CASE REPORT

Clivus and Dural Involvement in a Case of Multiple Myeloma: A Rare Complication of Multiple Myeloma

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Background

Multiple myeloma (MM) represents a malignant proliferation of plasma cells derived from a single clone. The tumor, its products, and the host response to it result in a number of organ dysfunctions and symptoms, including bone pain or fracture, renal failure, susceptibility to infection, anemia, hypercalcemia, and occasionally clotting abnormalities, neurologic symptoms, and manifestations of hyperviscosity. Intracranial lesions of MM commonly involve the cranial vault, the skull base and the orbit [1]. The clivus and petrous regions are far less commonly affected [2].

We report an unusual presentation of multiple myeloma with neurological involvement in the form of clivus mass causing progressive external ophthalmoplegia which was treated with whole brain radiotherapy (WBRT).

Case Presentation

A 55 year old male a known case of multiple myeloma since Sep 2009, presentation at onset included fatigability, low backache and M band on SPEP, bone marrow showed numerous plasma cells (30 %) and was labelled as symptomatic multiple myeloma Stage II with CRAB 3/4 (serum calcium—10.8 mg/l, Hb—9.5 g %, lytic lesions present) and had no evidence of extramedullary disease. His serum immunofixation reports were not available. Initially he received thalidomide and dexamethasone and achieved

remission after six cycles. He had multiple relapses from 2011 to 2013, clinically as well as biochemically. In June 2013 he was started on melphalan, prednisolone and lenalidomide (MPL) following relapse without any extra-medullary spread, CRAB 2/4 (serum Calcium 11 mg/dl lytic lesions) and had received three cycles of MPL till presentation. Presently he came with a 7 day history of left frontal headache followed by diplopia. Headache was subacute in onset, continuous, dull aching type with no photophobia, phonophobia, nausea, vomiting, lacrimation, diminution of vision with minimal amelioration with NSAIDs. Diplopia was subacute in onset, progressive, painless and persistent. He also noticed progressive inward and downward deviation of his left eye. He denied history of trauma, fever, rash, weakness, and any other complaints.

On examination he had pallor, ptosis in right eye and lateral rectus palsy of the left eye (Fig. 1). There were no other neurological deficits or features of meningism. Fundus was normal and the rest of his systemic examination was normal.

On evaluation he had two of the CRAB criteria (normochromic normocytic anaemia, Hb 10 g/dl, normal renal functions, serum Calcium >11 mg/l, no lytic bone lesions) and a raised ESR (46 mm/h). His renal function tests, 24 h urine protein and serum electrolytes were normal. MRI Orbits and Brain (Fig. 2) showed loss of normal fatty marrow of the clivus with thickening and abnormal enhancement of overlying dura suggesting infiltration of clivus. Cerebrospinal fluid analysis (CSF) was negative for malignant cells.

A diagnosis of refractory multiple myeloma with clivus mass was made and the patient was taken up for whole brain radiation. Ten cycles of WBRT were administered to a total dose of 3000 Gy over a period of 2 weeks. Post radiotherapy his headache and diplopia subsided.

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Fig. 1 Lateral rectus palsy *left eye* and ptosis *right eye*

Discussion

Although multiple myeloma commonly involves the nervous system, intracranial involvement is rare [3]. In 1932 Cushing found only four cases of multiple myeloma amongst 2000 intracranial tumours [4]. Clarke has divided cranial myelomas into three groups [5]. In Group 1, the tumor involves the skull base and characteristically manifests with cranial nerve palsies. Group 2 tumors are termed “intracranial tumor syndromes” because the myeloma extends into the parenchyma of the brain with or without involvement of the skull. Tumors in Clarke’s Group 3 are “intra-orbital tumor syndromes.” By the 1950s, there were only 25 histologically verified cases of multiple myeloma associated with cranial nerve palsies [5]. 15 of these 25 cases involved generalized multiple myeloma, whereas the other 10 were solitary plasmacytomas. The sixth nerve as in our case is the most common cranial nerve affected by multiple myeloma, followed by the fifth and eighth nerves [5]. The body of sphenoid and apex of the petrous pyramid are the most commonly involved sites for multiple myeloma affecting the skull [6]. The differential diagnosis for clivus based masses includes chordoma, metastatic disease, chondrosarcoma, and plasmacytoma. Multiple myeloma may involve the region of the sella turcica and mimic a pituitary adenoma [5]. Leptomeningeal involvement is associated with a very poor prognosis, however patients with dural involvement show a more favourable response to combined chemoradiotherapy [7]. Cranial nerve palsies, particularly sixth nerve paresis may be initial presenting features of plasmacytoma and multiple myeloma or may manifest in patients on follow up. Although dural infiltration in multiple myeloma is very rare, it may closely resemble meningioma [8]. Local irradiation is the primary mode of treatment for extramedullary plasmacytoma and multiple myeloma, occasionally followed by surgical

Fig. 2 T1WI sagittal image showing dural thickening overlying the posterior aspect of clivus which shows post contrast enhancement on FLASH 3D sagittal image. Also noted is the loss of T1 hyperintensity in clivus consistent with loss of normal fatty marrow

resection of the residual tumor [9]. The prognosis is poor, and most patients die within 2 years of their diagnosis; 3-year survival is only about 10 % [10].

Conclusion

In a case of multiple myeloma presenting with progressive headache and diplopia, a possibility of CNS involvement in the form of clivus and dural involvement must always be

thought of and subjected to appropriate neuroimaging. WBRT is the palliative modality of choice in such cases.

Compliance with Ethical Standards

Conflict of interest All authors have nil to declare.

Informed consent Informed consent was obtained from the patient for whom identifying information is included in this article.

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