Unusual Case of Solitary Intraparenchymal Brain Plasmacytoma

Case Report

A 50-year-old, previously healthy woman was referred to the emergency unit of our institution for sudden motor aphasia resolving spontaneously in 15 minutes. Magnetic resonance imaging (MRI) of the brain showed infiltrative contrast-enhancing tissue in the left temporal-insular lobe and thalamus, with surrounding edema and a mild axial median shift (Figs 1A and 1B). The tissue also infiltrated the corpus callosum with initial diffusion to the white matter of the right hemisphere. No dura mater or skull involvement was documented. MRI spectroscopy confirmed the presence of infiltrating lymphoma-like tissue. Electroencephalography showed nonspecific wave slowing in the left frontotemporal region.
The patient was admitted to the neurosurgery department in which she underwent a stereotactic computed tomography (CT)–guided brain biopsy. Histologic examination of the biopsy specimen showed mature plasma cells in the brain parenchyma with areas of perivascular infiltration (Fig 2A). Immunohistochemistry analysis revealed CD138+/H11001 elements expressing K light-chain restriction (Fig 2B), which confirmed the presence of a mature plasma cell neoplasm. The Ki-67 proliferation index was 5%. Cytogenetic analysis was not performed as a result of tissue paucity.

On the basis of the diagnosis of brain plasmacytoma, the patient underwent additional assessments. A morphologic and flow-cytometry analysis of CSF showed no clonal plasma cells. Spinal cord MRI was negative. Laboratory tests showed a normal hemogram, and no serum or urinary monoclonal components were detected. Bone marrow aspirate and trephine biopsy were negative. Bone marrow cytogenetics and fluorescent in situ hybridization analysis revealed no abnormalities. A complete skeletal radiogram failed to reveal osteolytic bone lesions; chest and abdominal CT scans were negative. Positron emission tomography–CT scans showed only an area of hypometabolism in the left cerebral hemisphere, which corresponded to the lesion observed with MRI. At the end of the staging assessments, the diagnosis was consistent with a solitary intraparenchymal brain plasmacytoma. Neurologic examination at that time showed a mild delay of verbal fluency, mild motor aphasia, and mild gait dis-equilibrium. Neuropsychological tests, including the Rey-Osterrieth complex-figure test, Rey’s auditory verbal learning test, a verbal fluency test, and Beck’s depression inventory, revealed mild impairment of short-term and visuospatial memory and minor depression. The patient started therapy with intravenous dexamethasone (16 mg per day) and prophylactic antiepileptic therapy with oral levetiracetam (500 mg twice a day). She then received whole-brain radiotherapy with 36 Gy (20 fractions) plus a 9-Gy boost in the area of the lesion (five fractions), which made a total radiation dose of 45 Gy.

Over a 48-month follow-up period, repeated brain and spinal cord MRI investigations yielded no evidence of disease progression. The results of the last brain MRI performed are shown in Figures 1C and 1D. Bone marrow examination, serum and urine electrophoresis, and chest and abdominal CT scans consistently yielded negative findings, thus excluding progression to multiple myeloma or evidence of systemic disease. Neurologic examination showed a mild delay of verbal fluency, mild gait dis-equilibrium, and mild motor aphasia. The patient is self-sufficient as regards daily life activity, working in regular employment, and taking care of two adolescent boys as a single mother. The patient is taking oral dexamethasone (0.75 mg every other day) and levetiracetam (500 mg twice a day). She is currently being treated with oral diphosphonates, calcium, and vitamin D to prevent steroid bone damage.

**Discussion**

Plasma cell neoplasms account for 1% of malignant tumors, including multiple myeloma, solitary plasmacytomas, and syndromes caused by tissue immunoglobulin deposition. A solitary plasmacytoma is a localized plasma cell neoplasm that usually affects bone (bone plasmacytoma) and rarely involves other tissues (extramedullary plasmacytoma). In general, a bone plasmacytoma or plasmacytoma with serum paraprotein at diagnosis is believed to progress toward multiple myeloma more frequently than extramedullary plasmacytomas.

Extramedullary plasmacytomas account for 3% to 5% of plasma cell neoplasms, which mostly affect patients in the sixth or seventh decade of life. In 80% of cases, extramedullary plasmacytoma are localized in the upper respiratory tract; more rarely, they may affect the GI tract, lymph nodes, bladder, and CNS. Solitary plasmacytomas involving the CNS usually result from the spread of malignant plasma cells from the dura mater or skull and are generally referred to as...
intracranial plasmacytomas. Among the latter, primary intraparenchymal brain plasmacytomas (ie, solitary intracranial plasmacytomas without dura mater or skull involvement) have been reported as extremely rare. Because plasma cells are not found in the brain tissue in normal conditions, an isolated intraparenchymal involvement is difficult to explain in the absence of contiguous dural or bone lesions.

As regards treatment, several single-center and multicenter retrospective studies have reported effective treatment of solitary plasmacytomas (both bone and extramedullary) with radiotherapy. Solitary extramedullary plasmacytomas, in particular, are highly radiosensitive tumors that can be controlled in 80% to 100% of cases with moderate doses of radiotherapy. Accordingly, complete surgical removal should be considered only if feasible, and adjuvant chemotherapy might be a valid option only in selected high-risk patients. Data from the literature have suggested an effective median radiation dose in the range from 40 to 60 Gy. However, whether the radiation dose correlates with better local control remains a debatable issue.

As far as primary intracranial plasmacytomas are concerned and, particularly, in the case of primary intraparenchymal brain plasmacytomas, information regarding treatment is rare and consists of descriptions of single case reports or small series. Cures have been reported by using complete surgical resection alone, subtotal resection with radiotherapy, or complete surgical resection with adjuvant radiotherapy. In contrast, intracranial plasmacytomas occurring as initial presentations or spread of multiple myeloma usually have a poor outcome unless new agents are provided. In this report, we describe an unusual case of solitary intraparenchymal brain plasmacytoma treated with a combination of steroids and whole-brain radiotherapy with a boost on the lesion. In our patient, the massive infiltrative pattern of the disease would have hampered the possibility of surgical resection. The treatment was well tolerated and caused no adverse radiation effects. After 48 months of follow-up, the patient was stable and presented only mild cognitive impairment, which did not compromise her daily living activity. Moreover, no progression to multiple myeloma has been documented.

According to our experience, in the rare occurrence of an extensive primary intraparenchymal brain plasmacytoma, whole-brain radiotherapy with a boost on the lesion is capable of affording optimal local control of the disease with no major adverse effects.

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AUTHORS’ DISCLOSURES OF POTENTIAL CONFLICTS OF INTEREST
The author(s) indicated no potential conflicts of interest.

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DOI: 10.1200/JCO.2012.43.0215; published online ahead of print at www.jco.org on October 1, 2012