Extramedullary left temporal plasmacytoma resembling as meningioma: a case report and literature review

Gopal Raman Sharma[1'2], Alok Deo[1], Samir Aryal[1,2], Ram Chandra Shrestha[1], Maya Bhattachan[1], Sumit Joshi[1,2], Dambar Bikram Shah[1,2], Rakesh Pathak[3]

[1]Department of Neurosugery, Dirghayu Neuro hospital and Allied sciences, Chabahil, Kathmandu, Nepal.

[2]Department of Neurosciences, Nepal Mediciti, Bhaisepati, Lalitpur, Nepal

[3]Department of pathology, Dirghayu Neuro Hospital and Allied sciences, Chabahil, Kathmandu, Nepal

ABSTRACT

When plasmacytoma occurs outside bone marrow, it is called extramedullary plasmacytoma (EMP), a rare malignant disorder due to excessive proliferation of abnormal plasma cells in other system of the body like the gastrointestinal tract, thyroid glands, breast, kidney, and central nervous system (CNS). Extramedullary plasmacytoma involving the central nervous system is very rare and may involve skull, dura mater, brain parenchyma and spines. Hence, we present a very rare care of EMP of left temporal lobe involving bone, dura and brain parenchyma on a 56-year-old righthanded gentleman who presented with left-sided V2 trigeminal neuralgia. In this case report, we will be discussing on natural course, clinical pathological characteristics, diagnosis, treatment, and prognosis of intracranial EMP.

KEYWORDS

Extramedullary, Intracranial, Mutiple Myeloma, Outcome, Plasmacytoma, Surgery

*Corresponding Author |
Dr. Gopal Raman Sharma
Department of Neurosciences, Nepal Mediciti, Lalitpur, Nepal
Email: gopal47@hotmail.com

INTRODUCTION

Extramedullary plasmacytoma (EMP) is one of the uncommon plasma cell tumors that occur outside the bone marrow 1,2,3. It is a form of multiple myeloma (MM), a hematological malignancy of bone marrow due to excessive proliferation of abnormal plasma cells within bone marrow. The majority of EMP affect gastrointestinal tract, thyroid, breast, kidney, spine, skull and central nervous system3,4. EMP of skull and dura mater without previous history of MM is rare, and solitary intracranial plasmacytoma (SIP) without involvement of skull and dura mater is extremely rare4,5,6,7. The most common site of extramedullary intracranial plasmacytoma (EMIP) are the temporal and clivus areas which may mislead to a diagnosis of meningioma or chordoma, however, it may involve any part of skull, dura or brain parenchyma inside the calvarium7,8,9,10,11.

Because of limited experiences and paucity of literature definite management of EMIP is questionable. Available treatment options are surgery, radiotherapy, chemotherapy alone and or combination of them12,13,14,15,16.

A CASE REPORT

A 56 -year-old righthanded gentleman was presented to our neurosurgery OPD with history of persistent headache and left sided episodic facial pain for last 3 months. There was no history of vomiting, fainting attacks and weakness of extremities. He has been treated for multiple myeloma (MM) three years ago and was in remission, however recently chemotherapy was restarted for recurrence of disease. On clinical examination, he was fully conscious and oriented of time, place and person. Funduscopic examination of both eyes revealed bilateral papilloedema. He has hyperesthesia over the left V2 dermatome. He had no other focal neurological deficit. MRI of brain with Gadolinium enhancement showed heterogeneously enhancing dural based extra axial lesion within left temporal fossa with gross perilesional edema compressing ipsilateral ventricle with midline shift to right and multiple heterogeneously enhancing skull lesions of various sizes also noted in the left tempero parietal area. Based on the age of the patient and radiological features our first clinical diagnosis was left temporal convexity meningioma involving overlying bone (Figure. 1a, b, c). As planned left pterional craniotomy and excision of mass was carried out under general anesthesia. Intraoperatively, the mass was extra axial, solid, fleshy, very vascular and attached to the dura and left temporal bone. The solid tumor was gradually devascularized and deattached from the dura and removed in piecemeal fashion using CUSA (computerized ultrasonic aspirator). Gross total removal of tumor was achieved. Looking at the morphological feature of the tumor during procedure our impression was left temporal convexity meningioma with hyperostosis of overlying bone.

Post operative events were smooth without any complication and CT scan of brain did not show any residual mass.

Histology feature was suggestive of poorly differentiated malignant neoplasm (Figure. 2). Immunohistochemistry (IHC) makers like CD138, MUM- 1, Kappa were raised to score 3, 20-25 % of tumor cells were immunoreactive to Ki-67 and all these features were consistent with plasma cell neoplasm which favor plasma cell neoplasm with plasmablastic differentiate (Figure. 3). Since the patient had gross total excision of the tumor we along with medical oncologists decided to continue chemotherapy and MRI to be repeated after three months.

At three months followed up patients was clinically normal and had no complain. As protocol we repeated MRI of brain with IV contrast which revealed recurrence of mass with gross involvement of adjustment temporal and frontal bone along with dura (Figure. 1d, e, f, g, h). Oncologists reviewed the case and decided to start a course of cranial radiation along with continuation of chemotherapy.

DISCUSSION:

Multiple myeloma (MM) is less common malignancy but second most common hematological neoplasm which is characterized by uncontrolled proliferation of abnormal plasma cells in the bone marrow6. MM is a disease of the bone marrow however, it occasionally infiltrates the other systems of the body which is known as extramedullary plasmacytoma (EMP)17. Incidence of plasmacytoma to MM range from 7 to 17% at diagnosis to 6-20% during the course of disease3. EMP account about 3% of all plasma cell tumors. Majority of EMP occurs in head, neck, and gastrointestinal tract, central nervous systems, thyroid, kidneys and the breast3,4. Intracranial involvement of MM is rare affecting <10 % of patients with MM18. Extramedullary intracranial plasmacytoma (EMIP) is rare plasma cell tumor that may involve skull, meninges and brain parenchyma3,4,5,6,7. Rarely EMIP may present with acute intracranial bleed and one case has been reported in literature19.

Patients with EMIP usually present with features of raised ICP, seizure and focal neurological deficit and cranial nerve palsies8,20,21,22,23. The patient may or may not have a previous history of the treatment of MM.

Radiological diagnosis is made by CT/ MRI of head with IV contrast. The lesion is usually extra axial and enhances homogenously or heterogenously with IV contrast. There may be involvement of bone and dura, which confuses with meningioma or chordoma5,9,10,11,17,24,25.

Systemic MM should be ruled out by doing bone marrow biopsy, serum B 2 macroglobulin, serum and urine protein electrophoresis and skeletal survey if patient has single solitary EMIP3,5,14. There is no clearcut guidelines for the management of EMIP. Surgical excision should be the first line treatment for symptom relief as well as for histological diagnosis 4,5,7,14.

Histological features of plasmacytoma are cellular atypical, pyroninophilia of tumor cells and presence of amyloidosis15,16.

In immunohistochemistry (IHC) description tumor cells are positive for k chain and negative for Lambda chain are strongly favor for immunomodulatory proliferation of tumor plasma cells18.

IHC also reveals that abnormal cells are immunoreactive for CD138, EMA and strongly positive for MUM-116.

Solitary intracranial plasmacytomas are radiosensitive can be treated only by radiotherapy alone12,13,14 however, for other varieties of EMIP surgery alone or a combination of both are more effective3,4,5,9,10,14,21. Regarding chemotherapy, Lenalidomide with low dose of dexamethasone is recommended for multiple myeloma with EMIP3,16,23. Other chemotherapies like thalidomide and bortezomib have been used for MM with EMIP16. Autologous stem cell transplantation is recommended for refractory recurrent and multiple plasmacytoma1. Radiotherapy achieves 80-100% local control with 50-65% disease free longer than 10 years5,14,16.

The survival of patients with MM with EMIP ranges from several months to more than 10 years depending on status of disease at the time of discharge3,5,14,16.

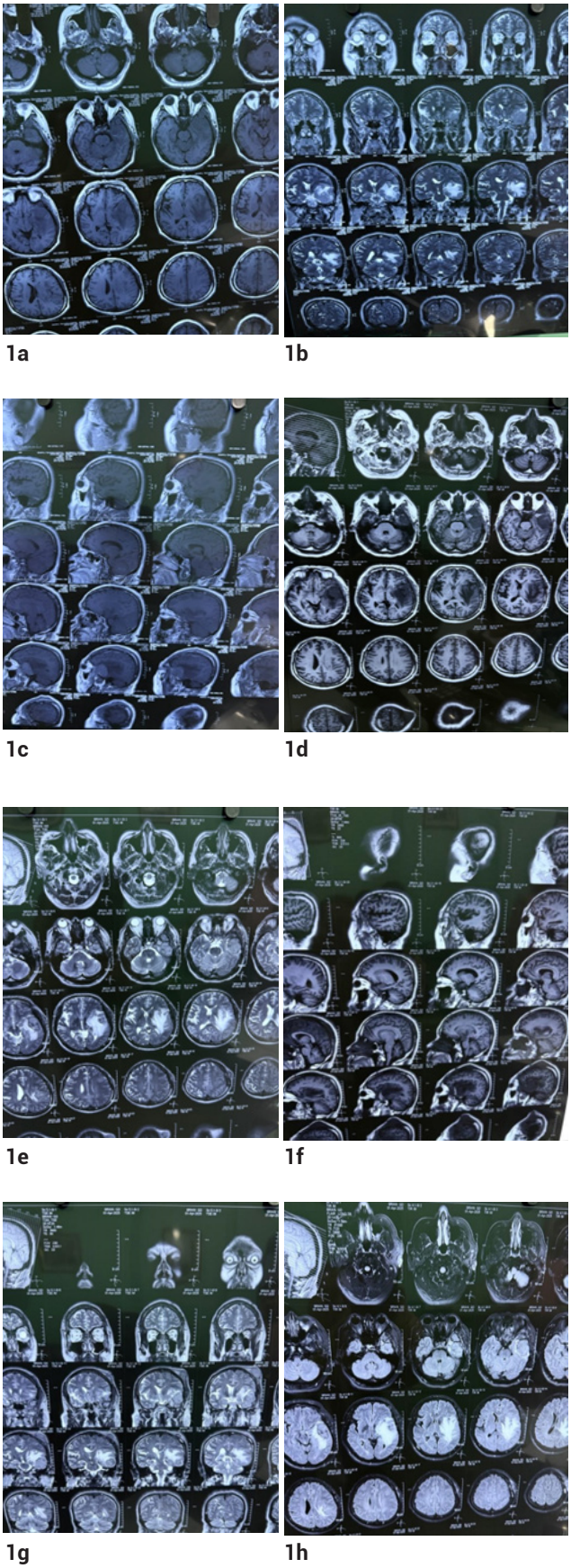
CONCLUSION:

Any intracranial extra axial mass with or without bone involvement on the background of MM should be considered as EMIP besides meningioma or chordoma.

There is no established treatment for EMIP and available treatment are surgery, radiotherapy and chemotherapy, alone or in combination of them.

Treatment should be individualized after discussion among treating surgeon, the Oncologist, patient and his/her family members.

Figure. 1



1a **1b**

1c **1d**

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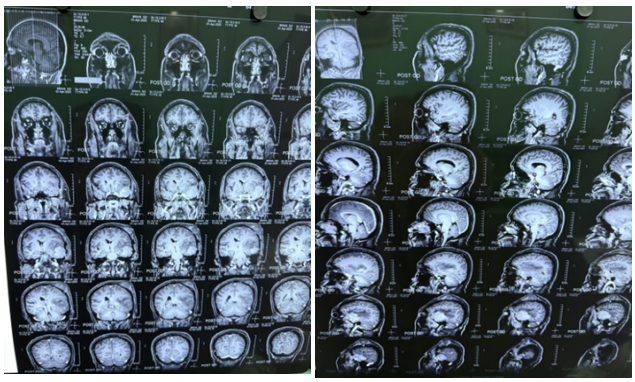
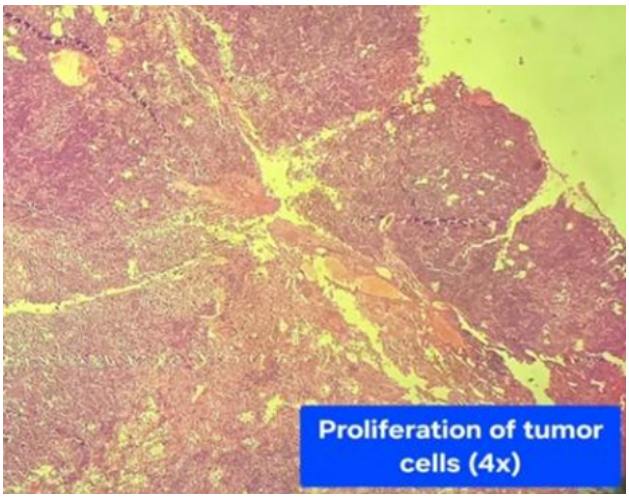


1i 1j

Figure 1. (1a, b & c) Preoperative MRI of brain of a 56year man showing low intensity on T1W1 images and high intensity on T2W2 images left temporal axtra axial mass involving overlying dura and bone, there is homogenous enhancement of lesion with intense perilesional edema.

Postoperative (after three months) MRI of brain of same patient (1d, e, f, g, h) showing recurrence of tumor with surrounding edema compressing the ipsilateral ventricle.

Figure. 2



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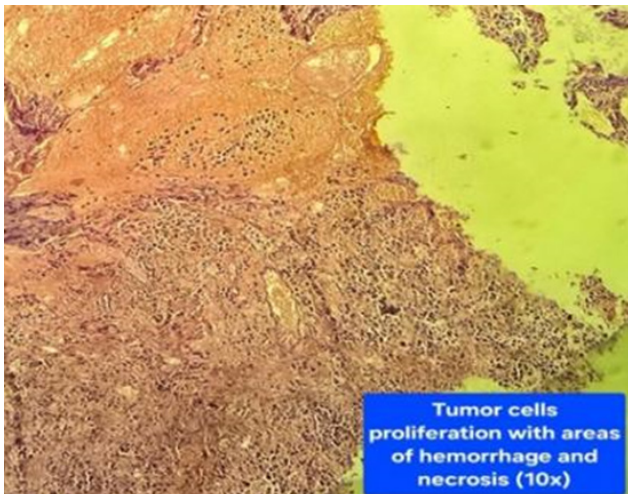

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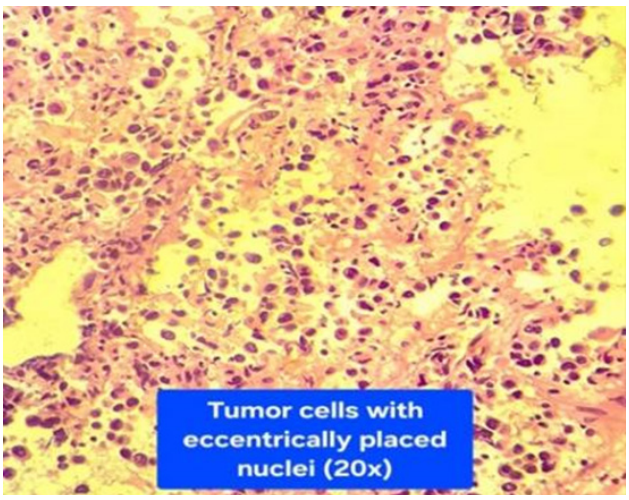

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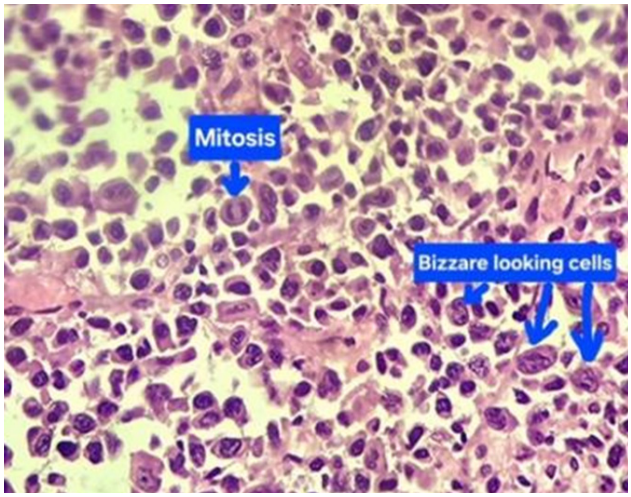

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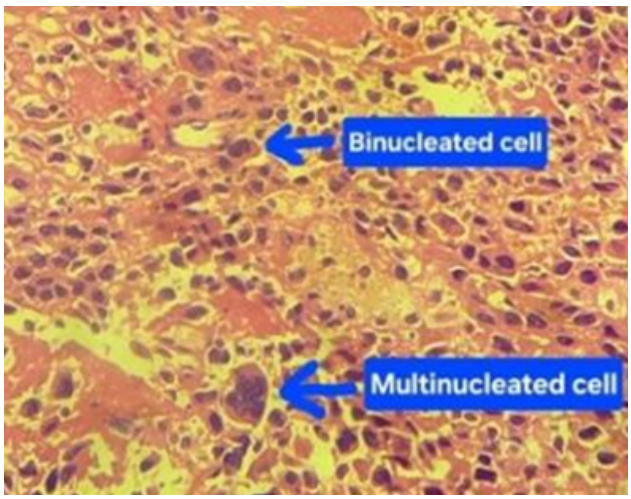
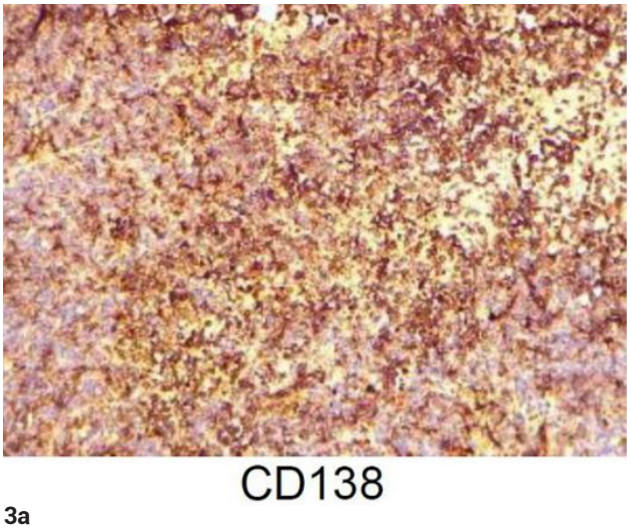


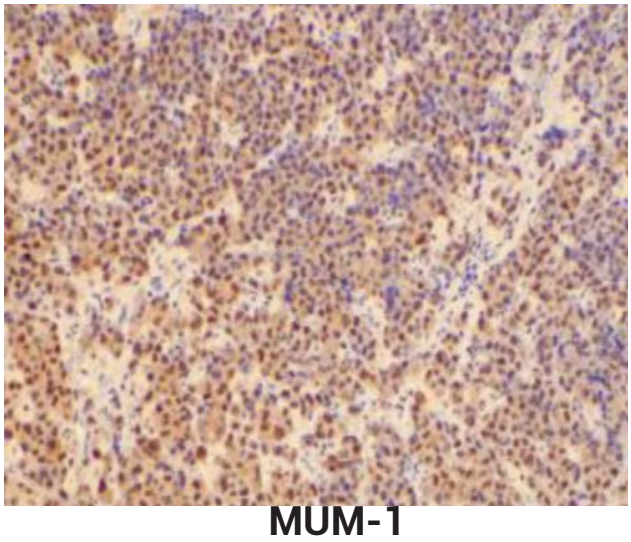
2b

Figure 2. Histological features (a, b, c, d. e): Tumor comprised of atypical cells with eccentrically placed nucleoli. Sheets of tumor cells revealed moderate to marked pleomorphism with frequent mitoses. There is associated hemorrhage, bizarre cells and marked inflammatory cells including plasma cells. These microscopic features are suggestive of a poorly differentiated malignant neoplasm.

Figure. 3



3a



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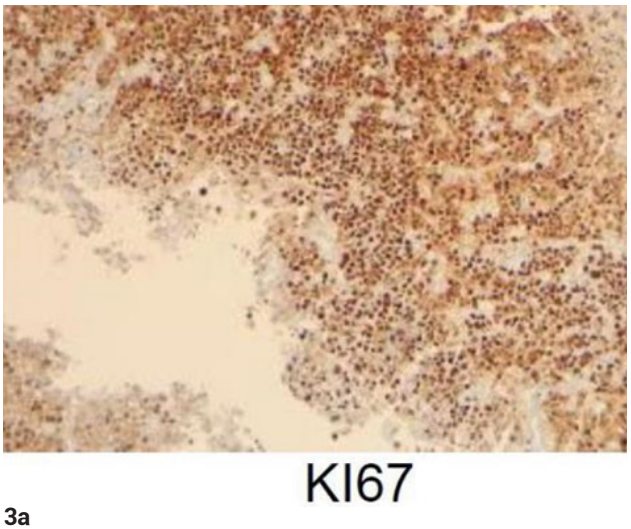

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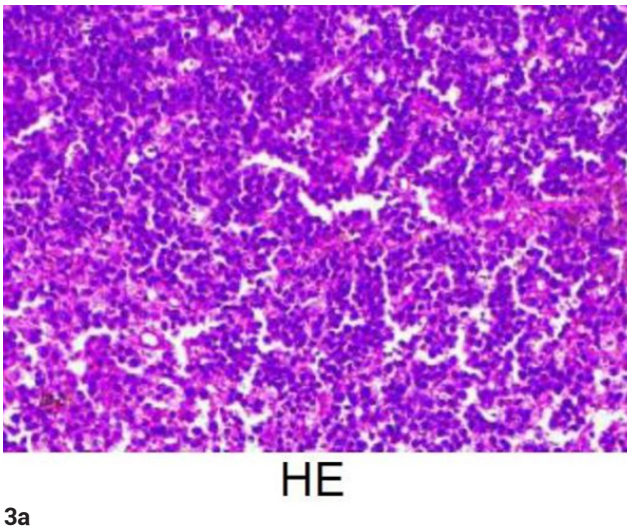

3a

Figure 3. Immunohistochemistry IHC): (a, b. c, d): IHC examination showed CD138 and MUM-1 immunoreactive of 3+ scores. Ki – 67 is positive in 20-25% of tumor cells and cells are immunoreactive to Kappa of 2+ Score. These all features are suggestive of plasmablastic malignant neoplasm.

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