



# Primary Cerebral Plasmacytoma: A Rare Case Report with Review of Literature

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## Abstract

Solitary craniocerebral plasmacytoma is the least common form of extramedullary solitary plasmacytoma (SP). Cerebral SP is very rare. The world literature counts only seven intracranial cases. The authors report a case of cerebral tumor of SP in a 52-year-old female who presented to a hospital with headache and difficulty in walking for 6 months. Contrast-enhanced computed tomography (CECT) brain showed left occipital intracranial space occupying lesions (ICSOL) of size 26 mm × 14 mm adjacent to superior sagittal sinus with disproportionate perilesional edema and midline shift toward the right. She underwent gross total tumor excision with left fronto-temporo-parietal decompressive craniectomy. Microscopic sections showed highly cellular tumor infiltrating white matter and gray matter. Histopathology revealed plasmacytoma. These plasma cells were seen infiltrating the adjoining brain parenchyma. Immunohistochemistry study showed the following pattern of antigens: most of the cells were negative to CD138 antibody, and majority of cells were positive (++ to +++) to CD56 antibody, more groups of cells were positive to lambda antibody than KAPPA. Bone marrow biopsy showed only 1% polyclonal plasma cells. Whole body bone scan showed no evidence of any osteoblastic skeletal metastasis. The patient recovered well, and cranioplasty was done after 6 weeks.

## Keywords

- ▶ solitary plasmacytoma
- ▶ extramedullary plasmacytoma
- ▶ solitary plasmacytoma of bone
- ▶ multiple myeloma (MM)

**Conclusion** Cerebral SP is rarely found in clinical practice and if diagnosed properly can save patients life. This case report would definitely address many unexplored facts about cerebral SP and set milestone in the field of clinical research.

## Introduction

The primary solitary plasmacytoma (SP) comes into mind when there is localized accumulation of neoplastic monoclonal plasma cells in the tissue with absence of other features of systemic plasma cell proliferative disorder (i.e., anemia, hypercalcemia, renal insufficiency, or multiple lytic bone lesions). SP can be classified into two groups depending upon its location: primary solitary plasmacytoma of the bone (SBP) if the tumor involves an osseous site and extramedullary

plasmacytoma (EMP) if it involves an extra osseous site. The differences in cellular adhesion molecules or chemokine receptor expression profiles of the malignant plasma cells may have impact on the development of SP or multiple myeloma (MM). EMP is a very rare disease. EMP is most often located in the head and neck region, mainly in the upper aerodigestive tract, such as the nasal cavity and nasopharynx, but may also occur in the gastrointestinal tract, urinary bladder, central nervous system, thyroid, breast, testes, parotid gland, lymph nodes, and skin. Primary craniocerebral plasmacytoma is the

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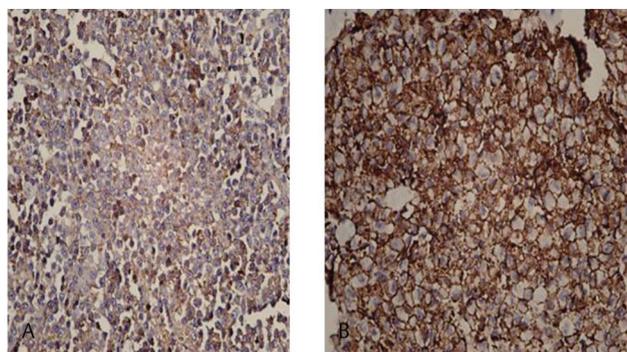


least common form of extramedullary primary plasmacytoma. The most common sites of primary plasmacytomas arising from the skull are the parietal bone and the bones of the skull base. The dura is the most common site of craniocerebral plasmacytoma not originating in the skull, but occasionally involving the calvaria and brain parenchyma secondarily. Primary plasmacytoma can arise within brain tissue very rarely. World literature revealed only seven such cases.<sup>1</sup> Here, the author reports a cerebral primary plasmacytoma.

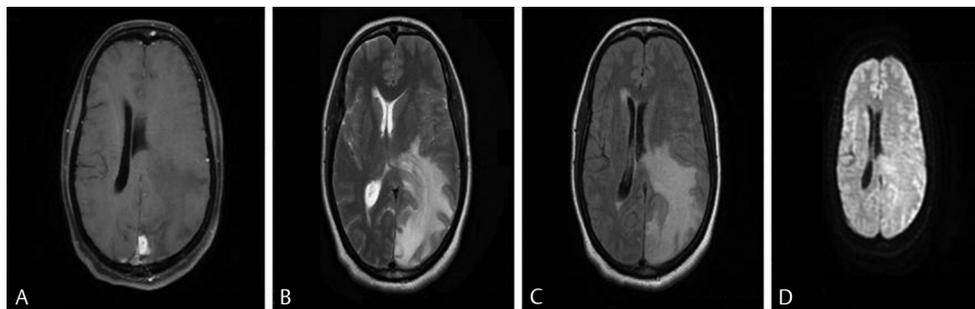
### Case Presentation

A 52-year-old female presented to our hospital with chief complaints of headache and difficulty in walking for 6 months. There was a history of loss of consciousness once for few minutes. There was no history of fever. On examination, she was conscious but confused, the Glasgow Coma Scale (GCS) score was E4V4M6, heart rate (HR) 50/min, and blood pressure (BP) 128/68 mm Hg; pupils were reacting to light but were unequal, and power in upper and lower limbs was 4/5 plantar–extensor bilateral. Magnetic resonance imaging (MRI) of the brain was done for study of lesion in left occipital lobe with disproportionate edema, which showed a tubular, relatively well-defined, heterogeneous, T2-hyperintense, T1-isointense, diffusion unrestricted, noncalcified, nonhemorrhagic lesion. Intense post-contrast enhancement was seen. MR spectroscopy revealed prominent lipid lactate peak at 1.33 and no obvious elevated perfusion was noted on perfusion-weighted imaging (PWI) (►Fig. 1). After clinical and radiological evaluation, it was decided to decompress the tumor in view of mass effect and also for proper histopathological diagnosis, and at the same time, to decrease the deleterious mass effect. After all preoperative investigations, she underwent craniotomy, gross total tumor excision with decompressive craniectomy of the left fronto-temporo-parietal region due to brain swelling at the time of closure. Intraoperatively, tumor was grayish white in color, highly vascular and infiltrating surrounding brain tissue. Microscopic sections showed highly cellular tumor infiltrating white matter and gray matter. The tumor was composed of sheets of plasma cell and a few lymphocytes in between; the plasma cells had large nuclei, some were vesicular, and few of them were binucleated. Nucleoli were also seen in some

of the tumor cells. Nuclear atypia and mitotic figures were seen at places. These plasma cells were seen infiltrating the adjoining brain parenchyma. Hemorrhagic areas were seen amidst these fragments of tumor cells. Immunohistochemistry showed CD 138—most of the cells were negative except a few cells occasionally, CD 56—majority of cells were positive (++) to (+++), kappa—some groups of cells were positive and others were negative, and lambda—more groups of cells were positive as compared with kappa (►Figs. 2A and B). Immunohistochemistry antibodies used were manufactured by Dako mouse monoclonal antihuman antibody CD138-MI15 clone. Antibodies used were manufactured by Dako mouse monoclonal anti human antibody CD56-123C3 clone, Biogenex mouse monoclonal antibody anti-kappa light chain K88 clone, and Biogenex mouse monoclonal antibody anti-lambda light chain HP6054 CLONE. DAB chromogen were used. After confirmation of plasmacytoma on histopathology, investigations were performed on the line of MM diagnosis. Routine blood investigations were within normal limit except serum alkaline phosphatase 180 IU/L (range 50–136), serum beta 2 micro globulin 1660 µg/L (range 670–1,310), and erythrocyte sedimentation rate (ESR) 52 mm/h (range 0–20). We excluded MM by serum electrophoresis and urine examination for Bence–Jones protein. Whole body bone scan did not show any scintigraphic evidence of osteoblastic skeletal disease. Bone marrow biopsy showed only 1% polyclonal plasma cells. The patient recovered well after operation, and cranioplasty was



**Fig. 2** Immunohistochemistry showed CD 138—most of the cells were negative except a few cells occasionally, CD 56—majority of cells were positive, kappa—some groups of cells were positive and others were negative, and lambda—more groups of cells were positive as compared with kappa.



**Fig. 1** (A–D) MRI brain showed lesion in left occipital lobe with disproportionate edema, which showed a tubular, relatively well-defined, heterogeneous, T2 hyperintense, T1 isointense, diffusion unrestricted, non-calcified, non-hemorrhagic lesion with post contrast enhancement. MRI, magnetic resonance imaging.

done after 6 weeks. The patient has taken palliative external beam radiotherapy to whole brain, with a total dose 39 Gy, in 13 fraction, for 2.5 weeks, 3Gy per day for 5 days per week and been doing well at follow-up. Postoperative MRI brain after 6 months showed correction of edema with a decrease in the size of plasmacytoma.

## Discussion

Cranio-cerebral plasmacytomas lesions are called primary when they arise primarily from the skull or its contents. SP is excluded when it arises as the result of extension from extracranial skeletal sites of myeloma and when it is associated with plasmacytosis within bone marrow or extracranial myeloma. Cranio-cerebral SP is a rarer disease than the more common plasma cell tumor that arises within the cranium as a result of disseminated MM. The distinction between the two variants makes the difference in outcome because primary cranio-cerebral plasmacytoma is a relatively benign entity, which is potentially curable, whereas MM generally has a poor prognosis.<sup>2</sup> Primary cranio-cerebral plasmacytoma is also a different disease from plasma cell granuloma, which is an inflammatory pseudotumor characterized by a benign proliferation of polyclonal plasma cells (unlike the monoclonal population seen in plasmacytoma), lymphocytes, histiocytes, and many small blood vessels. As cranio-cerebral SP is not associated with extracranial disease, but rarely an abnormal monoclonal immunoglobulin (g) can be detected within serum or urine. Once the histologic diagnosis of plasmacytoma is reached, a systemic plasma cell dyscrasia must be excluded by bone marrow evaluation, serum protein electrophoresis for presence of a monoclonal gammopathy, and bone scan or skeletal survey. A monoclonal population of cells excludes both plasma cell granuloma and meningioma with conspicuous plasma cell components. Primary cranio-cerebral plasmacytoma is a radiosensitive tumor that can be treated by radiation therapy alone, surgery alone, or a combination of the two. When increased serum Ig is present, levels can be followed as a measure of therapeutic success. Patients are maintained on long-term follow-up especially in the late postoperative period to monitor generalized plasma cell dyscrasia. In this case, we excluded MM by serum electrophoresis and urine examination for Bence-Jones protein. Immunohistochemistry showed CD 138—most of the cells are negative except a few cells occasionally, CD 56—majority of cells are positive (++ to +++), kappa—some groups of cells are positive and others are negative, and lambda—more groups of cells are positive as compared with kappa. Bone marrow biopsy showed only 1% polyclonal plasma cells. We also did bone scanning to rule out MM. Moreover the patient had symptoms for 6 months and recovered well after operation, so this case is unlikely to be MM. Mancilla et al in 1976 reviewed literature and considered only three primary intraparenchymal plasmacytoma (→Table 1).<sup>3-5</sup> Bindal et al in 1995 reported a series of eight cases and concluded that MM is unlikely to develop during the long term in patients with intracranial plasmacytoma who do not develop MM or myelomatous changes in the early postoperative period;

**Table 1** Enumeration of some cases of previously described primary cerebral plasmacytoma

Authors	Age/sex of the patient	Localization of plasmacytoma
Cabot et al <sup>3</sup>	72/M	Intracerebral, diffuse involvement
French <sup>5</sup>	42/F	Hypothalamus
Goriachkina <sup>7</sup>	18/F	Hypothalamus
Ferrari et al <sup>6</sup>	50/F	Left temporal-insular lobe and hypothalamus

Abbreviations: F, female; M, male.

however, lesions that infiltrate the skull base are not likely to be primary, and patients who harbor these neoplasm should undergo complete evaluation and close follow-up review to exclude MM. Ferrari et al in 2012 reported an unusual case of primary intraparenchymal brain plasmacytoma and concluded that primary cerebral plasmacytoma has good prognosis in contrast to plasmacytoma, which occurs as initial presentations or spread of MM.<sup>6,7</sup>

Histopathological examination of the tumor is required for definitive diagnosis of central nervous system (CNS) plasmacytomas because they have no pathognomonic clinical or radiologic features. These are usually dense, homogeneous tumors with infiltrates of plasma cells. Amyloid deposition may be present in 15 to 38% of the cases. Immunoglobulin G (IgG) is the most common Ig expressed by these tumors with kappa chain restriction. Plasmacytomas can be graded low (grade 1), intermediate (grade 2), and high grades (grade 3) based on the cellular atypia.<sup>8</sup> The differential diagnosis in these patients should include plasma cell granulomas; these can be distinguished from plasmacytomas microscopically. Histologically, plasma cell granulomas are characterized by an admixture of plasma cells, lymphocytes, or macrophages on a background of partly necrotic tissue, including some areas of fibrosis and other areas of granulation tissue.<sup>9</sup> One should consider meningioma, metastasis, lymphoma, dural sarcoma, and leptomeningeal carcinomatosis as an important differential diagnosis. The treatment of CNS plasmacytomas changes according to the extent of the disease, and in primary tumors, local surgical debulking followed by radiotherapy has shown good outcomes, but once the disease becomes systemic, chemotherapy is the main treatment.<sup>10-12</sup> It has been reported that extramedullary localizations of myeloma originating from the cranial bone usually respond well to new drugs (e.g., thalidomide, bortezomib and lenalidomide) compared with intraparenchymal plasmacytomas. So, our case is one of the rare case reports of primary plasmacytoma of brain. After proper diagnosis and management, this rare tumor can be cured.

## Conclusion

To summarize, cerebral primary plasmacytoma is rarely found in clinical practice and if diagnosed properly can save patients life. This case report would definitely address many

unexplored facts about cerebral primary plasmacytoma and set milestone in the field of clinical research.

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No financial support of any kind from anybody for this study was received.

#### Conflict of Interest

There is no conflict of interest with anybody.

#### Contribution of the Authors

Dr. Arun Kumar and Dr. Biswaranjan Nayak conceived the original article design, drafted the original manuscript reviewing current literature, and corresponded with the editorial team. Dr. BH Krishnamurthy and Dr. Sushant Kumar Patro summarized the case and helped substantially with the drafting and critiquing of the original manuscript.

#### Declaration of Authors

The manuscript has been read and approved by all the authors, the requirements for authorship as stated earlier in this document have been met, and each author believes that the manuscript represents honest work.

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