CASE REPORT

MULTIPLE MYELOMA PRESENTING AS THIRD CRANIAL NERVE PALSY: A RARE CASE REPORT

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HOW TO CITE THIS ARTICLE:
Lohit Kumar Kalita, Chayanika Kalita, Pabitra Kumar Gogoi, Umesh Ch. Sarma. "Multiple Myeloma Presenting as Third Cranial Nerve Palsy: A Rare Case Report". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 15, April 13, 2015; Page: 2375-2378.

ABSTRACT: Compression of cranial nerves (CNs) by an intracranial plasmacytoma is considered to be an unusual presentation of multiple myeloma (MM). Here, we report a case of right 3rd cranial nerve involvement as an initial presentation, which emphasizes the fact that third CN palsy can be the first presenting feature of Multiple Myeloma (MM).
KEYWORDS: Multiple myeloma, plasmacytoma, third cranial nerve, palsy.

INTRODUCTION: Central nervous system (CNS) involvement in multiple myeloma (MM) is rare and accounts for only 1% of cases.[1] Again neurologic symptoms due to plasmacytomas located either in the base of the skull or at intracranial locations are extremely rare. The median interval from initial diagnosis of multiple myeloma to the CNS involvement is about 11 to 13 months.[1] Though multiple cranial neuropathies in late stages of MM is not uncommon, the report of isolated cranial neuropathies as presenting symptoms of solitary clival plasmacytoma is few.[2] Very rarely multiple cranial nerves (CN) involvement by plasmacytoma may be presenting feature of MM.[3,4,5] Here, we report a case of right third CN palsy as first presentation of intracranial plasmacytoma.

CASE REPORT: A 70-year-old male patient presented with the complaints of drooping of right upper eye lid for three days. There was history of mild headache, weakness, recurrent fever and back pain since 3 months, for which he was receiving conservative treatment from local treating physician. His physical examination showed severe pallor and generalized bony tenderness. The neurological examination revealed pupil sparing right third cranial nerve paresis [Figure : 1]. His blood picture showed haemoglobin of 4.2 g/dl, total leucocytes count was 9990 cells/cu mm with neutrophil 81.2% and lymphocyte 14.1%, monocytes 4.1% and eosinophil count was 0.7%. The erythrocyte sediment rate was 160 mm at first hour. The peripheral blood smear showed normal in count and morphology of WBC series with moderate anisocytosis with microcytic hypochromecia and target cell in RBC series. There was mild rouleaux formation and a few NRBC. The platelets were adequate in number and morphology. The routine examination was normal and urinary Bence-Jones protein was absent. The blood chemistry showed serum creatinine of 1.46 mg/dl, urea of 58.48 mg/dl, serum calcium was 9.5 mg/dl. There rheumatoid factor and antinuclear antibody were absent. Bone marrow aspiration showed more than 20% plasma cells. Serum electrophoresis showed presence of M-Spike in the gamma globulin region. His X-ray skull showed multiple punched out lytic lesions [Figure: 2]. Computed tomography of brain showed (a) multiple extensive punched out osteolytic lesions involving all bones skull as well as the skull base. (b) A heterogenous lesion involving right petrous apex and basi sphenoid which extends to

bilateral sphenoid sinus [Figure: 3]. Once the diagnosis of MM with multiple plasmacytoma was confirmed, the patient was put on treatment with bortezomib and dexamethasone.

DISCUSSION: Plasmacytoma and multiple myeloma represents the spectrum of the same disease, where plasmacytoma refers to the localized form and MM implies systemic dissemination. The plasmacytoma may be intramedullary or extramedullary type. Again it can be primary in origin or can arise as secondary to disseminated MM. MM may show evidence of intracranial or orbital encroachment at any stage of the disease process, and occasionally this may be the first clinical manifestation. Our patient presented initially with right 3rd CN palsy, and subsequent investigation confirmed the diagnosis of plasmacytoma with MM. Among the neurologic complications in 277 myeloma patients studied by Silverstein and Doniger et al, the spinal cord compression was the most frequent (27 cases, 9.7%) presentation, while CN involvement was unusual (7 cases, 2.5%) and it was the initial manifestation in only five patients (1.8%).[1] Among these seven patients one patient had isolated 6th CN palsy and another one had 3rd, 4th and 6th CNs palsy. Two other patients had papilledema one of which was associated with 7th and 12 CN palsy. Reast of the patient had retro-orbital myeloma associated with proptosis or extraocular involvement. Among the 25 proven cases of myeloma with CN palsies studied by Clarke et al, the 6th CN was the most common involvement followed by 7th, 8th and 5th CNs.[2] The present case belongs to Group I of Clarke's clinical classification of myelomas. There are several cases of isolated cranial neuropathies as symptoms of solitary plasmacytoma. The multiple cranial neuropathies in combination such as Foster Kennedy syndrome, cavernous sinus syndrome, Gradenigo syndrome and jugular foramen syndrome had been reported in late stages of MM.[6,7,8] Multiple CNs involvements as the initial presentation for plasmacytoma had been reported very rarely.[3,4,5] Ko et al. reported a case of the intracranial plasmacytoma in the cavernous sinus presented as left 6th and partial 5th CN palsy.[3] Doniger et al. has reported a case of plasmacytoma in spinoclival region with disseminated MM who presented as third and fourth complete CN palsy.[4] Tappin et al. has reported a case of lower CN palsy in the form of Collet-Sicard syndrome, which turned out to be the intracranial plasmacytoma.[5] In-spite of being in stage II (International Stagging System) MM in terms of areas of spread/conversion with deranged laboratory data, the patient in this report did present with isolated cranial neuropathies with involvement of 3rd CN as a sign at presentation leading to the diagnosis. CT scan study in this patient revealed a single plasmacytoma in the base of the skull involving right petrous apex, basi sphenoid extending to involve bilateral sphenoid sinus causing compression of right 3rd CN. The extent of the lesion co-relate with our 3rd CNs involvements. CN involvement is almost always due to direct compression of CNs by tumor arising from skull.[1] When plasmacytoma appears at different locations in the context of MM, the histological diagnosis of all these lesions is not needed.[4] We are not certain whether plasmacytoma was the initial manifestation of MM or plasmacytoma has progressed to MM. Both circumstances carry a worse prognosis and it should be treated aggressively.[9]

CONCLUSION: The intracranial plasmacytoma is a rare manifestation of multiple myeloma and 3rd CN palsy can be the rare first clinical presentation. Though rare, multiple myeloma should be considered in differential diagnosis of such patients.

REFERENCES:

1. Silverstein A, Doniger DE. Neurological complications of myelomatosis. Arch Neurol 1963; 9: 102-12.
2. Clarke E. Cranial and intracranial myelomas. Brain 1954; 77: 61-81.
3. Ko S, Huang SY, Liu CY. Extramedullary plasmacytoma masquerading as Tolosa-Hunt syndrome: A case report. BMJ Case Rep 2009.
4. Montalban C, Martín-Aresti J, Patier JL, Millan JM, Cosio MG. Unusual cases in multiple myeloma and a dramatic response in metastatic lung cancer: Case 3. Intracranial plasmacytoma with cranial nerve neuropathy in multiple myeloma. J Clin Oncol 2005; 23: 233-5.
5. Tappin JA, Satchi G, Corless JA, Ashworth F. Multiple myeloma presenting as the Collet-Sicard syndrome. J Neurol Neurosurg Psychiatry 1996; 60: 14.
6. Morgan WK. Foster Kennedy syndrome produced by myelomatous deposits in the skull. Br Med J 1955; 1: 708.
7. Kyoda K, Nakamura S, Takeshima M, Matano S, Masuda S, Matsuda T, et al. Cavernous sinus syndrome associated with nonsecretory myeloma. Intern Med 1999; 38: 988-90.
8. Bourne RR, Maclaren RE. Intracranial plasmacytoma masquerading as Gradenigo's syndrome. Br J Ophthalmol 1998; 82: 458-9.
9. International Myeloma Working Group. Criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders: A report of the International Myeloma Working Group. Br J Haematol 2003; 121: 749-57.

Fig. 1: Patient with right 3rd CN palsy

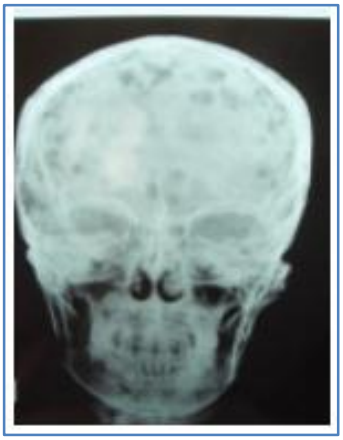

Fig. 2: X- ray of skull of the patient shows multiple radiolucent punched out lytic areas

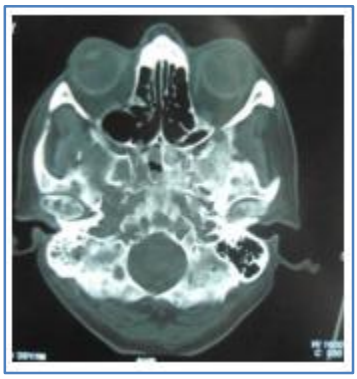

Fig. 3: A heteroechoic lesion involving right petrous apex, basi sphenoid extending to involve bilateral sphenoid sinus

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Date of Submission: 18/03/2015.
Date of Peer Review: 19/04/2015.
Date of Acceptance: 02/04/2015.
Date of Publishing: 13/04/2015.