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Case Report

Amyloidogenic occipital mass obscuring plasmacytoma: a rare presentation and review of literature

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ABSTRACT

Solitary plasmacytoma of bone are a spectrum of plasma cell disorder which are defined by presence of clonal plasma cell in the bone including skull. The occurrence of true plasmacytoma of skull with large amyloid deposits in young patient is a rare presentation. We present case of 25-year-old man with a 3-month history of a slowly growing mass in the occipital region associated with headache. Neurological examination was within normal limits. Magnetic resonance imaging revealed a large extra-axial mass, the inner table of the skull were partially destroyed by the tumour, but the dura was not involved. The tumour removed and the skull defect was reconstructed followed by radiotherapy. Histopathological examination confirmed plasmacytoma with amyloidosis, positive for amyloid P component on immunohistochemistry. Laboratory investigations revealed no systemic myelomatosis. After 2 years of close follow up patient had no signs of recurrence or progression to multiple myeloma. The different management stratagies for the two entities of plasma cell disorder spectrum i.e. solitary plasmacytoma of bone and multiple myeloma necessitates there differentiation by thorough clinical, radiological and hematological studies. Surgical treatment followed by radiotherapy is an effective treatment option.

Keywords: Amyloid, Amyloid P, Plasma cell disorder, Solitary plasmacytoma, Solitary plasmacytoma of bone

INTRODUCTION

Solitary plasmacytoma of bone (SPB) are solitary lesion of bone with evidence of clonal plasma cells, normal bone marrow with no evidence of clonal plasma cells and normal skeletal survey and absence of end-organ damage.[1] They present in>40years of age, only 2-3% cases are younger than 30 years.[2] We report a case of solitary plasmacytoma of occiput with amyloidosis in 25-year-old male presenting with recurrent headache. Magnetic resonance imaging revealing a large extra-axial lobulated mass lesion of occipital bone. The lesion was excised and patient underwent radiotherapy with good response, no evidence of recurrence or multiple myeloma after 2 years of follow up. SPB should be distinguished from multiple myeloma due to different treatment guidelines, SPB is a radiosensitive tumor with good response. Skull plasmacytoma with amyloidosis occurring in less than 30 years is a rare presentation and may easily be missed on histology if massive amyloid deposits are evident that tend to obscure the underlying plasmacytoma, hence careful search for underlying plasma cell dyscrasia should be performed.

CASE REPORT

A 25-year-old male patient presented with a 3-month history of occipital swelling associated with mild to moderate headache. No neurological abnormality was evident. Haemoglobin, blood counts, blood urea, serum

creatinine and electrolytes including serum calcium were within normal limits. Magnetic resonance imaging (MRI) of the brain with contrast demonstrated a well-defined heterogeneously enhancing extra-axial lobulated lesion measuring 85×87×70 mm arising from the left half of occipital bone, heterogeneously hyperintense on T2/FLAIR and isointense on T1 WI (Figure 1 A, B).

The lesion was destroying the inner table occipital bone, compressing and buckling the left cerebellum with evidence of CSF cleft. Mass effect seen in form of compression of 4th ventricle and dilatation of 3rd ventricle. MR angiography was also performed showing no evidence of arterial aneurysm or vascular malformation; however, few vascular channels were seen supplying the extra-axial lesion. Skeletal survey of the body revealed no other lytic lesions anywhere in the body.

A subtotal excision of mass was performed and sent for frozen section with clinical diagnosis of meningioma, a frozen section revealed a cellular neoplasm, with possibility of round cell neoplasm was rendered, amyloid deposits were not appreciated and plasmacytoid morphology was hampered due to crushing artifact (Figure 2A). Histopathological study demonstrated tumor tissue composed of amorphous pink material with few interspersed plasma cells invading the surrounding adipose tissue and skeletal muscle (Figure 2B-D). The cells are large with central to eccentric nuclei, coarse chromatin, few with prominent nucleoli and moderate cytoplasm. Few binuceated forms were also seen along with fair number of giant cells.

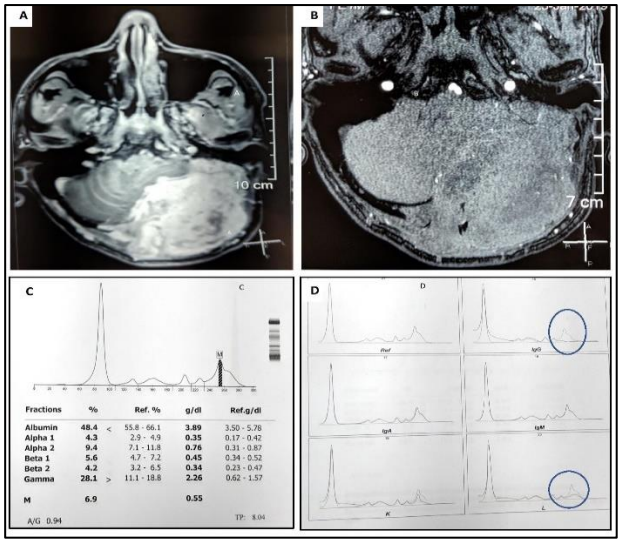

Figure 1: Magnetic resonance imaging (MRI) of the brain with contrast showing extra-axial lobulated mass, heterogeneously hyperintense on T2/FLAIR (A) and isointense on T1 WI(B). Serum electrophoresis revealed M band in gamma region (0.55g/dl)(C), immunofixation showing IgG lambda type of M-band in gamma region(D).

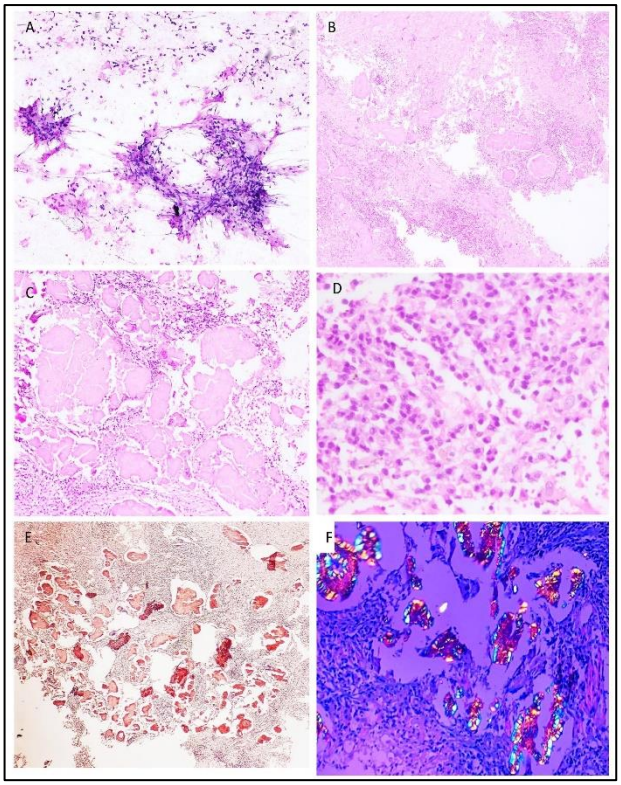

Figure 2: Histopathology findings. Crush smear showing clusters of cells with marked crushing, (A) tumor tissue composed of amorphous pink material with few interspersed plasma cells and fair number of giant cells. (B-D) The amorphous pink material positive for congo red (E) with apple green birefringence on polarising microscopy (F). (Hematoxylin-eosin; original magnification 50x (A, B), 100x (C), 200x(D-F)).

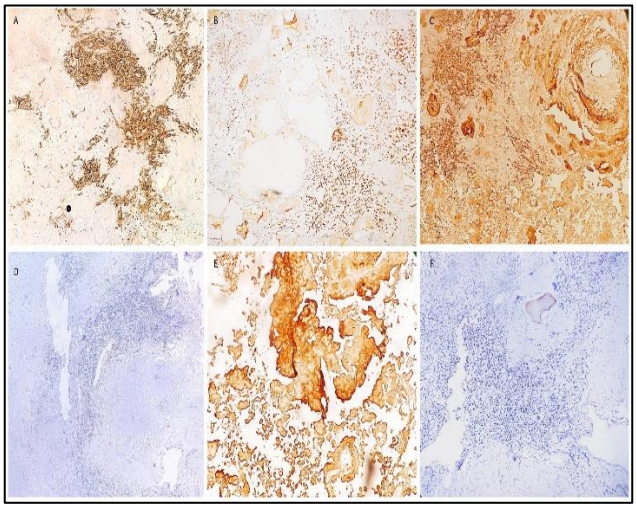

Figure 3: Immunohistochemical findings: Tumor showing diffuse expression for CD 138, MUM-1, Lambda(A-C), loss of expression for lambda (D), positive expression for amyloid P(E) and negative for amyloid AA(F), [Original magnification DAB 100x (A–F)].

The amorphous pink material was positive for congo red (Figure 2E-F). For definite diagnosis immunohistochemical (IHC) staining was performed, tumor showed expression for CD138, MUM-1, lambda and amyloid P (Figure 3A-C, E) with negative expression for kappa (Figure 3D), Amyloid AA (Figure 3F) leukocyte common antigen (LCA), Tdt, synaptophysin, chromogranin, myogenin and Sal- like protein-4 (SALL-4) a final diagnosis of solitary plasmacytoma with primary amyloidosis was rendered. Serum electrophoresis showed M band in gamma region (0.55 g/dl) followed by immunofixation showing IgG lambda type of M-band in gamma region (Figure 1C-D). Urinalysis for Bence-Jones proteins showed no abnormal findings. Bone marrow examination was within normal limits. The patient underwent external beam radiation therapy, a radiation dose of 50 Gy was given in 25 fractions over 5 weeks and patient was kept on regular follow, after two years patient has no signs of recurrence or progression with negative serum immunofixation and<5% plasma cell in bone marrow examination.

Table 1: Brief review of solitary plasmacytoma of skull.

Study	Age/gender	Site	Size (mm)	Amyloid	Treatment	Follow up	Recurrence
Barone et al[10]	55, F	Frontal	90×80	Absent	Gross total resection	9 months	No
Nakamoto et al[11]	56, M	Temporoparietal	60×40	Absent	Gross total resection+ RT (40Gy)	NA	NA
Okamoto et al[12]	72, F	Occipital	NA	Absent	Gross total resection	4 years	No
	64, M	Occipital	60×90	Absent	Gross total resection	8 months	No
Matsuda et al,[13]	55, F	Temporal	70×50	Absent	Gross total resection+RT (50Gy)	2 years	No
Tanaka et al[14]	55, M	Frontal	80×80	Absent	Gross total resection+RT (50Gy)	7 months	No
Singh et al[15]	38/F	Petrous bone	50×35	Absent	RT+Chemotherapy (thalidomide, dexamethasone)	3 months	No
	42/M	Middle cranial fossa, orbit	NA	Absent	RT+chemotherapy (melphalan, prednisolone)	4 years	No
	66/F	Left parietal region	5545	Absent	Surgery		Died of disease
Zigouris et al[16]	78, F	Fronto-temporo-parietal	98×80	Absent	Gross total resection	1 year	No
Bakar et al[17]	49, M	Frontal	90×85	Absent	Gross total excision		Died of disease
Gürbüz et al[18]	63, M	Parietooccipital	NA	Absent	Gross total resection+RT	NA	No
Dong et al[19]	70, M	Frontoparietal	60×50	Absent	Gross total resection+RT	2years	No
	75, F	Parietal	30×30	Absent	Gross total resection+RT	18 months	No
Singh et al[20]	25, M	Parietal	80×79	Present	NA	NA	NA
Mankotia et al[21]	36, M	Frontal	180×130	Absent	Gross total resection+RT	3 months	No
Yang et al[22]	42, F	Occipital	60×50	Absent	Surgery+Chemotherapy (VPDT)	2 years	Died of disease
Kuo et al[23]	40, M	Parietooccipital	NA	Absent	Gross total resection	1 year	No
Chen et al[24]	67, M	Temporoparietal	65×60	Present	Chemotherapy followed by RT (5400cGy)	25 months	Pathologic fracture humerus and died of pulmonary infection
Munjal et al[25]	63, F	Frontoparietal parasagittal	103×80	Absent	Embolization followed by surgery	NA	NA
Present case	25, M	Occipital	87×85	Present	Partial resection+RT (50Gy)	2 years	No

NA: Not available

DISCUSSION

Solitary plasmacytoma (SP) are rare with SPB accounting for 70% of all SP while 30% comprising of solitary extramedullary plasmacytoma of all SP, SPB has a cumulative incidence of 0.15/100.000.[3] occurring in vertebrae, pelvis and ribs with rare occurrence in the skull, even low incidence in young population with amyloid production as in the present case (Table 1). SPB present as mass lesion comprising of clonal plasma cell with normal bone marrow examination and absence of other symptoms (CRAB).

A complete work up comprising of clinical, laboratory and radiological assessment is required before diagnosing SPB, all patients should be recommended for bone marrow aspirate and biopsy in order to rule out>10% monoclonal plasma cells in BM along with regular follow up and repeat examination should be performed in case of baseline plasma cell infiltration as 50% of SPB may progress to multiple myeloma (MM) within 10 years.[4]

Histopathology of the lesion provides the definite diagnosis, however imaging studies including Positron emission tomography/Computerized tomography (PET/CT) and magnetic resonance imaging (MRI) are essentially required and recommended to support the diagnosis as well as rule out any additional lytic lesions in SPB.

The treatment of SPB includes complete or partial resection for debulking and diagnostic purpose followed by localized fractionated radiotherapy given at a dose of 40-50 Gy at daily dose of 1.8-2.0 Gy per fraction as given in current case which showed good response, however Tsang et al suggested that tumor>5cm showed only 38% local control rate so large tumor mass require a higher radiotherapy dose or combination therapy for local control is controversial due to rarity of SPB leading to lack of clinical trial and it remains matter of debate.[5]

Treatment assessment in SPB is done according to international myeloma working group (IMWG) criteria for multiple myeloma as no guidelines for SPB have been recommended, hence regular follow-up by blood and urine testing is required along with BM aspiration, biopsy and imaging to be done when a progression is suspected.[6] An interesting finding in the current case was detection of large amorphous, eosinophilic, hyaline, extracellular deposits on HE staining positive on congo red showing apple green birefringence on polarized microscopy with positive expression for amyloid-P component and negative expression for Amyloid AA on IHC, further categorizing deposit as AL amyloidosis (Primary amyloidosis).

AL type amyloidosis is usually associated with multiple myeloma and deposits are most commonly seen in kidney, liver and skin, only few cases of plasmacytoma of skull with amyloidosis have been reported, categorization of amyloid has not been done in any of the cases (Table 1). The amyloid deposits may be massive, outnumbering the plasma cells and detection of clonality in such cases may become difficult and represent burnout-plasmacytoma, hence careful examination of section with amyloid deposits should be performed as seen in the current case along with foreign body giant cell reaction often associated with AL type amyloidosis as reaction to amyloid deposit, however lack of giant cell reaction in systemic amyloidosis lead to the hypothesis that the giant cells may play a role in transformation of full length light chain into insoluble forms which are formed from N terminal light chain fragments and deposition of amyloid.[7,8]

The presence of amyloid deposit further necessitates the thorough examination to rule out any evidence of multiple myeloma as treatment guidelines would change, in the present case there was no evidence of multiple myeloma and patient responded well to the radiation therapy however, presence of amyloid deposit indicates that the patient should be kept on close regular follow as probability of progression is high in these cases.[9]

CONCLUSION

The current management and follow up guidelines of SPB are based on small scale studies due to low incidence of SPB, larger prospective clinical trials are needed for further devising the treatment and response guidelines along with improved imaging studies differentiating active and inactive lesions. Advance and sensitive methods may be used for detection of minimal residual disease in SPB as used in multiple myeloma. Studies with large group are required to identify and stratify the group having high risk of progression, may benefit from adjuvant treatment regimen, which may further delay the progression.

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