Atypical monoclonal plasma cell hyperplasia: its identity and treatment

Case report


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Atypical monoclonal plasma cell hyperplasia, like plasma cell granuloma, is an inflammatory pseudotumor. Both are extremely rare in the central nervous system. Atypical monoclonal plasma cell hyperplasia is a recently identified neuropathological entity described by Weidenheim, et al., in 1989. A second case of this disease entity is now reported. The histological findings that differentiate this lesion from plasma cell granuloma, plasmacytoma, and meningioma are discussed. The present case clearly demonstrates the complete resolution of the disease after a course of fractionated radiotherapy.

KEY WORDS • atypical monoclonal plasma cell hyperplasia • central nervous system • plasma cell granuloma • plasmacytoma • radiation therapy

Case Report

This previously healthy 60-year-old woman experienced frequent blurred vision for 6 months. After each attack her vision completely recovered. She had no other associated symptoms such as headache or nausea. Examination by an ophthalmologist showed that the patient had bilateral papilledema and that her visual acuity was 20/100 in the right eye and 20/50 in the left. No visual field deficit was detected during the examination. The results of additional physical and neurological examination were otherwise normal. A magnetic resonance (MR) image of the brain revealed an extraaxial lesion in the posterior fossa, extending supratentorially in line with the posterior interhemispheric fissure (Fig. 1). Admission laboratory studies, including complete blood counts and liver function tests, yielded normal results.

Operation. On December 27, 1994, the patient underwent occipital craniotomy for open biopsy of the lesion. Intraoperatively, the lesion was found to be infiltrating the dura mater and the tentorium. It was fibrous and adhered to the underlying brain parenchyma. It was impossible to achieve total resection and a small piece of lesion was removed for diagnostic purposes.

Pathological Studies. Microscopically, the lesion showed diffusely thickened dura with a dense infiltration of chronic inflammatory cells, consisting of lymphocytes, histio-
cytes, and plasma cells with fibrosis (Fig. 2A). Focally, germinal center formation was seen (Fig. 2B). In some areas, there was a conspicuous excess of plasma cells in the cellular infiltrates (Fig. 2C). However, a monomorphic population of plasma cells or sheets of plasmacytosis were not encountered. Immunohistological staining revealed κ light chain restriction in the plasma cells (Fig. 2D). Because of the monoclonal nature of plasma cell proliferation in a background of mixed cell infiltrates, the diagnosis of atypical monoclonal plasma cell hyperplasia was determined.

Further investigations seeking systemic involvement of hematopoietic proliferation were conducted. Examination of cerebrospinal fluid revealed a protein level that was less than 0.1 g/L, a glucose level of 4.6 mmol/L, and a white blood cell count that was zero; there were no organisms grown on culture including acid-fast bacilli. Serum immunofixation electrophoresis detected immunoglobulin (Ig)G-κ paraprotein. Serum Ig levels were normal. Ultrasonography of the abdomen showed a diffuse fatty change in the liver but there was no focal lesion. No paraaortic/iliac lymphadenopathy was detected. Bone scan revealed multiple areas of increased uptake of isotopes but these had no significant clinical implication. Bone marrow examination revealed normal hematopoietic elements with no excess of plasma cells. The results of all these studies indicated that there was no systemic involvement of plasma cell dyscrasias, making the diagnosis of multiple myeloma unlikely.

Radiation Therapy and Follow-Up Care. The patient received empirical radiation therapy. Her visual symptoms improved. Follow-up MR imaging of the brain was performed 6 months after the conclusion of radiation therapy. The MR imaging demonstrated total resolution of the infiltrate with lymphocytes, plasma cells, and histiocytes. Original magnification × 300. C: Focally, many plasma cells can be seen. They are still intimately intermingled with a mixed cellular infiltrate and adjacent sclerosis. Original magnification × 300. D: Same area as that shown in C, but immunostained for κ (K) and λ (L) light chains. A monoclonal nature for kappa is clearly demonstrated. Original magnification × 300.
Atypical monoclonal plasma cell hyperplasia

The occasional “lymphoid follicle”...

...light...

...we could not observe...

...could be found. Immunocytochemical staining of the lesion showed a monoclonal reactivity of the IgG κ light chain. This latter finding distinguishes atypical monoclonal plasma cell hyperplasia from plasma cell granuloma, which contains polyclonal reactivity of Ig chains. The lesion in our patient exhibited similar histological and immunocytochemical characteristics to the one described by Weidenheim and colleagues, except that our microscopic findings also revealed fibrosis.

Distinct from plasma cell granulomas, intracranial atypical monoclonal plasma cell hyperplasia must also be differentiated from other lesions with plasma cell and lymphocytic infiltration, such as plasmacytoma or meningioma. Our patient is unlikely to have had plasmacytoma because her lesion contained a significant lymphocytic component and germinal centers were present. The MR image of this lesion also resembled an en plaque meningioma, but a cerebral angiogram showed a clear avascular lesion displacing the occipital vasculature anteriorly, which would make an angiographic diagnosis of meningioma unlikely. Histologically, some meningiomas can have a marked plasmalymphocytic infiltrate. However, the absence of meningothelial cells in this case precludes the diagnosis of meningioma.

Atypical monoclonal plasma cell hyperplasia is a very rare entity. The first case was reported in 1989. Two earlier case reports describe similar histological features; however, the diagnosis of plasmacytoma was given. We agree with Weidenheim, et al., that atypical monoclonal plasma cell hyperplasia is a different neuropathological entity and the present case report should help to consolidate this proposal. However, the arguments that there is a spectrum of plasmacytic lesions in the CNS and that atypical monoclonal plasma cell hyperplasia is the preneoplastic stage of solitary plasmacytoma are difficult to prove.

Thus far, most of the plasma cell granulomas and the atypical monoclonal plasma cell hyperplasia reported were in solitary form and total resection was possible. The effectiveness of radiotherapy for plasma cell granuloma has remained uncertain. In our patient, we could not obtain a total excision of the lesion because of its location and adhesion to the underlying brain parenchyma. Complete resolution of the lesion was demonstrated 6 months after radiotherapy. This is the first reported case that clearly demonstrates the effectiveness of radiotherapy in the treatment of CNS atypical monoclonal plasma cell hyperplasia.

References

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