Intraventricular Fibroma: MR and Pathologic Comparison

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Summary: This case report describes clinical, surgical, MR, and pathologic findings of an intraventricular fibroma, a rare, benign mesenchymal neoplasm. Relatively isointense with normal brain parenchyma on T1-weighted images, the tumor exhibited hyperintense signal on T2-weighted pulse sequences and enhanced intensely and homogeneously after intravenous administration of gadopentetate dimeglumine. Choroid plexus hyperintense signal on T2-weighted pulse sequences and pathologic findings of an intraventricular fibroma, a rare, benign brain parenchyma on T1-weighted images, the tumor exhibited mesenchymal neoplasm. Relatively isointense with normal chymal neoplasms (1, 2). In 1985, Palma et al reported a case of an intracerebral fibroma and reviewed the literature (2). This report describes an intraventricular fibroma in a child. We report clinical, surgical, and magnetic resonance (MR) findings as well as the pathologic features by immunohistochemistry and electron microscopy.

Case Report

An 11-year-old girl presented with a 6-week history of intermittent vomiting and headaches. Physical examination was normal. MR revealed a 3-cm mass in the trigone of the right lateral ventricle. It was relatively isointense with the adjacent normal brain parenchyma on T1-weighted images, 1000/17/2 (TR/TE/excitations) (Fig. 1A) and of hyperintense signal intensity on T2-weighted images (2500/90/1) (Fig. 1B). The portion of the right lateral ventricle posterior to the mass was "trapped" as demonstrated by dilatation and signal intensity differing from the rest of the cerebrospinal fluid, probably due to its higher protein content. The ventricles were otherwise normal in size and signal intensity. Edema was noted in the brain parenchyma surrounding the mass. The mass enhanced intensely after intravenous injection of gadopentetate dimeglumine (Fig. 1C).

A choroid plexus papilloma was considered the most likely diagnosis preoperatively. At surgery, a firm, highly vascular encapsulated tumor within the trigone of the right lateral ventricle was removed. The tumor was intimately related to adjacent thickened, matted choroid plexus, and the trigone of the right lateral ventricle was obstructed. Analysis of fluid withdrawn from the "trapped" portion of the ventricle revealed an extremely elevated protein content, confirming the suspected etiology of the hyperintense appearance on T1-weighted images. Intraoperative frozen sections suggested benign fibrous and vascular tissue.

Microscopic examination revealed a mesenchymal-type neoplasm with rich vascularity. No significant mitotic activity, tumor necrosis, or nuclear pleomorphism was found. Special stains demonstrated no glial component. The initial diagnosis was angiofibromatous meningioma because of these findings. However, this tumor lacked the ultrastructural features of meningothelial or arachnoid cap cells, being composed of collagen-producing fibroblasts of varying degrees of maturation without interdigiting processes or papillary structures seen in choroid plexus or ependymal elements. The final diagnosis was benign fibroma with chronic inflammatory change and focal dystrophic calcification. The mass appeared to arise from the choroid plexus stroma.

Following resection, the patient has undergone clinical examination every 6 months and is doing well without symptoms or neurologic deficit 2 years after removal of the neoplasm. Two postoperative MR scans reveal postoperative change with no evidence of recurrence.

Discussion

True intracerebral fibromas are rare neoplasms that may be difficult to differentiate histologically from meningiomas. Indeed, this neoplasm was inseparable from an angiofibromatous meningioma by immunohistochemistry and required ultrastructural techniques for the final diagnosis. The rarity of intracerebral fibromas is usually attributed to the paucity of connective tissue within the brain. It has been noted that fibroblastic elements within the central nervous system tend to produce ma-
Fig. 1. Intraventricular fibroma.
A, Axial T1-weighted MR image (700/20/2), reveals a heterogeneous 3-cm mass within the right lateral ventricle (curved arrow) with a dilated "trapped" trigone (straight arrow). Note the increased signal intensity of the trapped portion secondary to increased protein content in the cerebrospinal fluid.
B, Edema (arrow) surrounds the mildly hyperintense mass and dilated ventricle on an axial T2-weighted MR image (2500/90/1).
C, Parasagittal T1-weighted MR image (700/20/2) after intravenous administration of gadopentetate dimeglumine demonstrates intense enhancement of the mass (arrow).
D, A representative area of benign fibroblastic proliferation (black arrows) with scattered chronic inflammatory cells (white arrows) present. The high collagen content was confirmed ultrastructurally and with the various immunohistochemical stains. No glial, papillary, choroid plexus, or ependymal elements were identified. (Original magnification: X410).

Lignant (fibrosarcomas) rather than benign neoplasms (2, 3). Their exact cell of origin is unknown but it has been speculated that these neoplasms could arise from perivascular connective tissue, the pia-arachnoid, or dural fibroblasts located in the cerebrum (4). The majority (75%) of the reported intracerebral fibromas have occurred in the first 2 decades of life, as in our case (2).

Other more common lesions that can demonstrate an appearance identical to this case include choroid plexus papilloma/carcinoma, meningioma, ependymoma, and subependymal giant cell astrocytoma.

References