EMIFACIAL spasm is most commonly caused by compression of the facial nerve RExZ by an aberrant vessel, usually a branch of the AICA, PICA, or vertebral artery. Rarer causes of compression include posterior circulation aneurysms, vascular malformations, various types of tumors, and cysts. Intracranial ependymal cysts are rare lesions and occur less frequently than arachnoid cysts. Zehnder reported the first case in 1938, and Friede and Yasargil reviewed the literature and collected 15 cases in 1977. The cysts are generally located in the supratentorial cerebral parenchyma, but are occasionally present in the intraventricular region, subarachnoid space, spine, or midbrain. In this article we report the histological and immunohistochemical findings in a rare case in which an ependymal cyst of the CPA caused hemifacial spasm.

Hemifacial spasm associated with an ependymal cyst in the cerebellopontine angle

Case report

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No previous case of hemifacial spasm associated with an ependymal cyst has been reported in the literature. In this article the authors report the first case in which hemifacial spasm accompanied an ipsilateral cerebellopontine angle ependymal cyst in a 27-year-old woman. Cyst fenestration and arterial decompression of the facial nerve at the root exit zone resulted in complete resolution of the patient’s symptoms. A histopathological study including immunohistochemical methods identified an ependymal cyst.

KEY WORDS • hemifacial spasm • ependymal cyst • microvascular decompression • cerebellopontine angle • posterior fossa

Case Report

History. This 27-year-old woman, with no other significant medical history, complained of twitching of the right facial muscles, which had been occurring for 3 years. The spasm originated in the orbicularis oculi muscle and extended to the lower part of the face. Before being referred to us, this woman had been treated with tranquilizers for several months without obvious benefit.

Examination. Neurological examinations revealed no deficits except for the right hemifacial spasm. Computerized tomography scans revealed a homogeneous lesion in the right CPA with a density similar to that of CSF. Metrizamide-enhanced CT cisternography demonstrated slow filling and slow clearance of contrast medium in the right CPA lesion. Magnetic resonance images demonstrated an extraaxial mass in the right CPA. The lesion was isointense with CSF on T1-weighted, T2-reversed, and diffusion-weighted images (Fig. 1). Vertebral artery angiography disclosed elongation of the right PICA. The diagnosis was vascular crosscompression of the facial REXZ associated with a right CPA cyst.

Operation. A right retromastoid craniotomy was per-

Abbreviations used in this paper: AICA = anterior inferior cerebellar artery; CPA = cerebellopontine angle; CSF = cerebrospinal fluid; CT = computerized tomography; GFAP = glial fibrillary acidic protein; MR = magnetic resonance; PICA = posterior inferior cerebellar artery; REXZ = root exit zone.
formed. After incision of the dura mater and gentle displacement of the cerebellar hemisphere, we uncovered a cyst with a thick, gray wall that was located between the internal auditory canal and the jugular foramen (Fig. 2). Excision of the outer wall of the cyst caused clear CSF to escape. At this time the abnormal muscle response in the orbicularis oculi and mental muscles did not disappear. The outer and inner walls of the cyst were partially resected for histological examination. The AICA, PICA, and seventh through 10th cranial nerves were found, as shown in Figs. 2 right and 3 left. After the AICA and PICA had been mobilized and held away from the RExZ, by gluing the AICA into the pyramis and inserting small pieces of Teflon felt between the pons and the PICA (Fig. 3 right), the abnormal muscle response disappeared.

Postoperative Course. Immediately after the operation, the spasm disappeared completely. The patient remains asymptomatic and has shown normal facial strength and function for 10 months.

Pathological Findings. A light microscopy examination revealed that the cyst wall was lined by a single layer of cuboidal cells that rested on glial tissue without a basement membrane (Fig. 4A). Immunohistochemical studies revealed that the surface-lining cuboidal cells were positive for epithelial membrane antigen (not shown) and GFAP (Fig. 4B), and were negative for cytokeratin (not shown). The glial layer was also positive for GFAP (Fig. 4B) and was demarcated from the fibrous tissues by a continuous basement membrane that was positive for laminin (Fig. 4C). The diagnosis of an ependymal cyst was made.

Discussion

Ependymal Cysts of the Posterior Cranial Fossa

Various kinds of cysts can occur in the posterior cranial fossa. In origin, they can be epithelial, such as endodermal (neurenteric) or neuroepithelial (ependymal and choroidal epithelial) cysts; mesenchymal, such as arachnoid cysts; or mixed, such as teratomatous cysts. Nevertheless, CPA ependymal cysts appear to be quite rare. To our knowledge, only two reports of this entity have been published and there are no cases in the literature in which hemifacial spasm was associated with an ependymal cyst.

Ependymal cysts are neuroepithelial cysts that are typically found in the central white matter of the temporoparietal and frontal lobes. These cysts are most often found within the parenchyma, and rarely, in the subarachnoid or intraventricular regions. The protein content of the cyst fluid is generally greater than that of the CSF; on MR images the cyst will typically appear isointense to CSF on T1-weighted images and isointense or slightly hyperintense to CSF on T2-weighted images and isointense or slightly hyperin-
tense on T2-weighted images. Clinical symptoms usually result from neurological deficits referable to these lobes, from seizures, or from chronic headaches associated with increased intracranial pressure. The diagnosis of an ependymal cyst is appropriate for cysts in which the epithelial lining rests directly on the brain parenchyma or a layer of astroglia, rather than a basement membrane and connective tissue.

Ependymal cysts display immunoreactivities for GFAP and S-100 protein, which are expressed by the normal ependymal epithelium. Immunoreactivity for epithelial membrane antigen has also been described in the normal ependymal epithelium. In our case, the histological and immunohistochemical features were those of an ependymal cyst.

Hemifacial Spasm

Hemifacial spasm is a symptom of hyperactive dysfunction usually caused by vascular compression of the facial nerve at the RExZ. It has been shown by many authors that the spasm is relieved by vascular decompression of the RExZ. Jannetta suggested that, as a result of the aging process, arterial elongation might compress the RExZ of the facial nerve and might alter the relationship between the nerve and the vessels. Hemifacial spasm most commonly occurs in middle age, typically during the fifth and sixth decades of life, and is extremely rare in young persons. Rarer causes of hemifacial spasm include aneurysms and vascular malformations as well as various mass lesions. In most reported cases the RExZ of the facial nerve was directly compressed by the lesions; however, Higashi and colleagues reported that an arachnoid cyst in the CPA produced a deviation in the PICA, causing it to contact the RExZ of the facial nerve. They performed not only cyst fenestration but also arterial decompression, resulting in complete disappearance of the hemifacial spasm. In our relatively young patient, the offending AICA and PICA might have been moved by the ependymal cyst to compress the RExZ of the facial nerve. Both cyst fenestration and arterial decompression resulted in complete relief of the hemifacial spasm, similar to the case described by Higashi and colleagues.

References

Hemifacial spasm with ependymal cyst


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