

"Imaging Features Of Extramedullary Plasmacytoma Of Skull Base With Multiple Myeloma"- A Rare Case

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Abstract

Imaging features of extramedullary plasmacytoma of skull base with multiple myeloma is reported in a 65 year old woman patient presented with multiple cranial nerve palsies. Contrast Enhanced CT revealed a large homogeneously enhancing mass in the left parapharyngeal region with extension in left petrous apex, left cavernous sinus and upper two cervical vertebrae. An extramedullary plasmacytoma associated with multiple myeloma was diagnosed after biopsy from the mass and laboratory investigations.

Ind J Radiol Imag 2006 16:1:29-32

Key words : - Multiple myeloma, Extramedullary plasmacytoma, Computed Tomography, Skull base.

INTRODUCTION

Extramedullary plasmacytomas are rare lesions to diagnose, 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 generalized disease. Extramedullary plasmacytomas occurring as solitary (primary) tumors or secondary manifestations of multiple myeloma most often involve the upper airways and paranasal sinuses. Skull base is one of the rarest sites of extramedullary plasmacytoma. The imaging findings associated with this rare site of extramedullary plasmacytoma involvement are reported.

CASE REPORT

A 65-year-old woman presented with a large mass in the left submandibular region in the neck with asymmetry of left side of face and diplopia for six months. On examination of neck, a single diffuse 6x10 cm sized firm, non-tender swelling was noted over left side of neck. Left sided multiple cranial nerve palsies (cranial nerves II, III, IV, VI, VII and VIII) were noted. Positive laboratory findings were moderate anemia, hypercalcemia and elevated ESR of 150-mm/1st hr.

Contrast-enhanced CT revealed large 7x12 cm sized homogeneously enhancing lobulated mass involving the base of the skull on left side with epicenter in left parapharyngeal space [Fig-1]. The majority of the mass

appeared to be extracranial. Laterally the lesion was extending in the parotid and masticator space. Invasion of pharyngeal mucosal space with contralateral shift of oropharynx was seen medially. Involvement of posterior cervical space and posterior paravertebral muscles was noted posteriorly. Superiorly the lesion involved the skull base- petrous apex and sphenoid bone on left side. Extra-axial intracranial extension of the lesion was noted in the sellar-parasellar region, causing complete encasement of the left cavernous sinus [Fig-2]. Inferiorly the lesion extended down to the level of hyoid bone. Invasion of carotid space with Encasement of carotid vessels and IJV is seen. Bone window images demonstrated destruction of multiple bones of skull base with non-sclerotic ill-defined margins. Clivus, petrous apex, sphenoid bone, lateral mass of C1, and the pedicle of C2 were involved on left side [Fig-2, 3]. Scanogram revealed multiple osteolytic lesions were noted in cranial vault with clivus and basiocciput as well as cervical vertebrae [Fig-4]. Extradural extension of the lesion was seen in the spinal canal at C2 level on the left side [Fig-1]. No areas of calcification within the mass, any bony expansion or sclerosis were noted.

Biopsy of neck mass showed scattered mononuclear cells with eccentrically placed nuclei and abundant basophilic cytoplasm with perinuclear halo. Binucleated and multinucleated cells were seen [Fig-5]. These features resembled those of mature and immature plasma cells consistent with a diagnosis of myeloma.

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Received 10 October 2005; Accepted 12 March 2006

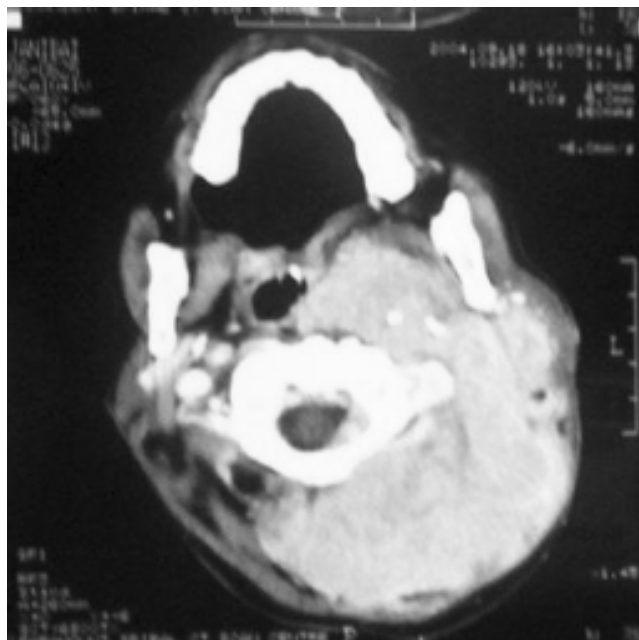


Figure 1: Contrast Enhanced CT image of the neck shows large, homogeneously enhancing mass involving the base of the skull on left side in parapharyngeal space. The lesion is involving the parotid and masticator space laterally, pharyngeal mucosal space with contralateral shift of oropharynx medially, posterior cervical space and posterior paravertebral muscles posteriorly, also invading the great vessels.

DISCUSSION

Plasma cell neoplasms include multiple myeloma, monoclonal gammopathies of unknown significance, plasmacytomas, and plasma cell leukemia [1]. Multiple myeloma is the most common of the plasma cell neoplasms. Whereas multiple myelomas represent systemic disease without the potential for cure, plasmacytomas represent local forms of plasma cell neoplasms [2]. Plasmacytoma is further classified into two groups: osseous [Solitary plasmacytoma of bone (SPB)] and non-osseous [Extramedullary plasmacytoma- (EMP)] primary lesions [3].

EMP and SPB each comprise < 4% of all plasma cell neoplasms [4, 5]. Solitary plasmacytoma of bone constitute approximately 70% of all plasmacytomas [3]. Extramedullary Plasmacytomas are four times more likely to occur in males than in females and 95% of tumors occur over the age of 40 yrs (mean age is 59 yrs) [6]. Multiple myeloma is a relatively rare cancer, which occurs predominantly in patients over 60 years of age [1]. Although EMP is known to frequently coexist in patients of MM at autopsy, clinically only 20% of the head and neck EMP present simultaneously with multiple myeloma [6, 7].

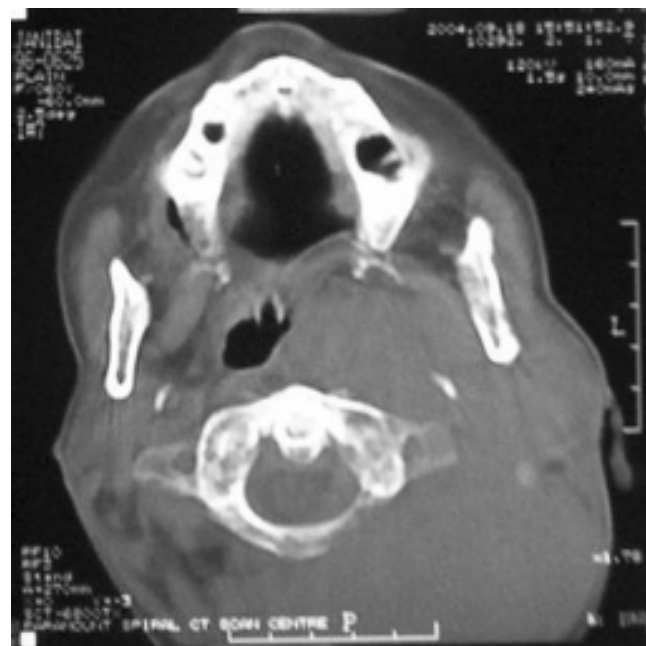
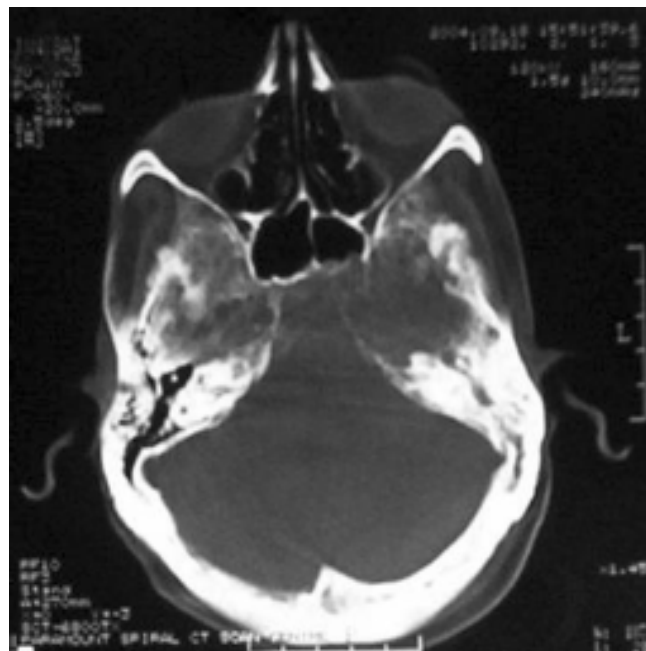


Figure 2 & 3: Bone window images demonstrates destruction of clivus, petrous apex, sphenoid bone, lateral mass of C1, and the pedicle of C2 on left side.

Plasmacytoma represent less than 1% of all head and neck tumors [4]. However, the majority (80%) of the EMP occur in the in the head and neck, especially the nasopharynx and the paranasal sinuses [3-6]. Rare cases of primary EMP have been described 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 [4, 5, 8]. When the skull is involved most occur in the calvarium and the skull base is rarely affected [9]. Local plasma cell neoplasms can involve skull base structures. They may occur at the clivus or petrous

apex where they represent a solitary plasmacytoma of bone, or they may originate within the submucosa of the sino-nasal and nasopharyngeal tracts where they represent an extramedullary plasmacytoma [2]. In the present case, the lesion involving the left petrous apex, clivus and sphenoid bone- and was part of disseminated disease, and multiple myeloma was diagnosed.



Figure 4: Scanogram of skull shows multiple lytic lesions involving vault and basiocciput.

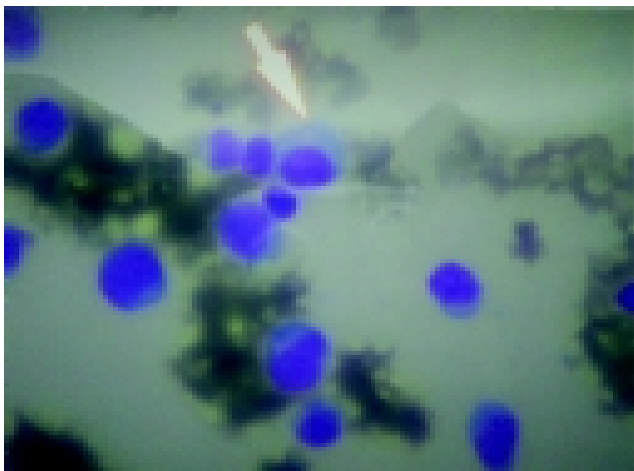


Figure 5: Photomicrograph of HPE of biopsy from the mass shows mature and immature plasma cells consistent with a diagnosis of myeloma.

Skull base lesions are usually asymptomatic, but larger tumors can cause symptoms [2]. Neurologic symptoms

due to plasmacytomas located either in the base of the skull or at intracranial locations are extremely rare [10]. The cause of the neuropathy is the direct compression of nerves or nerve groups either in their intracranial course, especially when there is involvement of the body of the sphenoid or the apex of the petrous bone, cavernous sinus or at the cranial outlets in the base of the skull [1, 10, 11]. 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, 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 [12].

On CT and MRI, plasmacytoma is of fairly homogeneous soft-tissue density (because of its cellularity) [13]. On CT plasmacytomas are well-demarcated tumors, occasionally aggressive with bone destruction and involvement of adjacent structures [14]. 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 [14]. CT and MRI can define the extent of the disease. The body of the sphenoid and the apex of the petrous bone are the most common sites of involvement of skull base plasmacytoma [12].

Whilst there is considerable overlap in the radiological appearances consideration of patients' age, sex, predilection for an anatomical site, and the presence of calcification, presence or absence of the expansion of the affected bones are the most important factors in suggesting the correct diagnosis.

Absence of expansion of the bone of the skull base differentiates extramedullary plasmacytoma from the plasmacytoma of bone. This distinction is important because the potential of development of multiple myeloma is higher in solitary plasmacytoma of bone than in extramedullary plasmacytoma [5]. Imaging 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 [14].

Though imaging features are non-specific, large homogeneous mass involving the skull base with moderate

to strong contrast enhancement, absence of calcification and clinical presentation of cranial neuropathy- possibility of extramedullary solitary plasmacytoma should always be kept as a differential diagnosis. When cranial nerve palsies are associated with lesions in cervical vertebral bodies or the petrous bone, a solitary plasmacytoma should be considered in the differential diagnosis.

The potential for malignant systemic progression is higher for solitary plasmacytomas of bone than for extramedullary plasmacytomas [2, 5]. 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 tumor. When extramedullary plasmacytoma with multiple myeloma is diagnosed, local treatment of the plasmacytoma should be followed by the systemic combination chemotherapy [2].

Five year survival rate of extramedullary plasmacytoma is 31%-75% [6]. The prognosis of extramedullary plasmacytoma with multiple myeloma is poor, and most patients die within 2 years of their diagnosis; 3-year survival is only about 10% [4, 6].

CONCLUSION

The imaging features of the skull base plasmacytoma; a rare location and its differential diagnosis- are presented which may aid our understanding of the disease. Plasma cell neoplasms should be considered in the differential diagnosis of skull base tumors associated with cranial nerve palsy. This case again highlights the importance of obtaining a biopsy of skull base tumors to determine the appropriate treatment as excellent clinical and radiological results can be obtained with radiation of plasma cell tumors of the skull base. Preoperative biopsy will obviate the need of an extensive surgery of this difficult anatomical region.

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