A case of multiple myeloma with cranial nerve palsy

SURESHBABU THIRUMAL
Department of Neuro Surgery,
MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract:
We report a case of multiple myeloma with right sixth cranial nerve palsy. A 50 years old male tailor by occupation presented with complaints of back pain for one and half years, headache for 2 months, swelling in the scalp for one month and double vision on the right side for three weeks. The patient was symptomatically treated by a general practitioner and referred with MRI scans. A 3 x 2 cm bony swelling in the left parietal region was excised and HPE shows features of multiple myeloma. The radiology, bone marrow aspiration study, immunoelectrophoresis and blood parameters support the diagnosis of multiple myeloma. The patient was referred to haematologist and radiotherapist for further management.

Keyword: Multiple myeloma, plasmacytoma

A 50 years old male, tailor by occupation was admitted with complaints of backache for one and half years, headache for 2 months, swelling in the scalp 1 month and double vision on seeing to right side for 3 weeks. He took analgesics for the back pain on and off. The double vision in the right eye made him to attend an ophthalmologist. He took MRI brain and spine and referred the patient to our institute for the left parietal swelling. There is no history of seizures, loss of consciousness, vomiting, fever, weight loss and any symptoms of tuberculosis.

On examination he was moderately built, well nourished, no pallor, not icteric, no generalized lymphadenopathy and no neurocutaneous markers. His higher mental functions, lobar functions, spinal motor system, sensory system, cerebellum and autonomic nervous system examinations were normal. Except for the right sixth cranial nerve involvement, examination of other cranial nerves was normal. On the left parietal region 3 x 2 cm bony hemispherical, smooth, non tender swelling was present.
MRI brain shows parietal bony lesion with abnormal marrow signal involving basisphenoid clivus associated with destruction of soft tissue component causing expansion of bone with bulging into prepontine cistern posteriorly and the sphenoid sinus anteriorly. Multiple abnormal signal deposits present in the skull base and also in the left parietal bone. Brain parenchyma was normal. MRI spinal screening shows collapse of D2, D7, D8, D10 and L1 to L5 vertebra. Multiple abnormal hyper intense lesions were seen in pelvic bone, sacrum and both femur.

Figure 1c preoperative MRI showing clival enlargement with compression of prepontine cistern Figure 1d Figure 1d X-ray skull showing osteolytic lesions

Figure 1a-1b T2WI & T1WI - preoperative MRI showing lesions in the parietal bone and skull base.

Figure 1e CT skull clival involvement extending into prepontine cistern
The patient underwent total excision of the left parietal bony lesion. Surgical procedure – under ETGA, patient in supine position head turned towards the right inverted U shaped incision made over the swelling in the left parietal region, scalp flap raised. The parietal bone was found eroded and a 3x2cm soft, greyish non suckable, non hemorrhagic hard lesion was seen protruding through the defect in the parietal bone. The lesion was excised along with the bone that was eroded. The underlying dura was intact. The specimen which was sent for HPE showed features suggestive of multiple myeloma.

The histopathological examination of the tumour showed cells bearing characteristic morphologic features of plasma cells i.e., round, and oval cells with an eccentric nucleus with coarsely clumped chromatin, densely basophilic cytoplasm and a perinuclear zone containing the golgi apparatus. Bone marrow aspiration study also showed features of multiple myeloma with plasma cells more than 30%. The peripheral blood smear shows features of microcytic, hypochromic anaemia. Serum electrophoresis showed the characteristic M BAND. However urine is negative for Bence-Jones protein. The urine may be negative for bence jones protein in 1/3 of the patients with multiple myeloma. The serum calcium is 8.4mgs. Serum LDH is 648u/l and serum B2 MICROGLOBULIN is 8950ng/ml. Serum B2 microglobulin is the single most powerful predictor of survival in patients with multiple myeloma.

Patient was treated with steroids for the sixth cranial nerve palsy. Since multiple myeloma responds very well to chemotherapy the patient has been referred to hemat-oncologist – expecting recovery of sixth cranial palsy. Incase of failure of chemotherapy to relieve the cranial nerve palsy with, the same can be corrected by clival resection.
Multiple myeloma, also known as plasma cell myeloma or Kahler's disease, is a cancer of plasma cells, a type of white blood cell normally responsible for producing antibodies. In multiple myeloma, collections of abnormal plasma cells accumulate in the bone marrow, where they interfere with the production of normal blood cells. Most cases of myeloma also feature the production of a paraprotein, an abnormal antibody which can cause kidney problems. Bone lesions and hypercalcaemia (high calcium levels) are also often encountered. Myeloma develops in 1–4 per 100,000 people per year. It is more common in men.

The classic triad of multiple myeloma is marrow plasmacytosis, lytic bone lesions and serum or urine M component. Marrow plasmacytosis results in normocytic normochromic anemia, rarely granulocytopenia and thrombocytopenia. There is increased risk of infection due to immune deficiency. Apart from replacement of marrow the above said signs may also be due to tumour induced inhibition of haematopoiesis.

Lytic bone lesions cause bone pain, hypercalcaemia and pathological fractures. Bone pain affects almost 70% of patients and is the most common symptom. Myeloma bone pain usually involves the spine and ribs, and worsens with activity. Persistent localized pain may indicate a pathological bone fracture. Involvement of the vertebrae may lead to spinal cord compression.
M component an abnormal monoclonal immunoglobulin occurring in the serum in plasma cell dyscrasias, formed by proliferating concentrations of immunoglobulin-producing cells. M protein can be IgG, IgA or IgD. Serum M component can be detected by electrophoresis. Two-thirds of patients with serum M components also have urine light chains. The proteins are immunoglobulin light chains (paraproteins) and are produced by neoplastic plasma cells. They can be kappa (most of the time) or lambda. The light chains can be immunoglobulin fragments or single homogeneous immunoglobulins. Bence Jones proteins are particularly diagnostic of multiple myeloma in the context of end-organ manifestations such as renal failure, lytic (or "punched out") bone lesions, anemia, or large numbers of plasma cells in the bone marrow of patients. Bence Jones proteins are present in 2/3 of multiple myeloma cases. Serum b2 microglobulin is a protein of 11000 mol wt with homology to the constant region of immunoglobulins i.e. the light chain of class 1 MHC antigens (HLA A,B,C) on the surface of every cell. Serum B2 microglobulin is the single most powerful predictor of survival in patients with multiple myeloma. Common problems are weakness, confusion and fatigue due to hypercalcaemia. Headache, visual changes and retinopathy may be the result of hyperviscosity of the blood depending on the properties of the paraprotein. Finally, there may be radicular pain, loss of bowel or bladder control (due to involvement of spinal cord leading to cord compression) or carpal tunnel syndrome and other neuropathies (due to infiltration of peripheral nerves by amyloid). It may give rise to paraplegia in course. However, isolated sixth nerve palsy can be explained by involvement of the clivus.

Due to close proximity of the cranial nerves at the skull base and to the cavernous sinus and multifocal nature of multiple myeloma the disease tends to affect multiple cranial nerves rather than single cranial nerve as it has happened in this patient. In our case sixth nerve palsy can be explained by the involvement of the clivus. Abducens nerve palsy is the most common cause of acquired extraocular muscle paralysis. It contains somatic motor fibres that arise from their nucleus in the pons and innervate the lateral rectus to control abduction of the eyes. The nerve travels along the base of the brain and enters the cavernous sinus, after traversing the petrous part of the temporal bone and ascending the clivus. The clivus is a shallow depression formed by the sphenoid and occipital bones and is located just posterior and slightly inferior to the sella turcica behind the dorsum sellae. It is continuous with the basilar portion of the occipital bone and supports the upper part of the pons. The abducens nerve pierces the dura of the clivus, passes through the Dorello’s canal formed by the petroclinoid ligament and then enters the cavernous sinus. This nerve exits the cavernous sinus through the superior orbital fissure and innervates the lateral rectus muscle. Because of its small size, long oblique course and anatomical location the 6th nerve is particularly susceptible to insult. This case is being presented because of the paralysis of the sixth cranial nerve alone in spite of the extensive involvement of the skull base with multiple myeloma. It is commonly held that eye signs in multiple myeloma are unusual. (Fung S et al. ophthalmic manifestations of multiple myeloma. Ophthalmologica 2004;219;43-8). Among these eye signs dysfunction of 2, 3, or 6 cranial nerves is common and in fact the sixth nerve is the most commonly affected because of its long course. It may give rise to paraplegia in course. However, isolated sixth nerve palsy is a late presenting case.
involvement alone as the initial presenting symptom of skull base multiple myeloma is rare. As multiple myeloma responds better to chemotherapy and irradiation this patient was referred to haemato-oncologist and radiation oncologist for further management.

In literature, cases of skull base multiple myeloma have been reported with multiple cranial nerve involvement. In our case isolated involvement of the sixth cranial palsy has been noted. This is due to the involvement of the greater wing of sphenoid extending only up to the lower compartment of the superior orbital fissure. The upper and the middle compartments of the superior orbital fissure are free which explains the intactness of the 3rd and 4th cranial nerves.

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