Castleman's disease of the leptomeninges

Report of three cases

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Castleman's disease is a rare, benign lymphoproliferative disorder that usually arises in lymph nodes, most commonly in the mediastinum. The authors report the clinical and pathological features of three patients with localized Castleman's disease of the leptomeninges. There were two women, aged 63 and 82 years, and one 25-year-old man. Two patients had progressive focal motor seizures of 3 and 24 months' duration, and the third patient presented acutely with generalized seizures. The clinical diagnosis was meningioma in each case, based on computerized tomography scans, cerebral arteriography, and the operative findings. All three lesions arose in the leptomeninges, compressed the underlying cerebral cortex, and infiltrated the overlying dura to a variable extent. Surgical excision of the tumor resulted in marked clinical improvement in all three patients. Histologically, two cases were classified as the hyaline-vascular type and one as the plasma cell type. Immunohistochemical stains of the latter case revealed a monoclonal population of mature plasma cells. Only a few scattered polyclonal plasma cells were seen in the other two cases. The authors conclude that Castleman's disease involving the leptomeninges is a rare disorder that may mimic meningioma clinically and radiographically.

KEY WORDS • Castleman's disease • angiofollicular lymphoid hyperplasia • meningioma • leptomeninges

Case Reports

Case 1

This healthy 25-year-old white man developed localized motor seizures of the left arm. An electroenceph-
alogram showed mild asymmetry of electrical activity, but a cerebral angiogram was interpreted as normal. The seizures became more frequent over a 2-month period, and then progressed to grand mal seizures despite the use of various oral antiseizure medications. A computerized tomography (CT) scan of the head revealed a round, circumscribed mass in the right parietal region (Fig. 1). A clinical diagnosis of meningioma was made. At surgery, a 6.5-cm subdural mass arising from the leptomeninges was removed in its entirety. The patient had an uneventful postoperative course. A repeat electroencephalogram was normal and no further seizure activity has been reported.

Case 2

This 82-year-old white woman was admitted to the hospital because of nausea, recent episodes of syncope, and amaurosis fugax. She had a 24-month history of focal motor seizures, which were controlled with oral phenytoin therapy. Cerebral angiograms showed a subdural mass in the right parietal region near the central sulcus. A CT scan of the head revealed edema of the adjacent cerebral cortex and a shift in the position of midline structures. A clinical diagnosis of subdural hematoma was made. Right parietal craniotomy revealed a firm, tan-brown mass beneath the dura of the right parietal lobe. The mass measured $5 \times 3 \times 1.2$ cm and was firmly attached to the underlying arachnoid tissue, but it did not infiltrate the cerebral parenchyma. Clinically, the mass was thought to be a meningioma, and it was removed in its entirety. Postoperatively, the patient exhibited slight weakness of the left arm, but subsequently recovered uneventfully with resolution of the nausea, syncopal episodes, amaurosis fugax, and seizures.

Case 3

This 63-year-old white woman experienced progressive right homonymous hemianopsia for approximately 18 months. The patient then developed a generalized seizure that required intravenous phenytoin and diazepam for control. Upon awakening from the seizure, she was confused and agitated. On neurological examination, a dense right visual field deficit and mild right hemiparesis were noted. A CT scan of the head revealed a mass in the left occipital region that extended along the straight sinus, with mild edema of the adjacent cerebral tissues. A clinical diagnosis of meningioma was made. A left occipital craniotomy was performed, and a spongy, lobulated, tan-pink tumor measuring $3.5 \times 2.5 \times 2.0$ cm and arising from the leptomeninges was removed. The overlying dura was intact, and the underlying brain was compressed but not infiltrated. Postoperatively, the patient had a persistent right homonymous hemianopsia, but the other symptoms resolved.

Histological Findings

Cases 1 and 2 had the histological features of the hyaline-vascular type of Castleman's disease. Both tumors consisted of benign lymphoid tissue with scattered lymphoid follicles having small, atrophic germinal centers with radially penetrating blood vessels. Some germinal centers were surrounded by concentric layers of small lymphocytes (Fig. 2 left). The follicles were separated by fibrotic tissue that was infiltrated by numerous small lymphocytes and scattered immunoblasts and plasma cells. Interfollicular microvascular proliferation was prominent (Fig. 2 right). Immunoperoxidase stains performed on paraffin-embedded tissue showed that the plasma cells were polyclonal.

The tumor in Case 3 consisted of benign lymphoid tissue with scattered lymphoid follicles, similar to those described above. However, the interfollicular areas were infiltrated by large clusters and sheets of mature-appearing plasma cells and only scattered small lymphocytes, corresponding to the plasma cell type of Castleman's disease (Fig. 3). Immunoperoxidase stains performed on paraffin-embedded tissue showed the plasma cells to contain monoclonal cytoplasmic immunoglobulin G of the kappa light chain type.

Discussion

While most authors agree that the plasma cell and hyaline-vascular types of Castleman's disease represent histological variants of the same basic disease process, an understanding of the etiology of this rare disorder is presently lacking. Keller, et al., suggested that Castleman's disease was a chronic inflammatory process, possibly of infectious origin. The clinical manifestations usually associated with the plasma cell type of Castleman's disease, such as fever, sweating, chronic anemia, leukocytosis, thrombocytosis, splenomegaly, bone marrow plasmacytosis, and occasional multicentric adenopathy, also support an inflammatory etiology. Some authors believe that the unusual histological appearance of the hyaline-vascular type, the absence of systemic symptoms, and the occurrence of lesions at
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FIG. 2. Photomicrographs of Castleman’s disease of the hyaline-vascular type.  
Left: A lymphoid follicle is shown with a hyalinized germinal center and radially penetrating blood vessels, surrounded by a concentric layer of small lymphocytes. H & E, × 10.  
Right: An interfollicular area is shown with small lymphocytes and numerous hyperplastic small blood vessels. H & E, × 20.

sites where lymphoid tissue is usually not found favor a hamartomatous process.2 However, the two types overlap clinically and histologically.7,20 Some patients with the plasma cell type, such as in Case 3, lack the usual systemic signs and symptoms, whereas cases of the hyaline-vascular type may be multicentric and have systemic manifestations.13,14,19,20 Although the fact that lymph nodes are not normally present within the central nervous system might favor a hamartomatous process, the advanced age of two of the patients is more in keeping with a chronic inflammatory process.

Localized Castleman’s disease is generally considered to be a benign condition and is usually cured by surgical excision of the mass19 or by radiotherapy.19 The use of corticosteroids or systemic chemotherapy for the multicentric variant may be of benefit in some cases.7,17,20 The presence of a monoclonal population of plasma cells in Case 3 suggests a malignant potential. Indeed, the evolution of Castleman’s disease to plasmacytoma and malignant lymphoma has been reported.7,16,20

All three of the patients in this report had clinical symptoms that were attributed to a slowly expanding tumor mass. Based on CT scans, cerebral angiography, and the operative findings, the clinical diagnosis was meningioma in each case. Rare cases of meningioma with a prominent lymphocytic and/or plasmacytic component1,8 and other rare cases of meningeal masses with the features of “sinus histiocytosis with massive lymphadenopathy”11,12 have been reported previously. In such cases, however, the tumors have consisted of a variable mixture of meningotheelial and inflammatory cells. In contrast, the three cases reported here had the typical histological features of Castleman’s disease without a meningotheelial component. In all three patients, the symptoms were alleviated by surgical excision of the mass, and there has been no evidence of recurrent disease during a short clinical follow-up period. The authors conclude that Castleman’s disease of the leptomeninges is a rare disorder that may mimic meningioma both clinically and radiologically.

FIG. 3. Photomicrograph of the excised mass in Castleman’s disease of the plasma cell type. There is an interfollicular cluster of mature-appearing plasma cells. H & E, × 40.
References


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