Sinus histiocytosis with massive lymphadenopathy: A case of multiple dural involvement

Abstract An exceptional case of Rosai-Dorfman disease (sinus histiocytosis with massive lymphadenopathy) arising from the meninges in a 60-year-old Japanese man is presented. Computerized tomographic scans and magnetic resonance images demonstrated well-circumscribed tumorous lesions that were homogeneously enhanced with contrast medium. Systemic examination revealed no abnormalities except for a cervical lymphadenopathy and diabetes mellitus. Microscopic examination of the resected specimens showed proliferated histiocytosis and infiltration of plasma cells and lymphocytes. The histology was characterized by the presence of histiocytes demonstrating lymphophagocytosis and immunoreactivity for S-100 protein staining. Immunohistochemical studies and electron microscopy were useful in confirming the diagnosis. The clinical and histopathological features of this disease are discussed.

Key words Emperipolesis · Lymphadenopathy · Rosai-Dorfman disease · Meninges · Sinus histiocytosis with massive lymphadenopathy

Introduction

Sinus histiocytosis with massive lymphadenopathy (SHML) (Rosai-Dorfman disease) is a rare idiopathic entity that is characterized histopathologically by proliferation of mature histiocytes, which show lymphophagocytosis, and infiltration of plasma cells and lymphocytes. Rosai and Dorfman first described this disease in 1969, and over 400 cases with various features of this condition have been identified. Most of the patients had cervical lymphadenopathy. Extranodal SHML may occur as part of a generalized process involving lymph nodes or may involve extranodal sites independent of lymph node status. Forty-three percent had at least one site of extranodal SHML. SHML involving the central nervous system (CNS) has been identified in 28 cases, and the lesions were always in close relation to the meninges. We describe a case of SHML with multiple dural involvement resembling meningioma.

Clinical summary

A 60-year-old Japanese man, medically treated for diabetes mellitus during the past 10 years, presented with a 2-month history of tinnitus.

Examination

Neurological examination revealed tinnitus and hearing disturbance in the left ear. There was a painless lymphadenopathy on the left side of the neck. Laboratory evaluation of blood and cerebrospinal fluid showed no abnormalities. A computerized tomographic (CT) scan of the head disclosed well-defined high-density masses in the left cerebellopontine angle region, the right temporal fossa, the right temporal convexity, the right frontal convexity, and the left frontal convexity, which were homogeneously en-
hanced with contrast material (Fig. 1). Magnetic resonance (MR) images demonstrated dural-based enhanced masses after administration of gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA) (Fig. 2), which were hyperintense to white matter on T₁-weighted images and hypointense on T₂-weighted images. In addition, Gd-DTPA-enhanced MR images showed the enhanced dura mater around the mass in the left frontal convexity (Fig. 2c,d). Cerebral angiography documented avascular lesions. There were no bone abnormalities on plain X-ray films. MR images of the spinal region disclosed nothing abnormal. The preoperative diagnosis was multiple meningioma.

Operation

On December 3, 1992, the mass in the left frontal convexity was removed via a left frontotemporal craniotomy. It was adherent to the inner surface of the dura and demarcated from the brain. The overlying bone was not involved. After the operation, administration of steroid resulted in a slight decrease in the size of the cervical lymphadenopathy; however, the size of the intracranial masses did not decrease and the patient's symptoms remained. On December 25, 1992, the mass in the left cerebellopontine angle region was totally removed via a left suboccipital approach. On January 12, 1993, other masses at the right temporal base, the right frontal convexity, and the right temporal convexity were totally removed, and the enlarged lymph node at the neck was also removed.

Pathological findings

Macroscopically, the mass was firm, elastic, and white to yellow in appearance (Fig. 3a). It looked like an intracranial meningioma. The surgical specimens were fixed in formalin and embedded in paraffin. Light microscopically, the meningeal lesions showed proliferation of histiocytes with intense infiltration by plasma cells and lymphocytes (Fig. 3b). In some areas, the plasma cells predominated over the histiocytic proliferation. Stromal fibrosis was present. The histiocytic cells had abundant, ill-defined, and occasionally foamy cytoplasm, with one or more round to oval, vesicular nuclei containing small nucleoli. Well-preserved lymphocytes were occasionally found in the cytoplasm of several histiocytes (Fig. 3c), a phenomenon referred to as lymphophagocytosis or emperiploisis. In addition to the aggregates of histiocytes, lymphoid cell clusters were found. The lymphocytes had no atypia. Russell bodies were occasionally observed within the cytoplasm of the plasma cells. No eosinophils were identified. There were scarcely mitotic figures, and no invasion of the dura mater nor the adjacent cerebral cortex. Meningothelial cells, whorl formation, and psammomatous bodies were absent. The histopathological appearance of the neck lymph nodal lesion was similar to those seen in the meningeal lesions. Lymph sinuses were expanded, with numerous inflammatory cells composed of histiocytes, plasma cells, and lymphocytes. Reactive germinal centers were not seen. There was necrosis and marked fibrosis of the lymph node capsule; the intranodal fibrosis was, however, minimal compared with the meningeal le-