Intracranial Cystic Hemangiopericytoma
—Case Report—

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Abstract

A rare case of intracranial hemangiopericytoma associated with a large cyst was treated by gross total removal and local irradiation. The tumor has not recurred for 16 months, although the effectiveness of radiation therapy for hemangiopericytoma is unclear. Histological examination of the tumor specimen showed aggregation of the microcystic components, possibly contributing to the cyst formation. Hemangiopericytoma should not be classified as meningioma because of the different neoplastic and cytological properties. Complete surgical removal is essential for this tumor because of its malignancy.

Key words: intracranial hemangiopericytoma, cyst formation

Introduction

Intracranial hemangiopericytomas originating in the meninges are rare, and cystic hemangiopericytomas are even rarer. We recently encountered a hemangiopericytoma associated with a large cyst originating in the cerebellar tentorium. The possible mechanisms of cyst formation are discussed, with a review of the literature.

Case Report

A 38-year-old male began to suffer from syncopal attacks in spring, 1988. He was admitted to our hospital with generalized convulsion on March 9, 1989. On admission, he showed no abnormal neurological signs. However, computed tomographic (CT) scans showed a supra- and infratentorial brain tumor associated with a large supratentorial cyst. The tumor was highly and homogeneously enhanced, and part of the cyst wall was also enhanced (Fig. 1). Gadolinium-enhanced magnetic resonance (MR) images showed both the tumor and a portion of the cyst to be highly enhanced (Fig. 2). Intra-arterial digital subtraction angiography (DSA) revealed a tumor stain fed by the right occipital artery from the arterial phase (Fig. 3). The diagnosis was cystic meningioma.

On March 28, an operation was performed using the combined supra- and infratentorial approach. The tumor originated in the cerebellar tentorium in contact with the transverse sinus and had grown supra- and infratentorially. The tumor was grossly totally removed, and the dural adhesion near the transverse sinus was electrocoagulated carefully. The cyst wall was also removed, excluding the part adhering tightly to the brain. The cyst contained a...
xanthochromic fluid.

Light microscopic examination of the tumor specimen showed fusiform-shaped cells with indistinct margins. The nuclei were clear with scanty chromatin. Formation of vascular lumens and aggregation of microcystic components were present in some places. Gitter stain showed well-developed reticulin fibers (Fig. 4). Tumor cells were also present in the cyst wall.

Postoperative postcontrast CT scans found no residual tumor (Fig. 5). Local irradiation of 50 Gy was given to prevent tumor recurrence. He was discharged on May 12, but is being followed as an outpatient. No tumor recurrence was observed up to 16 months after the postoperative radiation therapy.

Discussion

Hemangiopericytomas are rare in the central nervous system, with an incidence of 1-3.8% of all primary intracranial meningeal tumors.9,14,15,24)
Hemangiopericytomas originating in the intracranial meninges were first reported by Begg and Garret, and Fisher et al. Some authors have classified this tumor as an angioblastic meningioma, but others consider it is distinct from meningioma and represents a type of vascular neoplasm. This question remains unresolved.

Hemangiopericytomas frequently recur and metastasize extracranially in contrast to ordinary meningiomas. Goellner et al. reported local recurrence in 80% and extracranial metastasis in 23% of 26 patients with hemangiopericytomas. We consider that hemangiopericytomas should not be classified as meningiomas, because the electron microscopic appearance is very different to meningiomas and the incidence of recurrence and extracranial metastasis is much higher.

The incidence of cystic hemangiopericytoma is unclear, because of few definite diagnoses of this tumor by CT scanning. Blank et al. reported a pediatric case of cystic hemangiopericytoma, and Arita et al. reported a hemangiopericytoma containing necrotic, microcystic components and a macrocyst containing xanthochromic fluid. Possible mechanisms of cyst formation in meningioma include: 1) central degeneration and necrosis of the tumor due to insufficient blood flow, 2) exudation of plasma components, 3) intratumoral bleeding, 4) reforming process of the subarachnoid spaces, and 5) aggregation and enlargement of microcysts. In our case, some microcystic components were present, so we speculate that the aggregation and enlargement of microcysts were the contributing factors.

The effectiveness of radiation therapy for hemangiopericytoma has not been established. Some authors believed it to be efficacious, while others questioned its effectiveness. We consider the radiation therapy was effective in this case, since there was no tumor recurrence for 16 months after postoperative local irradiation. Complete surgical removal is essential for treatment of hemangiopericytoma, because the effect of radiation therapy remains unclear and the incidence of local recurrence is extremely high.

References


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