Multiple myeloma manifesting as an intraventricular brain tumor

Case report

*JOO-HUN DAVID EUM, M.D.,1 ASTRID JEIBMANN, M.D.,2 WERNER WIESMANN, M.D.,3 WERNER PAULUS, M.D.,2 AND HEINRICH EBEL, M.D.1

Departments of 1Neurosurgery and 3Radiology, St. Barbara Hospital, Hamm; and 2Institute of Neuropathology, University Hospital of Münster, Germany

Primary intracerebral manifestation of multiple myeloma is rare and usually arises from the meninges or brain parenchyma. The authors present a case of multiple myeloma primarily manifesting within the lateral ventricle. A 67-year-old man was admitted with headache accompanied by slowly progressing right hemiparesis. Magnetic resonance imaging showed a large homogeneous contrast-enhancing intraventricular midline mass and hydrocephalus. The tumor was completely resected, and histopathological examination revealed plasmacytoma. After postoperative radio- and chemotherapy, vertebral osteolysis was detected as a secondary manifestation of multiple myeloma.

(KEYWORDS: multiple myeloma, plasmacytoma, ventricle)

Plasma cell tumors are characterized by monoclonal proliferation of immunoglobulin-secreting plasma cells. Intracranial plasmacytoma is rare and may occur as metastatic spread of multiple myeloma, solitary extramedullary plasmacytoma, or initial manifestation of multiple myeloma. Intracranial plasmacytoma may be localized in the meninges, brain parenchyma, or skull base. We present a case of primary manifestation of multiple myeloma in the lateral ventricle, a location which has not been described before.

Case Report

Examination and Operation. This 67-year-old man with no significant medical history presented with headache and right hemiparesis that slowly worsened over a 2-month period. His family had noted a change in his personality. Neurological examination revealed right hemiparesis with facial involvement and hemihypesthesia. Cranial MR imaging displayed a large homogeneous contrast-enhancing intraventricular mass and hydrocephalus (Fig. 1). The patient underwent left occipital craniotomy. Intraoperatively, the tumor appeared to be an encapsulated solid mass. Gross-total resection was achieved, and an external ventricular drain was placed.

Histopathological Examination. Histopathological examination demonstrated a tumor of high cellular density and plasma cell differentiation displaying large cells with basophilic cytoplasm, often round eccentric nuclei with “spoke wheel” chromatin, and a perinuclear halo (Fig. 2A). Mitotic activity was increased. The tumor was sharply demarcated from adjacent reactive brain tissue. On immunohistochemical analysis, tumor cells showed expression of epithelial membrane antigen and a plasma cell marker (clone VS38c). Stainings for common leukocyte antigen CD45 (LCA), the T-cell antigen CD3, and B-cell antigen CD20 (L26) were negative. Positive staining for lambda light chain and absence of kappa light chain expression demonstrated plasmacytoma with light chain restriction (Fig. 2C and D). The Ki 67/MIB1 proliferation index was high (30%, Fig. 2B).

Postoperative Course. Postoperatively, the patient underwent chemotherapy administered intrathecally, comprising 18 cycles of arabinosylcytosine (Ara-C) and dexamethasone followed by cranial radiotherapy with 39.6 Gy and a boost of 10.8 Gy. Seven months after the operation, follow-up examination revealed osteolysis of T-7, which was treated with local radiotherapy (30 Gy) and...
adriamycin in combination with thalidomide and dexamethasone. Follow-up cranial MR imaging examinations performed 10 months postoperatively demonstrated a left temporal dynamic liquor block. A ventriculoperitoneal shunt was placed, and the patient received 3 additional cycles of thalidomide and dexamethasone. Subsequently, stem cell mobilization therapy with epirubicin, etoposide, and ifosfamide was carried out; bisphosphonate therapy was continued. Eighteen months after surgery there is no imaging or clinical evidence of intracranial relapse.

Discussion

To our knowledge this is the first documented case of multiple myeloma primarily manifesting as an intraventricular brain tumor. Tumors encountered in the vicinity of the ventricles comprise a variety of entities including choroid plexus papilloma, ependymoma, subependymoma, subependymal giant cell astrocytoma, neurocytoma, meningioma, and cerebral metastasis. Indeed, the imaging findings in the present case were initially interpreted as being most consistent with choroid plexus papilloma, meningioma, or neurocytoma.

Intracranial plasmacytoma may represent metastatic spread of multiple myeloma; however, it may also occur as solitary extramedullary plasmacytoma, which accounts for only 3–5% of plasma cell neoplasms. Solitary extramedullary plasmacytoma of intracranial location most frequently affects the meninges, the brain parenchyma, or the skull base.

An intraventricular location has not been described

![Fig. 1. Coronal (A) and axial (B) T1-weighted MR images, and axial CT scans (C and D). A: Image obtained after contrast administration revealing an intraventricular tumor mass reaching the roof of the lateral ventricles. B: Gadolinium-enhanced image showing a homogeneous lesion located in the left ventricle with extension to the opposite lateral ventricle. C and D: Postoperative images showing complete removal of the intraventricular tumor mass.](image)

![Fig. 2. Photomicrographs. A: An H & E–stained section displaying a tumor of high cellular density with plasma cell differentiation, basophilic cytoplasm, and frequently eccentric nuclei. There are several mitoses. B: The Ki 67/MIB-1 proliferation index is 30%. C and D: Staining for lambda light chain with distinct cytoplasmic positivity (C), while kappa light chain expression is absent (D). Original magnification × 400.](image)
Intraventricular plasmacytoma

in multiple myeloma or in solitary extramedullary plasmacytoma. The pathogenesis of intracranial plasmacytoma has been debated, and several hypotheses have been proposed. Primary central nervous system hematopoietic tumors may arise on B-cell transformation anywhere in the body, and subsequent attraction by cerebral adhesion molecules or chemokines/chemokine receptors. On the other hand, systemic tumor cells could be eradicated by the immune system, but may hide within the immunoprivileged central nervous system. Finally, polyclonal inflammatory lesions may develop into monoclonal neoplasias.

The exact relationship between multiple myeloma and solitary extramedullary plasmacytoma remains poorly understood. Some authors believe that these entities are manifestations in the spectrum of plasma cell tumors, and others suggest that extramedullary plasmacytoma is a separate entity. A significant percentage (up to 85%) of patients with solitary plasmacytoma will eventually develop disseminated multiple myeloma, if followed for a sufficient period of time. The clinical course in our patient is most compatible with the notion that most cases of intracranial plasmacytoma heralded systemic manifestations of multiple myeloma.

Surgical removal followed by postoperative irradiation is the treatment of choice in cases of intracranial plasmacytoma. In a few cases a successful curative approach was achieved by complete surgical resection alone or biopsy followed by radiotherapy. Gross-total resection followed by radiotherapy was well tolerated by our patient.

In conclusion, plasmacytoma may rarely manifest as an intraventricular tumor, radiologically and clinically mimicking other tumor entities more frequently encountered in the vicinity of the ventricular system.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References