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Skull Base Primary Solitary Plasmacytoma: A Case Report

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ABSTRACT

Primary plasmacytomas are rare and not many cases have been reported in literature. These are characterized by solitary bony lesions, neoplastic plasma cells, less than 5 % plasma cells in the bone marrow, less than 2.0gm/dL monoclonal protein in serum and a negative Bence Jones test for urine. These primary plasmacytomas involving the skull base are rare, as compared to surface bones of the skull and only 4-5 cases have been reported.

We report a primary plasmacytoma involving the skull base producing lower cranial nerve deficit and trigeminal neuralgia.

Keywords: plasmacytoma, primary, skull base, neuralgia.

INTRODUCTION

54 year old right handed male presented with complaints of pain over right ear, radiating to the upper and middle part of face, which used to get aggravated, during mastication. He also complained of ringing sensation in the right ear, giddiness, and double vision, which was more on gazing towards the right side. The tinnitus was followed by decreased hearing in the right ear.

On examination, patient was well built, higher functions were normal. Cranial nerves examination suggested, sensory involvement of right V2 and V3 and motor involvement of right sixth nerve and Rinne's test showed that air and bone conduction was affected on right side. Hence, clinically patient had involvement of 5th, 6th and 8th nerve on the right side. Motor and sensory systems were normal.

MRI revealed a large, 6.7cm(AP) x 3.8cm(sup-inf) x 4.7cm sized, irregular shaped, midline altered signal intensity space occupying lesion involving the clivus, central and adjacent portion of posterior skull base, with total destruction of basisphenoid, more on right side. It also involved the right internal acoustic meatus with displacement and distortion of right 7th and 8th nerve complex, with effacement of pre-pontine and pre-medullary cistern. The mass was noted to extend into anterior skull base involving right posterior ethmoid sinus; and also to the apex of right orbit. It was also noted to involve cavernous sinuses on either side with encasement of both internal carotid arteries which showed normal flow voids. The SOL was isointense to hypointense on T1, hypointense on T2 and isointense on FLAIR images. It showed homogemous strong enhancement after intravenous gadolinium contrast administration. No areas of cyst or

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hemorrhage were noted. Brain parenchyma including the pituitary gland was normal.

Patient was operated via right extra-temporal approach and tumor was noted to involve the temporal skull base extending into clivus, and infiltrating the 5th, 6th cranial nerves of the same side including the Gasserian ganglion. The tumor was excised with CUSA and complete decompression of 5th and 6th nerves was achieved. Gross appearance of the tumor was fleshy with areas of vascularity.

Histopathology revealed a highly cellular tumor composed of discohessive sheets of plsma cells of varying degree of maturation ranging from immature to mature. The mature cells had an eccentric pyknotic nuclei and moderate eosinophillic cytoplasm. The immature cells had open vesicular nuclei with prominent nucleoli. Mitotic activity was low and vascularity was observed; thus features suggestive of Plasmacytoma.

Post-operative period was uneventful and Trigeminal neuralgia subsided completely, 6th nerve paresis also started to recover.

The urine examination revealed absence of bence-jones proteins. Serul Immunoglobulin electrophoresis suggested absence of M-band. Serum examination for CEA and á-fetoprotein was negative.

Bone marrow biopsy revealed no abnormal plasma cells. As all the above work up for systemic myelomatosis was negative, diagnosis of primary skull base plasmacytoma was made.

Patient has received post-op adjuvant radiotherapy of 200 cGy per day for 25 days.

DISCUSSION

Solitary plasmacytomas of bone tend to disseminate or progress to multiple myeloma even as long as 7 to 23 years after presentation. Skull base plasmacytomas have a less favourable prognosis as compared to skull vault plasmacytomas, especially when there is no evidence f systemic myelomatosis[1]. Plasmacytomas are four times more likely to occur in males than in females and 95% of tumors occur over the age of 40 years (mean age of 59 years)[2,3]. They represent less than 1% of head and neck tumors[4]. A Pancholi et al[3] reported a case of plasmacytoma of skull base with multiple myeloma which involved the neck and skull bone along with multiple cranial nerves. Our case did not have any evidence of multiple myeloma, and there was no external swelling seen in the neck. When the skull is involved, most occur in the calvarium, and skull base is rarely affected[5]. Skull base lesions are usually asymptomatic, but larger tumors can cause symptoms. Neurological symptoms are the result of direct compression of nerves or nerve groups either in their intracranial course or at the cranial base outlets; the most common involvement being 6th, 2nd, 5th, 7th and 8th, in that order[6,7,8]. Our patient had involvement of 5th, 6th and 8th nerves involvement on right side along with giddiness.

Figure 1: Hematoxyllin-Eosin stain.

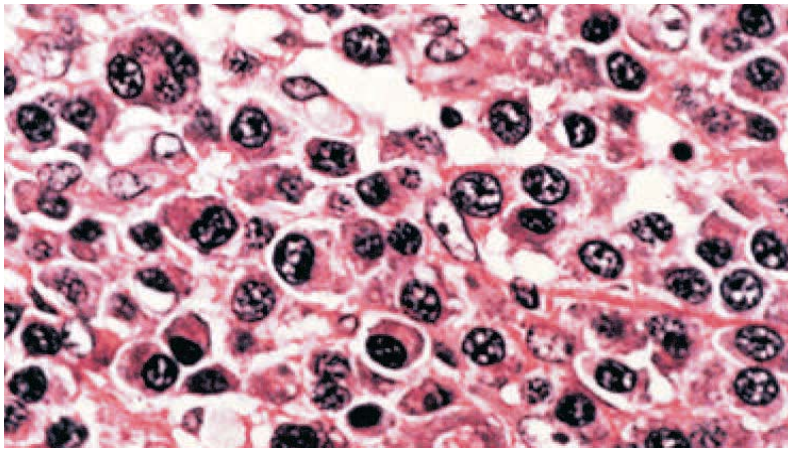


Figure 2: Contrast Enhanced CT Scan-Axial.

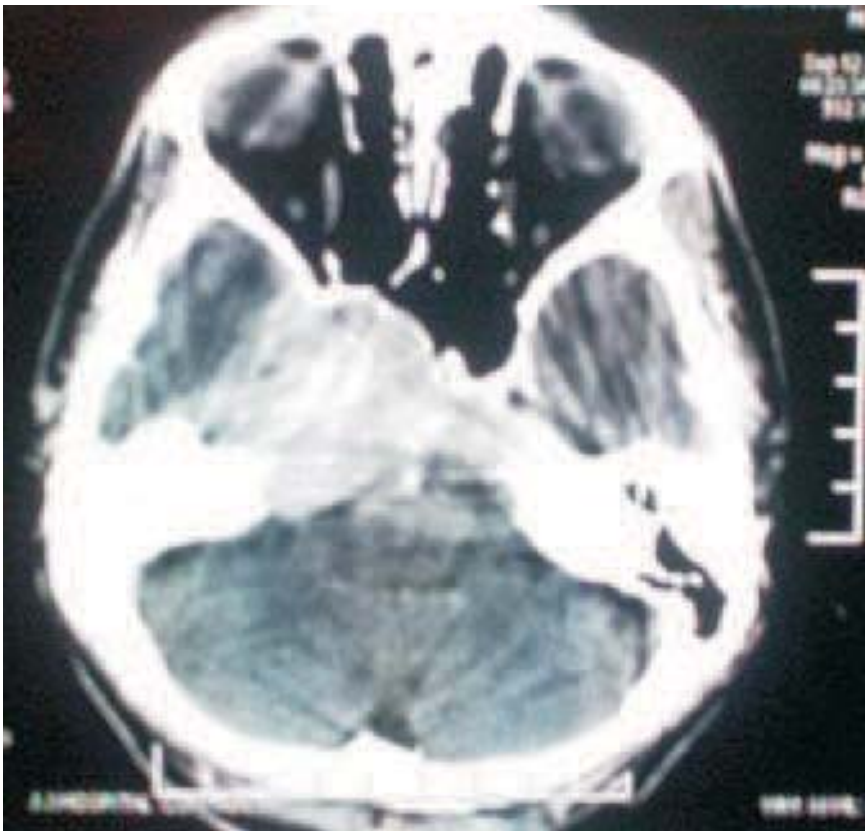


Figure 3: Contrast enhanced CT scan-Coronal.

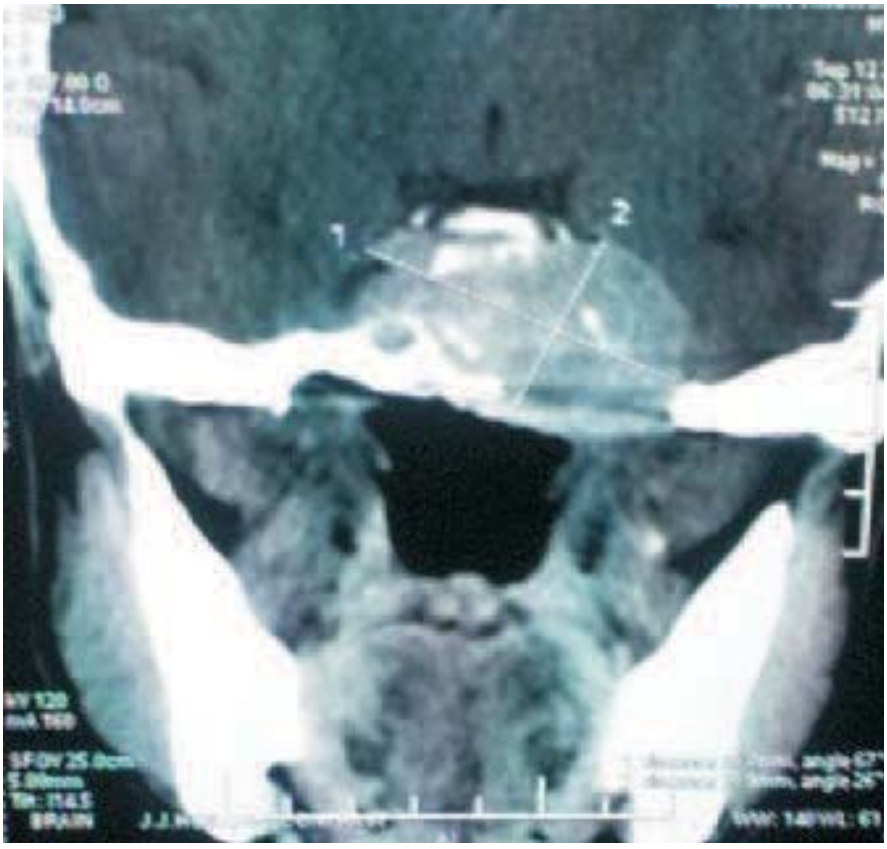


Figure 4: CT Scan : Bone Cuts showing osteolysis in the region of tumor

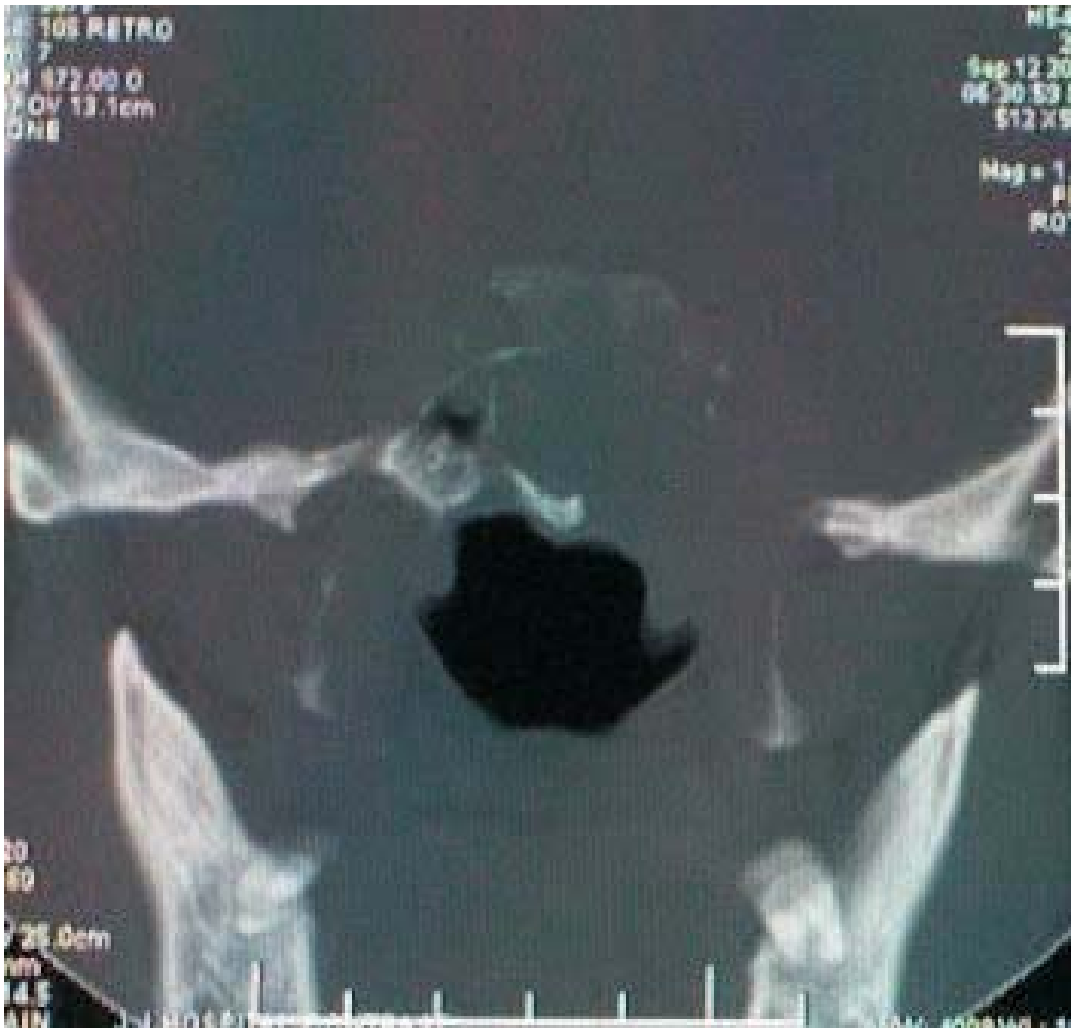
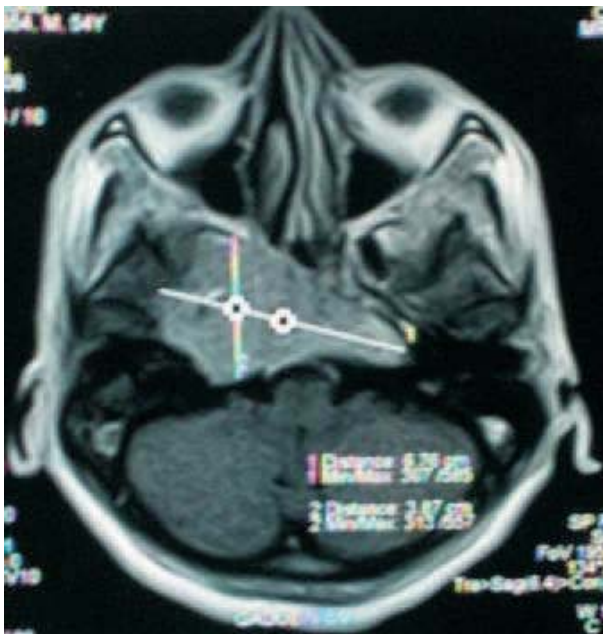


Figure 5: MRI images: Gadolinium enhanced.



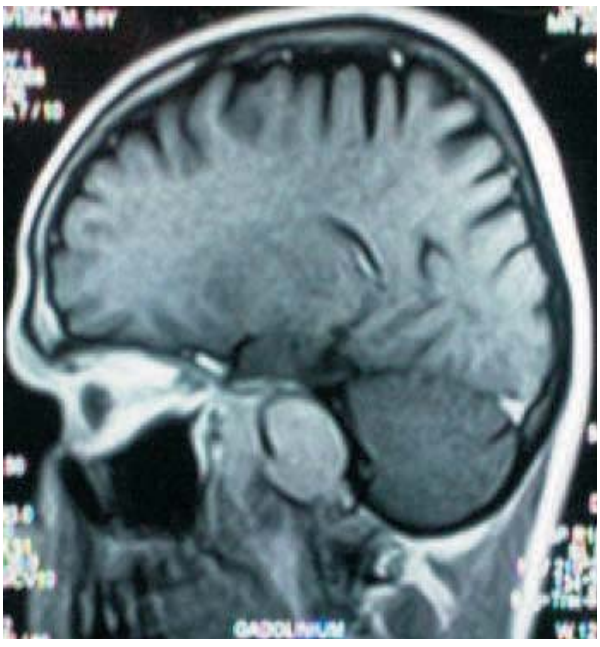
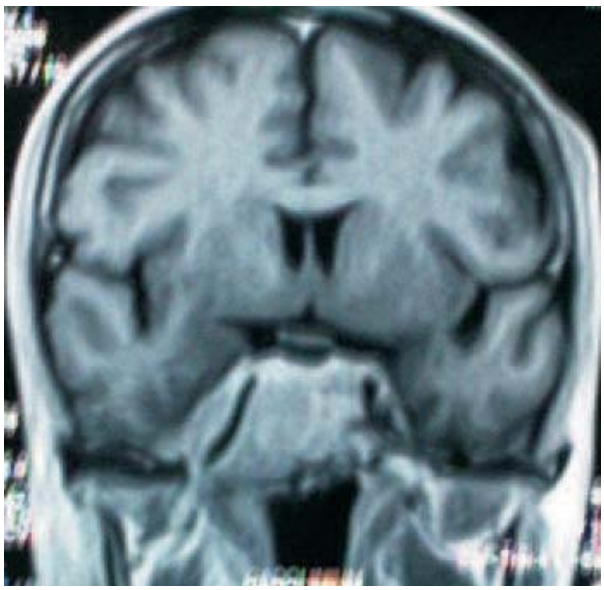
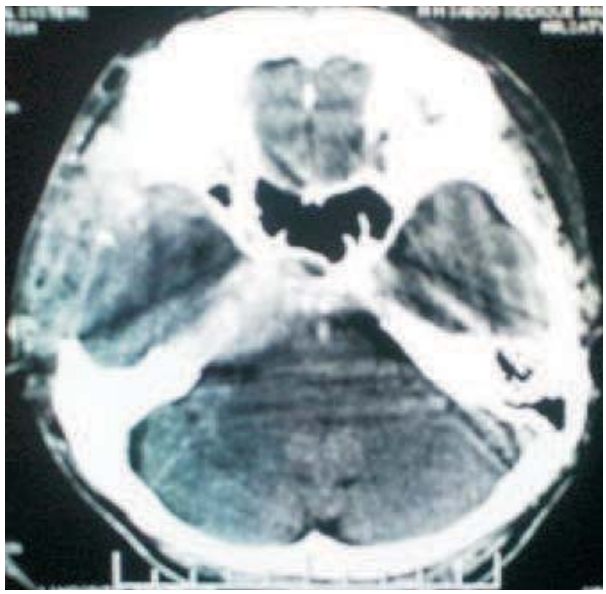


Figure 6: Post-op Contrast Enhance CT scan.





Skull base plasmacytomas may involve PICA, leading to lateral medullary syndrome[9].

CT scan findings of solitary plasmacytoma are of osteolytic lesion without a sclerotic rim, hyperdense tumor, and homogenous post-contrast enhancement[10,11,12,13].

MR features are isointense, hyper or heterogenous intensity on T1W, and homogenous enhancement on Gd-DTPA[11,12,13,14].

Surgical removal followed by radiotherapy is the best treatment at the moment[13,15]. Our patient has undergone surgery and is receiving radiotherapy at the moment.

REFERENCES

1. Tanaka M, Soichiro S, Nomura K, Nakanishi Y. Solitary Plasmacytoma of the Skull: a Case Report. Japanese Journal of Clinical Oncology, 1998. 626-630.
2. Som PM, Brandwein MS. Tumors and tumor-like conditions. In: Som PM, Curtin HD, eds. Head and Neck Imaging. Vol. 1, 4th ed. St. Louis: Mosby. 2003: 261-373.
3. A Pancholi, S Raniga, P A Vohra, V Vaidya, A Prajapati, S Mansingani. Imaging Features Of Extramedullary Plasmacytoma Of Skull Base With Multiple Myeloma- A Rare Case . Ind J Radiol Imag 2006 16:1:29-32.
4. Nofsinger YC, Mirza N, Rowan PT, Lanza D, Weinstein G. Head and neck manifestations of plasma cell neoplasms. Laryngoscope. 1997; 107:741-746.
5. Okamoto K, ITO J. Solitary Plasmacytoma of the Occipital Bone: A Report of Two Cases. European Radiology 1997; 7(4): 503-6.
6. Pollard JD, Young GAR. Neurology and the bone marrow. J Neurol Neurosurg Psychiatry 1997; 63:706-717.
7. Montalban C, Martín-Aresti J, Patier JL. Intracranial Plasmacytoma with Cranial Nerve Neuropathy in Multiple Myeloma. Journal of Clinical Oncology 2005; 23:233-235.
8. Spaar FW. Paraproteinaemias and multiple myeloma. In: Vinken PJ, et al, eds. Handbook of clinical neurology. Amsterdam: North Holland, 1980; 131-181.
9. Mill WB, Griffith R. The role of radiation therapy in the management of plasma cell tumors. Cancer. 1980; 45:647-652.
10. Jakubowski J, Kendall BE, Symon L. Primary plasmacytoma of the cranial vault. Acta Neurochir (Wien) 1980; 55:117-34.
11. Marais J,Brookes G, Lee CC. Solitary plasmacytoma of the skull base. Ann Otol Rhinol Laryngol 1992; 101:665-8.
12. Prasad ML, Mapapatra AK, Kumar L, Khosla A, Sarkar C, Prasad,A, et al. Solitary intracranial

plasmacytoma of the skull base. Indian J Cancer 1994; 31:174-9.

13. Bindal AK, Bindal RK, van Loveren H, Sawaya R. Management of intracranial plasmacytoma. J Neurosurg 1995; 83:218-21.
14. George JC, Caldemeyer KS, Kreipke DL, Chalian AA, Moran CC. Solitary plasmacytoma of the mastoid bone presenting as coalescent mastoiditis. Arch Otolaryngol Head Neck Surg 1994; 120:1393-4.
15. Du Preez JH, Branca EP. Plasmacytoma of the skull: case reports. Neurosurgery 1991; 29:902-6.