive for CD 138 with Lambda light chain restriction. Serum protein electrophoresis showed IgD lambda monoclonal band, the urine was positive for Bence Jones protein. Skeletal survey confirmed multiple osteolytic lesions.

Result: The patient was diagnosed as stage IIIB multiple myeloma with orbital plasmacytoma and transferred under the care of oncologist who started bortizomib based chemotherapy with very good clinical and laboratory response after the second cycle and almost 100% clinical recovery of the eye proptosis.

Conclusion: The diagnosis of multiple myeloma should be borne in mind in cases of unilateral proptosis as early diagnosis and treatment can be vision saving in these patients. This can be established with extensive histopathological and biochemical investigations.

Keywords: Proptosis; Orbital tumors; Multiple myeloma

Introduction

Multiple myeloma usually presents as a systemic, disseminated disease. It represents an uncontrolled proliferation of plasma cells with overproduction of proteins belonging to the immunoglobulin family. It accounts for 1% of all neoplasms [1]. The condition demonstrates an equal male to female occurrence with peak incidence in the fifth decade. The plasma cells frequently proliferate in the bone marrow and invade the adjacent bone resulting in bone pain with associated increase in the levels of paraproteins in serum and urine.

Plasma cells can cause a localized neoplastic proliferation like solitary plasmacytoma of the bone or extramedullary plasmacytoma. Orbital involvement can occur as a solitary plasmacytoma or as a part of systemic involvement in multiple myeloma.

Case Report

A 39 year old male patient from Bangladesh was referred to the Ophthalmology Department with a history of diminution of vision in the left eye accompanied by progressive proptosis for two weeks. Visual acuity of the Right eye was 6/6 and the left eye was 1/60. Examination of the left eye showed proptosis and 2 mm inferior dystopia with periorbital edema and conjunctival chemosis. Motility examination revealed limitation in movements in all gazes of the left eye).

Hertel exophthalmometry reading was 19 mm OD and 25 mm OS The proptosis was irreducible, and auscultation did not reveal any thrill or bruit. Intraocular pressure was 22 mm of Hg (Figure 1). The anterior segment examination was unremarkable. Funduscopic examination showed blurring of the disc with Choroidal folds were present in the left eye.

Two months prior to the eye complaint, the patient reported backache of two months duration for which he consulted a neurosurgeon. Magnetic resonance imaging (MRI) of the dorsolumbar spine showed diffuse abnormal marrow infiltrative process and a large extra- osseous soft mass opposite to D10 and D11. There was no cord compression. The patient also complained of buccal swelling on the left side of the hard palate.

He had a history of pulmonary tuberculosis and was treated with a course of ATT in 2002.

Routine complete blood count showed normocytic, normochromic anemia (Hemoglobin 9.9 g/dl). White blood cell and platelet counts were normal, but the erythrocyte sedimentation rate was elevated.

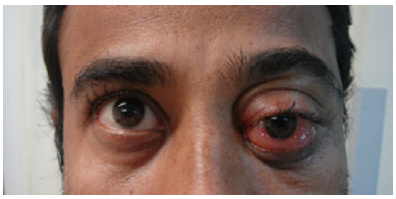

Figure 1: Photograph showing axial proptosis and inferior displacement of the left eye.

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Blood chemistry showed hypercalcemia and elevated BUN and reatinine.

Contrast enhanced computerized tomography (Figure 2) and MRI (Figure 3) of brain and orbit revealed a 6 mm soft mass in the left parietal bone causing lytic bony defect. A 3.1 x 1.3 x 2.7 cm extraconal, lobulated soft mass associated with bony erosion of the greater wing of the sphenoid is present in the left orbit. The mass is displacing the globe medially. MRI also showed two periodontal masses in relation to the roots of teeth at the left side of the upper jaw and left side of the lower jaw.

Based on the clinico-radiological findings, diagnostic possibilities of lymphoma, plasma cell related tumour, and metastasis were considered.

Fine needle aspiration of the orbital mass under CT guidance revealed atypical lymphoid cells with many bizarre plasma cells suggestive of plasma cell lymphoproliferative disorder.

Bone marrow aspiration (Figure 4) and biopsy (Figure 5) revealed sheets of plasma cells infiltrating the marrow and constituting 40% of the nucleated population. Cells were CD 138 positive with lambda light chain restriction. Karyotyping was normal. FISH (spell it out) study was not available.

Additional workup included serum protein electrophoresis which showed a monoclonal gammopathy of IgD Lambda. The serum free light chain lambda was 2734 mg/l (5.71-26.3), serum free light chain kappa 12.9 mg/l (3.3-19.4), and a Kappa to Lambda ratio of 0.005 (0.26-1.65). Twenty-four hour urine protein electrophoresis was positive for Bence-Jones protein. A skeletal survey showed generalized osteopenia, a large osteolytic lesion of the 10th rib and multiple small lytic foci involving pelvic bones, ribs and dorsolumbar vertebrae.

A final diagnosis of IgD lambda multiple myeloma stage IIIB manifesting as orbital involvement at presentation was reached.

The patient received bortizomib based chemotherapy (Bortizomib, Dexamethasone Lenalidomide, and liposomal doxorubicin every 21 days. After two cycles of this regimen; the left eye protrusion subsided completely (Figure 6) with regression of the buccal swelling.

After four cycles of the protocol; kidney function normalized (serum creatinine 89 µmol/l), hemoglobin returned to normal, and 13.7 gm/dl, the serum protein electrophoresis and immunofixation became normal with disappearance of Bence-Jones proteinurea.

The serum free light chain lambda was 22.8mg/l, serum free light chain kappa 21.8 mg/l, and the Kappa to Lambda ratio 0.965.

We plan for a total 6 cycles of courses of chemotherapy and then autologous stem cell transplantation followed by high dose therapy and autologus stem cell transplantation.

Discussion

Plasma cell dyscrasias are rare causes of proptosis. Dalrymple and Bence Jones in 1846 described neoplastic proliferation of plasma cells. The disseminated proliferation of plasma cells was characterized by marked proteinuria and bone pain [2]. Plasma cell tumours are classified into three main categories; multiple myeloma, medullary plasmacytoma and extramedullary plasmacytoma. Medullary plasmocytomas progress to multiple myeloma in 85% of patients; extramedullary plasmocytomas usually remain well localized. It is essential to perform systemic survey to search for multiple myeloma

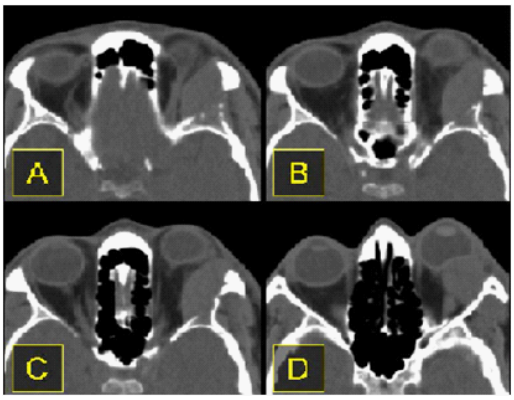

Figure 2: Orbital mass. A-D) Axial orbital CT scans show a solid mass at in the superolateral part of the left orbit causing globe displacement and bone destruction of the greater wing of sphenoid.

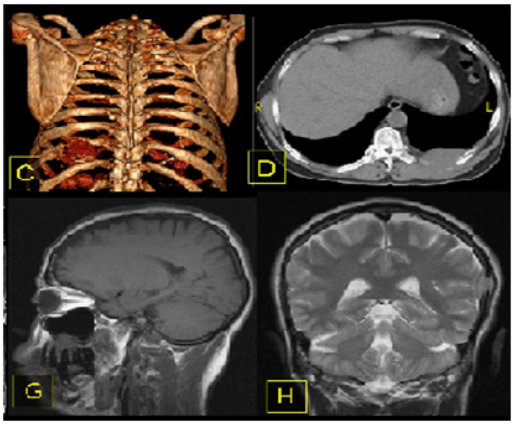

Figure 3: Extraorbital bony lesions. C) Volume rendered multi-detector CT image of the thorax, seen from the back, and D) transverse CT image reveals a focal osseous lesion of the left tenth rib. G) Sagittal T1-weighted MR image shows a lesion at the soft palate resting on the roots of the upper teeth. H) Coronal T2-weighted MR image of the head shows a focal lesion of the left side of the skull. The lesion has the same signal characteristics as that of the left orbital mass.

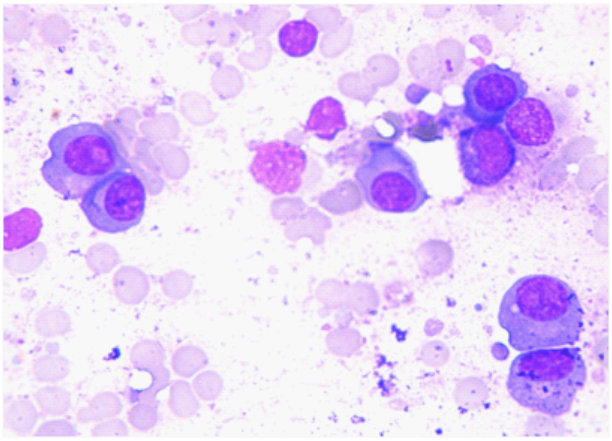

Figure 4: Bone marrow aspirate: Plasma cells of varying stages of maturation from mature plasma cell ⬅ to immature cells (Plasmablasts) ⬇ Wright stainX1000.

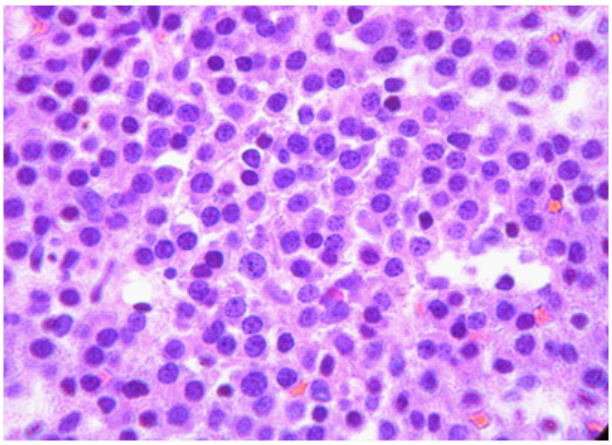

Figure 5: Bone marrow biopsy disclosing a uniform sheet of plasma cells. H&E stainX1000.

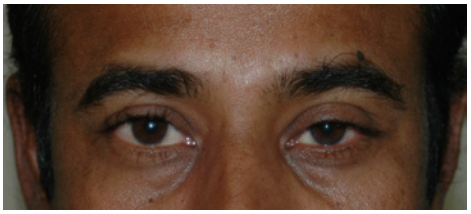

Figure 6: Resolution of proptosis and conjunctival chemosis after two cycles of chemotherapy.

when a plasma cell neoplasm is diagnosed. The evaluation includes radiological survey, serum and urine immunoelectrophoresis, and bone marrow biopsy.

Multiple myeloma is characterized by neoplastic monoclonal proliferation of the highly specialized B-lymphocytes producing a single immunoglobulin. It is engaged in the production of paraproteins or light chain moiety. Multiple myeloma represents 1% of all neoplasms. The principal clinical presentation of orbital multiple myeloma, plasmacytoma or primary (or solitary) extramedullary plasmacytoma in the orbit is proptosis, with a predilection for the superotemporal quadrant [1].

Orbital myeloma is rare and the incidence is 0.25% [3,4]. Bilateral proptosis with multiple soft tissue masses is an extremely rare presentation of multiple myeloma. Six cases of bilateral proptosis due to multiple myeloma have been reported [7-10].

Multiple myeloma cause a variety of complication some of which are life threatening such as hypercalcemia, impaired kidney function, infection, bone pain and fractures, spinal cord compression, anemia and hyperviscosity syndrome [11]. At the time of orbital presentation, our patient had skeletal involvement, positive bone marrow biopsy, anemia, hypercalcemia, and renal insufficiency.

Clinically, proptosis is not pathognomonic for MM, a feature that is seen in all orbital tumors, benign or malignant. Ultrasound and imaging features are suggestive of MM. The key feature is that this orbital soft tissue mass is associated with bone erosion. So, when a clinician sees this CT finding, MM should be in the differential diagnosis – before a biopsy is performed. MM is not a surgical disease. Once a biopsy is made, a systemic search should be done for staging. Chemotherapy is the usual treatment for MM. If chemotherapy fails, stem cell transplant is the next advancement in treatment.

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