Primary intracranial plasma-cell granuloma

Case report

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The authors report the fourth case of primary intracranial plasma-cell granuloma. The patient was a 16-year-old girl who presented with loss of vision as the major clinical feature. The tumor resembled a meningioma both preoperatively and grossly at surgery. Because the tumor did not respond to steroid treatment following subtotal surgical excision, radiation therapy was administered to the affected area. Major considerations in the differential diagnosis of this neoplasm are discussed.

KEY WORDS • plasma-cell granuloma • brain neoplasm • radiation therapy

Plasma-cell granuloma, a form of idiopathic inflammatory pseudotumor, is a relatively rare entity6,10 that is characterized histopathologically by a benign proliferation consisting predominantly of plasma cells, and including reticuloendothelial elements and intermediate "plasmacytoid" forms.4 Although reported most often in the lungs and conducting airways,4,24,27,28,35,37 these unusual lesions of obscure pathogenesis10 have been described in numerous other organs.1,2,6,11,15,17,31,33,38,41 Plasma-cell granulomas of the central nervous system (CNS) are extremely rare, having been reported only three times in the cranial vault5,25,40 and once in the cervical meninges.7 This report documents the fourth case of a primary intracranial plasma-cell granuloma. Other lesions of major consideration in the differential diagnosis include plasmacytoma, meningioma with extensive plasma-cell and lymphocytic infiltrates, granulomatous inflammations, histiocytosis X, and Wegener's granulomatosis.

Case Report

This previously healthy 16-year-old white girl from Barcelona, Spain, complained of mild occipital headaches, which occurred chiefly at night, and decreased visual acuity. These symptoms had lasted for 1 month, commencing in February, 1986. Visual testing in Spain revealed light perception only in the right eye, while superior and inferior pie-shaped defects were noted in the left eye. Computerized tomography (CT) revealed a homogeneously enhancing lesion in the right posteromedial frontal lobe adjacent to the anterior aspect of the suprasellar cistern, more prominent on the right side (Fig. 1). Carotid angiography showed slight elevation of the right anterior cerebral artery, but no tumor blush was seen. The patient was referred to Buffalo General Hospital for elective resection of a presumed meningioma.

Examination. General physical and neurological examinations at admission were normal, except for visual acuity of 20/400 in the right eye and 20/40 in the left eye. Ocular motility was within normal limits. The ocular media were clear and the optic discs unremarkable. Pupillary reactions were normal. Routine blood tests, urinalysis, serum cortisol, serum follicle-stimulating hormone and luteinizing hormone, and thyroid function tests were all normal. The serum prolactin level was elevated at 11.1 ng/ml (normal ≤ 4.3 ng/ml). Quantitative serum immunoglobulin (Ig) electrophoresis revealed slight elevation of IgM at 337 mg/dl (normal 53 to 323 mg/dl), with normal IgG and IgA levels. Plain x-ray films of the skull were normal.

Operation. On March 14, 1986, a right frontotemporal craniotomy was performed. A tumor was found arising from the dura beneath the anterior clinoid process just medial to the emergence of the intradural segment of the right carotid artery and lateral to the optic foramen. The tumor extended over the floor of
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FIG. 1. Computerized tomography scan showing a homogeneously enhancing lesion in the right posteromedial frontal lobe adjacent to the anterior aspect of the suprasellar cistern.

FIG. 2. Photomicrograph of the tumor specimen showing abundant plasma cells and lymphocytes, moderate numbers of large mononuclear cells, and occasional polymorphonuclear leukocytes. H & E, × 125.

Histopathological Examination. Histologically, the tumor consisted of fibrovascular tissue with heavy infiltrates of plasma cells and lymphocytes, moderate numbers of large mononuclear cells, and occasional polymorphonuclear leukocytes (Fig. 2). The plasma cells appeared reactive in type, with an appreciable number forming Russell bodies. There was no evidence of vasculitis or giant-cell formation. Special stains for fungi, bacteria, and acid-fast organisms, and extensive fungal and bacteriological cultures were negative. Electron microscopic studies revealed a mixture of plasma cells, lymphocytes, and histiocytes, with plasma cells predominating. Birbeck's granules were not seen. Immunoperoxidase studies of the plasma cells and lymphocytes revealed the presence of both kappa and lambda light chains, demonstrating the polyclonal nature of the plasma cells. Based on these studies, a diagnosis of plasma-cell granuloma was made.

Postoperative Course. The patient made a good recovery and was discharged from the hospital on the 6th postoperative day. Because surgical excision was not total, dexamethasone (8 mg daily) was administered on an outpatient basis for 10 days. Repeat visual testing on March 31, 1986, showed marked improvement, with a full visual field in the left eye and a residual inferior nasal field defect in the right eye; visual acuity was 20/25 in both eyes. Clinically, the patient continued to do well with no change on visual testing. However, repeat CT scanning on April 8, 1986, revealed an enhancing lesion involving the posterior aspect of the anterior interhemispheric fissure, extending to the superior aspect of the sella turcica. On October 15, 1986, another CT scan showed enlargement of the mass, with an additional smaller enhancing lesion noted lateral to the midline mass. Despite the increase in the size of the mass, there was no deterioration in vision. At this time, because the tumor did not respond to steroid treatment following subtotal surgical excision, radiation therapy to the area was recommended. Accordingly, in November, 1986, the patient received a total of 2000 roentgens over 10 sessions. In January, 1987, residual tumefaction was still evident on CT studies; however, she is asymptomatic and has normal vision.

Discussion

In 1973, Bahadori and Liebow4 used the term "plasma-cell granuloma" to describe localized benign proliferations in the lungs and conducting airways consisting predominantly of mature plasma cells, with Russell bodies, reticuloendothelial cells, and intermediate "plasmacytoid" forms. These lesions also contain occasional lymphocytes, polymorphonuclear leukocytes, mast cells, eosinophils, and large fat-filled mononuclear cells supported by a stroma of granulation tissue which may contain whorled masses of fibroblasts.4 The term "plasma-cell granuloma" was preferred to other synonyms such as inflammatory pseudotumor, xanthogranuloma, fibroxanthoma, or histiocytoma because of the predominance of plasma cells common to these lesions.5,7,40 The tumor in our case, which consisted of an admixture of polyclonal plasma cells with Russell bodies, histiocytes, lymphocytes, and occasional polymorphonuclear leukocytes and eosinophils, is consistent with the diagnosis of plasma-cell granuloma.

Primary plasma-cell granulomas of the CNS are extremely rare, with only four cases described prior to this
Primary intracranial plasma-cell granuloma

TABLE 1
Summary of clinical data in five reported cases of primary central nervous system plasma-cell granuloma

<table>
<thead>
<tr>
<th>Authors, Year</th>
<th>Age (yrs), Sex</th>
<th>Chief Complaint</th>
<th>Location of Tumor</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eimoto, et al., 1978</td>
<td>37, M</td>
<td>weakness, paresthesia of extremities</td>
<td>intradural extramedullary mass arising from cervical meninges</td>
<td>total excision &amp; irradiation</td>
<td>no recurrence at 9 mos</td>
</tr>
<tr>
<td>West, et al., 1980</td>
<td>17, M</td>
<td>fatigue, dyspnea on exertion, headaches, anemia</td>
<td>posterior fossa</td>
<td>total excision</td>
<td>no recurrence at 8 mos</td>
</tr>
<tr>
<td>Castell, et al., 1983</td>
<td>19, F</td>
<td>diabetes insipidus, panhypopituitarism</td>
<td>hypothalamus, optic chiasma, optic nerves</td>
<td>subtotal excision &amp; irradiation</td>
<td>no recurrence at 40 mos</td>
</tr>
<tr>
<td>Maeda, et al., 1984</td>
<td>36, F</td>
<td>unsteady gait, dysarthria</td>
<td>fourth ventricle</td>
<td>total excision &amp; irradiation</td>
<td>no recurrence at 32 mos</td>
</tr>
<tr>
<td>Cannella, et al., 1988</td>
<td>16, F</td>
<td>loss of vision</td>
<td>anterior fossa</td>
<td>subtotal excision &amp; steroids</td>
<td>recurrence at 1 mo, irradiation</td>
</tr>
</tbody>
</table>

In the absence of a demonstrable granulomatous inflammation of specific etiology, the major considerations in the differential diagnosis should include plasmacytoma, meningioma with extensive plasma-cell and lymphocytic infiltrates, histiocytosis X, and Wegener's granulomatosis. Solitary intracranial plasmacytoma is rare. These neoplastic lesions, characterized by variably atypical plasma cells, are monoclonal in nature. This is an important distinguishing feature from plasma-cell granuloma, which consists of a non-neoplastic proliferation of polyclonal plasma cells. Just as in our case, solitary intracranial plasmacytoma is often mistaken for meningioma preoperatively and at surgery. Although rare, both plasmacytoma and plasma-cell granuloma should be considered when an intracranial mass is encountered in an area where meningiomas commonly arise.

Foci of plasma cells or lymphocytes or both, which are found in meningiomas in a perivascular arrangement or diffusely scattered, are not uncommon, but meningiomas with conspicuous plasma-cell and lymphocytic components are rare. Horten, et al., reported five cases in which light microscopy revealed mixed populations of proliferating meningothelial cells together with well-formed plasma cells and lymphocytes. In our patient, no meningothelial component was identified despite a careful search with both light and electron microscopy. Stam, et al., reported a case similar to those described by Horten, et al., in which immunohistochemical analysis demonstrated the polyclonal nature of the plasma-cell population. One patient reported by Horton, et al., (their Case 1) was noted to have a panhypergammaglobulinemia. Thus, West and coworkers have raised the intriguing possibility that some or all of the tumors in the cases described by Horton, et al., might really be plasma-cell granulomas with incidental inclusions of meningial elements that would require electron microscopic studies for confirmation. Kepes, however, believes these tumors actually represent meningiomas being invaded by lymphocytes and plasma cells in response to an antigenic stimulus from the meningioma tissue itself.

Surgical excision is the primary treatment for the commonly found plasma-cell granulomas of the lung; however, when the lesion is locally aggressive and surgically nonresectable, radiation therapy has been used as an effective alternative. In the second reported case of primary intracranial plasma-cell granuloma (that of Castell, et al.), the patient, also a young woman from Spain, responded favorably to radiation therapy following incomplete surgical excision. In our patient, the lesion recurred rapidly following subtotal surgical excision and steroid administration, and radiation therapy was employed with incomplete regression but apparent cessation of further tumor growth or altered clinical status.

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References


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