Gliofibroma: CT and MRI

Abstract We describe CT and MRI appearances in two children with pathologically proven gliofibromas, in the cerebrum and cerebellum. A striking finding was lack of high signal on T2-weighted MRI.

Key words Brain, neoplasms · Gliofibroma · Magnetic resonance imaging

Introduction

Gliofibroma is an extremely rare bimorphic neoplasm composed of both glial and mesenchymal components [1, 2], whose biologic behavior is variable [1]. Gliofibromas have been reported to occur throughout the central nervous system (CNS), but imaging experience with these tumors is limited. We describe the imaging findings in two pathologically proven cases.

Case reports

Case 1

An 8-year-old boy was seen for abdominal pain, nausea, and vomiting. Abdominal ultrasound and upper endoscopy were normal. Three months later, the parents reported that he was experiencing paranoid spells consisting of frightened feelings, disorientation, confusion, dizziness, and visual hallucinations (dark shadows emerging from open desks at school). EEG showed left anterior temporal and right cerebral hemisphere spikes. The patient was given carbamazepine for complex partial seizures. He also complained of occasional headaches and double vision. He had slight anisocoria (left pupil 1 mm larger than the right) but examination was otherwise normal. CT of the head showed multiple contrast-enhancing mass lesions. The presumed diagnosis was infection.

On arrival at our center, the patient was afebrile. A chest radiograph was normal. White blood cell count was slightly elevated at 14.3 g/dl. Cerebrospinal fluid (CSF) gram stain, bacterial, fungal, and tuberculosis cultures, VDRL, cryptococcal antigen, and cytology were all normal. Human immunodeficiency virus was nonreactive. MRI confirmed the multiple intracranial masses (Fig. 1).

The patient underwent open biopsy of the left anterior temporal lobe lesion. The tumor was firm, smooth and pale tan. There was no evidence of hemorrhage. The bulk of the tumor was composed of collagen (Fig. 3a) and reticulin fibers; only focal areas contained glial fibrils. Two cell types were found within this extracellular matrix meshwork. The most common were elongated or spindle-shaped cells, devoid of cell processes. In most regions, these were sparse, but in some they were densely packed (Fig. 3b). Numerous mitotic figures were observed within these hypercellular zones. These cells showed negative immunostaining for glial
Fig. 1a–c Case 1. a T1-weighted spin-echo axial image demonstrates low-signal masses in the right aspect of the midbrain and the right temporal region. The temporal mass was shown to be within the choroidal fissure on a coronal section. b T2-weighted image shows that the masses give low signal, with surrounding edema. c A T1-weighted contrast-enhanced coronal image demonstrates that there are many masses which enhance intensely. Tumor nodules stud the superior surface of the cerebellum. Enhancement is also present along the tentorium (arrow). Intraventricular and subependymal tumor is shown (open arrows).

Fig. 2 Case 1. Gross pathology specimen of right temporal mass shows a firm, slightly lobulated, pale tan tumor fibrillary acidic protein (GFAP), S-100 protein, neuron specific enolase (NSE), and synaptophysin. Under the electron microscope, these cells appeared isolated, surrounded by collagen fibrils. Their cytoplasm contained abundant, rough, endoplasmic reticulum and numerous intermediate-type filaments. No basal lamina was observed around them (Fig. 3c). The second cell type was also rather sparse, although occasionally they formed small clusters. They were quite large cells, ranging from 10 to 40 μm in diameter, containing numerous cell processes (Fig. 3d) and one or two eccentric nuclei. They showed strong immunoreactivity for GFAP and S-100 protein, but none for NSE or synaptophysin. Synaptophysin staining also revealed the presence of clusters of cell processes with intense immunoreactivity, but no cell bodies were ever stained. This correlated with the electron microscopic observations in which neurites and synapses were occasionally seen, but no cell bodies could be found. We believe these represent pre-existing nerve elements of normal brain tissue entrapped by the tumor. The diagnosis was gliofibroma.

MRI of the spine demonstrated subarachnoid dissemination of the tumor (Fig. 4). The patient is currently receiving aggressive chemotherapy and the tumor foci have shown marked decrease in size and number.

Case 2
A 6-month-old girl’s mother noticed increasing head size, bulging soft spot, loss of head control, decreased crying, and an altered sleep pattern. Examination showed a head circumference of 49 cm (> 95%) with prominent scalp veins, a bulging anterior fontanelle, and bilateral papilledema. CT showed a large, midline, posterior cranial fossa mass causing obstructive hydrocephalus (Fig. 5). The infant was placed on steroids and acetazolamide. MRI showed a large mass arising from the inferior vermis, displacing the fourth ventricle anteriorly and superiorly, causing obstructive hydrocephalus, with periventricular interstitial edema (Fig. 6a). The mass was isointense with gray matter on both T1- and T2-weighted images (Fig. 6a, b). The preliminary diagnosis was medulloblastoma.

The tumor was resected in its entirety. The CSF showed no tumor cells. A ventriculostomy catheter was placed. Grossly the tumor consisted of a white firm and a soft red portion. Histologic examination revealed two separate cell types, glial and mesenchymal, neither of which showed anaplastic features. Swirled cells with elongated fibrillar processes which were strongly GFAP-positive were interrupted in places by nests of whorled cells or clusters of plump cells resembling gemistocytes. An ependymal covering was present along a portion of the surface of the tumor and fragments of choroid plexus were intimately associated with the tumor. There was some glial tissue beneath the ependyma, suggesting that the tumor originated from the subependymal region. The diagnosis was gliofibroma.

Postoperative CT demonstrated no evidence of residual tumor and undilated ventricles. The patient was discharged and is currently being followed.

Discussion
Bimorphic neoplasms of mixed mesenchymal and glial elements are rare [1, 2]. The most common is a gliosarcoma, which is thought to arise from a high-grade