# Solitary Plasmacytoma of Skull in 38 Year Old Woman A Rare Case

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## Abstract

**Solitary plasmacytoma of bone (SPB) is very rare and accounts for only 4% of all plasma cell tumours.[1,2] It has significant higher risk for progression to multiple myeloma. We describe a case of SPB in 38 year old woman (Median Age is 55 years and male preponderance) with 4 months history of painless swelling in right parietal region of skull. Clinical examination was normal and investigations revealed radiological solitary lesion with plasma cells on biopsy of the lesion. On further evaluation Immunoelectrophoresis, bone marrow aspiration, serum calcium, urine for Bence Jones proteins were all normal with Immunohistochemistry positive for CD 138 and Lambda light chains which confirmed our diagnosis. The level of β2 microglobulin was raised (2.11) mg/dl.**


**Key Words: Solitary Plasmacytoma, B2 Microglobulin, Protein electrophoresis**

## Introduction

Solitary plasmacytoma is an infrequent plasma cell neoplasm with no evidence of systemic multiple myeloma. Commonly SPB occurs in the axial skeleton whereas extra medullary plasmacytoma (EMP) occurs in the head and neck region. The ratio of SPB in males to females is 2:1 and median age of patients is 55 years.[3] SPB has significantly higher risk for progression to myeloma in comparison to EMP and progression is dependent on factors like lesion size/age/presence of spine lesions/M protein levels pre and post treatment/ radio therapy dosage given. Biopsy conformation of a monoclonal plasma cell infiltration from single site is required for diagnosis.

## Case Report

A 38 year old woman first noticed a soft progressive painless swelling of 4 X 4 cm in right parietal region of skull since 4 months. There were no complaints of backache, fever, burning micturition, lethargy, weakness, confusion, joint pains, headache or visual disturbances. Bladder and bowel control was normal. There was no history of trauma. History for seizure disorder was negative. On examination the swelling was fixed to underlying skull bone but the overlying skin was free and normal. There was no pallor and patient was anicteric. There was

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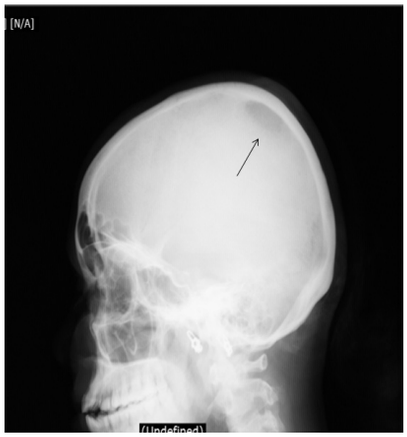

Fig 1. X Ray Skull - Solitary Punched out Lesion

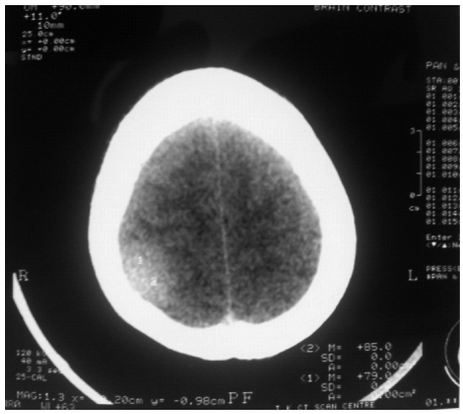

Fig 2. NECT HEAD - hyper dense lesion in high parietal convexity

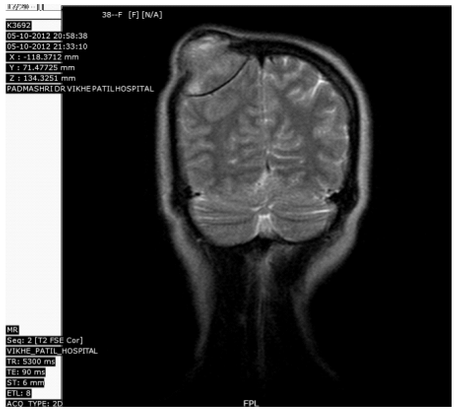

Fig 3.T2 FSE Coronal Section

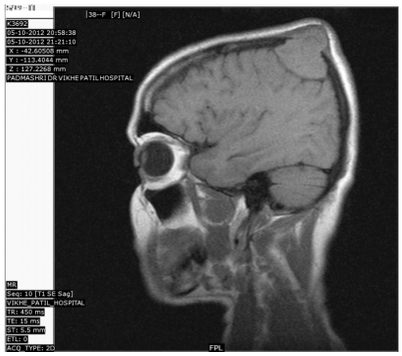

Fig 4.T1 SE Sagittal Section

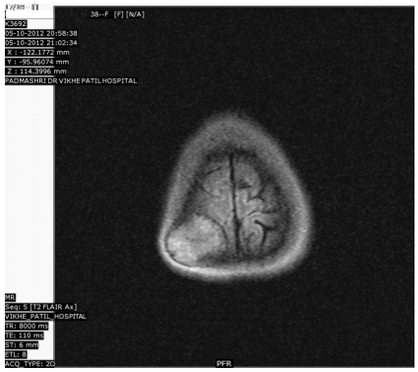

Fig 5.T2 FLAIRAxial Section

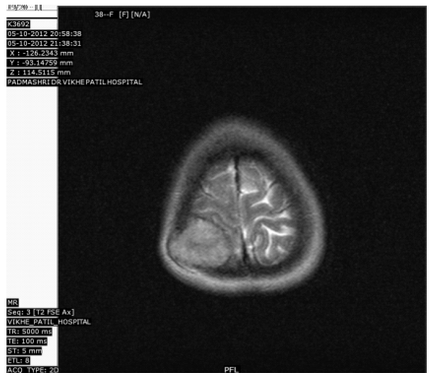

Fig 6.T2 FSE Axial Section

no lymphadenopathy, no skin rash, and no hyper-pigmentation. Her BP was 126/80 mm of Hg, temperature was normal. Her systemic examination was normal.

## Investigations

Laboratory investigations revealed haemoglobin -10.5 gm/dl, white blood cell count- 6600/mm$^3$, platelet count -2,00,000/mm$^3$, total serum protein-7 gm/dl, albumin-4 gm/dl, globulin-3 gm/dl and serum calcium-9.2 mg/dl. Peripheral blood smear and serum electrolytes were normal. Renal function and liver function tests were normal. Urine routine showed no albumin and no sugar. Urine test for Bence Jones proteins was negative. Serum protein electrophoresis showed normal values of albumin (3.6 gm/dl), alpha 1 globulin (0.1 gm/dl), alpha 2 globulin (0.9 gm/dl), beta 1 globulin (0.8 gm/dl), gamma globulin (1.4 gm/dl). Beta 2 micro globulin was 2.11 mg/dl. Urine protein electrophoresis and Immunofixation were normal. Bone marrow aspiration showed normo cellular marrow with minimal changes of micro normoblastic maturation with plasma cells 6%, adequate in number without any morphological abnormality. Patient's X ray skull (Fig 1) was suggestive of solitary punched out lesion. Computed tomography (NCCT) brain (Fig.2) showed evidence of well-defined hyper dense lesion in high parietal convexity with erosion of the adjacent parietal bone. Magnetic resonance (MR) brain (Fig.3-6) showed a large well defined, lobulated extra axial solid mass lesion measuring (4.5 X 3.5 X 3.3 cm) along right parietal convexity inferiorly indenting brain parenchyma and superiorly causing significant scalloping of overlying skull vault extending deep to scalp through vault defect measuring (2.5 X 2.1cm).

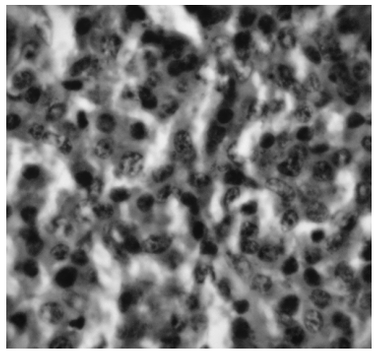

Fig 7. Microphotograph shows sheets of plasma cells with eccentric cytoplasm and cart wheel chromatin

Histopathology of resected bony tissue was suggestive of plasmacytoma (Fig.7). Sections from the bony tissue showed bony trabeculae with intervening cellular pro-

significant anisonucleosis or mitosis. Immunohistochemistry of the resected bony tissue was suggestive of multiple neoplastic cells strongly positive for CD 138 and lambda light chain and negative for cytokeratin and kappa light chain. Thus in view of clinical suspicion and with aid of laboratory and radiological investigations our diagnosis was confirmed and possibility of Multiple Myeloma was ruled out.

## Differential Diagnosis

Meningioma (Clinico-radiological), plasma cell granuloma, plasmacytoid lymphoma, large cell lymphoma of immunobastic type (histo-pathological).

## Treatment

Patient and relatives were explained the nature of the disease, its prognosis and outcome. Tumour was resected completely. There were no intra operative and post operative complications. In view of radiosensitive nature of the tumour radiotherapy was given.

## Images

Fig 3,4,5,6 - Large well defined mildly hyper intense on T2W images and isointense on T1W images, lobulated extra axial solid mass lesion along right parietal convexity inferiorly indenting adjacent brain parenchyma and superiorly causing significant scalloping of overlying skull vault extending deep to scalp through vault defect.

## Discussion

Plasmacytoma is a clonal neoplastic disorder of bone marrow originating from plasma cells. It may manifest as a spectrum of three different diseases like multiple myeloma (systemic disease), EMP and SPB. SPB may be the first manifestation of a following multiple myeloma. Clonal plasma cells involved in plasmacytoma produce monoclonal immunoglobulins as well as kappa or lambda light chains and hence their quantitative assays may be performed for diagnosis and to analyse tumour growth.

Typical characteristics of SPB include presence of radiological solitary bone lesion, biopsy of the lesion showing neoplastic plasma cells, bone marrow aspiration showing less than 10% plasma cells, negative Bence Jones protein in urine, lack of CRAB (increased serum calcium, renal insufficiency, anaemia, multiple bone lesions), low M component < 2 gm/dl, no osteoblastic response to bone destruction on skeletal survey.

The β2 microglobulin is usually normal in SPB. β2 microglobulin level is a prognostic indicator for development of SBP into MM. Studies suggest that prognostic significance increases when in addition to β2 microglobulin levels, other parameters like Hb%, RNA and DNA indices, thymidine kinase activity, serum albumin are also taken into consideration.

Progression of SPB to multiple myeloma can occur at a rate of 65 to 84% in 10 years and 65 to 100% in 15 years. In spite of definitive therapy given to cases of SPB median time of progression to multiple myeloma is 2 to 3 years. EMP in comparison has 10 year overall survival rate of 70% and thus has a good prognosis and approximately 65% of the patients have no recurrence and do not progress to multiple myeloma.

SPB expresses angiogenic factors, vascular endothelial growth factor and basic fibroblast growth factor and in accordance with the high vascularity of the tumour. Syndecan-1 may be an immune histochemical marker of solitary plasmacytoma of the skull, but this expression is not well characterised.[4,5] Syndecan-1 is a member of family of cell surface transmembrane heparan sulphate proteoglycan that can bind extra cellular matrix components and has an important function in biology of plasma cells. Treatment modalities for SPB include surgical resection of the tumour, radiotherapy and chemotherapy.

Review of literature reveals that total surgical resection followed by radiotherapy is an effective treatment in majority of SPB of skull. Some studies also suggest that if total resection has been achieved then radiotherapy should be reserved in case of tumour recurrence. But as SPB of skull is a highly radiosensitive tumour radiotherapy is given in most cases in a dose varying from 30-50 Gy (No current guidelines are available for the selection of dose) and thus recent reports emphasize complete surgical resection with radiotherapy as an acceptable treatment for SPB skull.[6] Adjuvant chemotherapy has no beneficial effect on disease control but certain studies suggest that this may delay the progression of SPB to multiple myeloma.

## Follow up and outpatient care

M protein measurement and complete blood count should be done every 6 weeks for the first 6 months. The amount of M protein is not a precise prognostic indicator but persistence of M Protein after treatment is a predictor for progression of SPB to multiple myeloma. A solitary plasmacytoma of bone tends to disseminate or progress to multiple myeloma, even as long as 7-23 years after its presentation.[7]

## Conclusion

If detected and treated early, progression of SBP to fatal neoplasm like multiple myeloma where the prog-

nosis is not as good as SBP can be prevented.[8] There is no single prognostic marker to predict the development of multiple myeloma in cases of SBP. There are no standard guide lines for choice of treatment in a case of SBP. In view of radiosensitive nature of tumour, radiotherapy should be given to patients of SBP, but there is no consensus on the dose of radiotherapy to be given.


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