

Case Report-III

Plasmacytoma of the Base of Skull - A Case Report

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ABSTRACT

Extramedullary plasmacytomas (EMP) are rare and may occur either as a part of a generalized disease (Multiple myeloma) or a local entity. Extramedullary plasmacytomas occurring as solitary (primary) tumour or secondary manifestations of multiple myeloma most often involve the upper airways and paranasal sinuses. Skull base is one of the rarest sites for extramedullary plasmacytoma. We present one such case.

INTRODUCTION

Plasma cell neoplasms constitute a group of disorders characterized by monoclonal proliferation of plasma cells and the presence of monoclonal immunoglobulin (M-Component) in the serum and or urine. Plasma cell neoplasms occurring in the head and neck region can be classified as extra-medullary plasmacytoma or solitary plasmacytoma. These disorders essentially represent distinct manifestation of a disease continuum. The clinical findings are critical to diagnosis, and distinguishing one disorder from the other. Findings or imaging

associated with this rare site of plasmacytoma involvement are reported.

CASE: A 58 year old male patient, presented to the ENT department with history of decreased hearing (more on the left) and ear pain for one month. History of vertigo and headache of 3 months duration was present. There was no history of vomiting, seizures, visual loss, fever, ear discharge or sinusitis. He was a known hypertensive.

On examination vitals were normal. Left sensory neural deafness was documented. There were no motor deficits. Blood parameters were within normal limits. Ultrasonography revealed fatty liver, pancreas and prostatomegaly. Patient was referred to radiology department to rule out middle ear / inner ear pathology.

Radiograph of skull revealed geographic lytic lesion with clear margins involving the base of skull. Contrast enhanced CT (CECT) revealed lytic destructive lesion with areas of spotty calcification involving the entire clivus, basi-occiput and occipital condyles infiltrating the bilateral petrous apices laterally, left internal auditory canal, jugular bulb and sinus plate of mastoid air cells. Left pterygoid plate and posterior aspect of nasal bone were involved. Fig.1 (a & b). Differential diagnosis of chordoma, lymphoma and plasmacytoma was considered.

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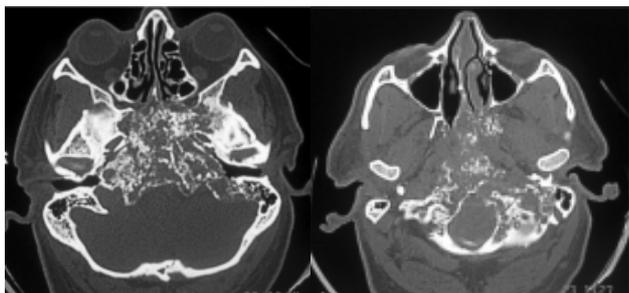


Fig. 1 CECT of the skull showing lytic destructive lesion with areas of spotty calcification involving the entire clivus, basiocciput and occipital condyles

MRI showed tonsillar herniation and a lobulated lesion appearing isointense on T1WI with mixed signal on T2WI in the region of clivus extending into the anterior lip of foramen magnum involving the basi-occiput and extending anteriorly into the base of the skull. Fig. 2

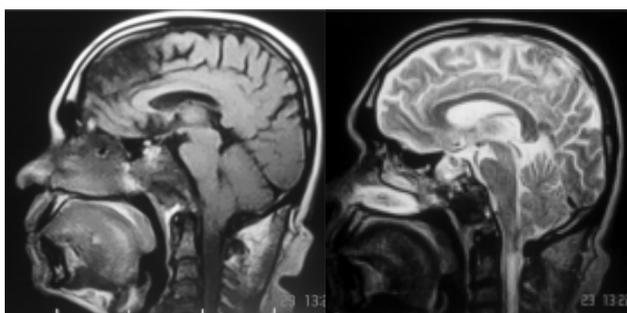


Fig. 2: MRI showing tonsillar herniation and lobulated lesion appearing isointense on T1WI with mixed signal intensity on T2WI in the region of clivus extending into the anterior lip of foramen magnum involving the basiocciput and extending anteriorly into the base of the skull.

Bone marrow aspiration showed hypercellularity with normoblastic maturation, basophilia and hyperlobated form of megakaryocytes and plasma cells with M: E ratio of 4:1 which confirmed plasmacytoma. No other lytic lesion was noted on skeletal survey.

DISCUSSION

Extramedullary plasmacytomas (EMP) are rare lesions and can occur either as a part of a generalized disease (Multiple myeloma) or a local entity. It is possible for an apparent solitary plasmacytoma to be the first presenting feature

of a generalized disease. Extramedullary plasmacytomas can involve either bone or soft tissue.

Plasmacytoma account for less than 1% of all head and neck tumours. 80% these occur in the nasopharynx and the paranasal sinuses. Rarely these may occur in the skull base, larynx, hypopharynx, parotid gland, submandibular gland, thyroid, mandibular region, trachea, esophagus, cervical lymph nodes, middle ear, orbit, scalp, forehead, palate, tongue and mastoid.¹⁻⁵

Extramedullary plasmacytoma are four times more likely to occur in males than in females and 95% of these tumours occur over the age of 40 years (mean age is 59 years).⁴

When the skull is involved, most occur in the calvarium and the skull base is rarely affected.⁵ Here, they may occur at the clivus or petrous apex where they represent a solitary plasmacytoma of bone (SBP) or they may originate within the submucosa of the sino-nasal and nasopharyngeal tracts where they represent an extramedullary plasmacytoma.²

Skull base lesions are usually asymptomatic, but larger tumours can cause symptoms. Neurologic symptoms due to plasmacytomas located either in the base of the skull or at intracranial locations are extremely rare.⁶ The cause of the neuropathy is the direct compression of nerves or nerve groups in their intracranial course. The most often involved cranial nerves are VI, II, V, VII and VIII in that order.¹ Skull base plasmacytoma may involve the posterior inferior cerebellar artery, which in turn leads to lateral medullary syndrome manifested by dysphagia, vertigo, vomiting, ipsilateral paralysis of soft palate, ipsilateral Horner's syndrome, ipsilateral hypotonia and ataxia, and dissociated sensory loss.⁷

Imaging studies include skeletal survey, computed tomography (CT),⁸ and MR imaging. More recently, position emission tomography (PET) with fluorodeoxyglucose (FDG) has been used to study relapsing patients in whom routine imaging is not very helpful. PET, in this instance, has been found to aid in detection of unsuspected sites of medullary and extramedullary disease.⁹

On plain radiographs, solitary bone plasmacytoma classically has a lytic appearance with clear margins and a narrow zone of transition to a nearly healthy surrounding bone. Rare occurrences of a cyst, a trabeculated lesion resembling a giant cell tumour, aneurysmal bone cyst, and sclerotic lesions have been described.

On CT scan plasmacytomas are well-demarcated tumours, occasionally aggressive with bone destruction and involvement of adjacent structures.¹⁰ On MR they are isointense on T1 weighted images and moderate signal intensity on T2 weighted images. There is significant contrast enhancement with central inhomogeneity.¹¹ CT and MRI can define the extent of the disease.

Fine needle aspiration of a plasmacytoma may be non-diagnostic because of the limited tissue available for special staining and for complete histologic examination. Therefore, incisional or excisional biopsy depending on the size and location of the mass is necessary. Histologically, plasmacytomas are characterized by a diffuse or sheet-like proliferation of plasma cells with varying degrees of maturity and atypia. The nuclei are oval to round and eccentrically located with a dispersed ("clock-face") nuclear chromatin pattern and a clear or halo area.¹²

Whilst there is considerable overlap in the radiological appearances, consideration of patient's age, sex, predilection for an anatomical site, the presence of calcification and presence or absence of the expansion of the affected bones are the most important factors in suggesting the correct diagnosis. It is important to distinguish MM from EMP or SPB because the treatment and prognosis are different. Evaluation for plasma cell neoplasm includes total body skeletal survey using plain radiographs, serum and urine electrophoresis complete blood count, calcium level and bone marrow biopsy. The diagnosis of EMP or SPB is

made by ruling out MM and with presence of a single plasmacytoma in soft tissue or bone, respectively.

The potential of development of multiple myeloma is higher in solitary plasmacytoma of bone than in extramedullary plasmacytoma.¹⁴

On radiology, differential diagnosis of a large soft tissue mass with bone destruction in the absence of gross expansion of the destroyed bone includes lymphoma, metastasis, adenoid cystic carcinoma, chondrosarcoma and osteosarcoma. Presence of calcification in the mass should suggest the possibility of chondrosarcoma, osteosarcoma and chordoma. The differential diagnosis of a clival lesion includes chordoma, chondrosarcoma and meningioma. The differential diagnosis of lesion of petrous apex, parasellar region includes chondrosarcoma, osteosarcoma & Ewing's Sarcoma. On CT and MRI an extramedullary plasmacytoma is indistinguishable from a solitary lytic metastasis.¹¹

The potential for malignant systemic progression (multiple myeloma) is higher for solitary plasmacytomas of bone than for extramedullary plasmacytomas.^{2,14} Plasma cell neoplasms are highly radiosensitive. Local irradiation is the primary mode of treatment for extramedullary plasmacytomas, occasionally followed by surgical resection of the residual tumour. When extramedullary plasmacytoma with multiple myeloma is diagnosed, local treatment of the plasmacytoma should be followed by the systemic combination chemotherapy.²

Plasma cell neoplasms should be considered in the differential diagnosis of skull base tumours associated with cranial nerve palsy. This case again highlights the importance of obtaining an adequate biopsy to determine the appropriate treatment as excellent clinical and radiological results can be obtained with radiation of plasma cell tumours of the skull base.

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