Diffuse fibrillary astrocytoma

Two diffuse astrocytoma variants have been deleted from the World Health Organization Classification of Tumors of the Central Nervous System 2016: protoplasmic astrocytoma, a diagnosis that was previously defined in only vague terms and is almost never made any longer given that tumors with this histological appearance are typically characterized as other more narrowly defined lesions; and fibrillary astrocytoma, since this diagnosis overlaps nearly entirely with the standard diffuse astrocytoma. As a result, only gemistocytic astrocytoma remains as a distinct variant of diffuse astrocytoma IDH-mutant.

Fibrillary astrocytomas also called low grade or diffuse astrocytomas, are a group of primary slow-growing brain tumors. They typically occur in adults between the ages of twenty and fifty.

Fibrillary astrocytomas arise from neoplastic astrocytes, a type of glial cell found in the central nervous system. They may occur anywhere in the brain, or even in the spinal cord, but are most commonly found in the cerebral hemispheres. As the alternative name of “diffuse astrocytoma” implies, the outline of the tumor is not clearly visible in scans, because the borders of the neoplasm tend to send out tiny microscopic fibrillary tentacles that spread into the surrounding brain tissue. These tentacles intermingle with healthy brain cells, making complete surgical removal difficult. However, they are low grade tumors, with a slow rate of growth, so that patients commonly survive longer than those with otherwise similar types of brain tumor, such as glioblastoma multiforme.

Case series

Age and histologic grade are interrelated characteristics of diffuse fibrillary astrocytomas, because the peak age incidence rises with increasing grade. The relationship between age and grade may be explained if age determines the rate of anaplastic progression in astrocytomas.

Shafqat et al., tested this hypothesis by determining the interval between diagnosis of low grade astrocytoma and progression to high grade astrocytoma in patients of various ages. A two-way scatterplot of age at initial diagnosis versus interval to anaplastic progression demonstrated a strong negative correlation ($n = 24$; Pearson correlation coefficient = -0.83; Spearman correlation coefficient = -0.79; $p < 0.001$ for both values). It was concluded that the rate of anaplastic progression in low-grade astrocytoma is directly correlated with patient age [1)].

Case reports

A 21-month-old patient had developed feeding difficulty and reactive airway disease at approximately 8 months of age. MRI showed a diffuse, nonenhancing tumor in the CM region. Following radical resection, and an unremarkable perioperative course, he aspirated, developed pulmonary insufficiency and expired. Postmortem examination revealed a low-grade diffuse fibrillary astrocytoma extending from C6 to the medulla. The medullary portion arose in a paramedian location and infiltrated dorsally into the fourth ventricle, the obex, the leptomeninges, and the adjacent cerebellum. This case demonstrates the growth pattern of a distinct subset of CM tumors that behave in a manner similar to intrinsic diffuse BST. Future identification of these subsets by a careful analysis of the clinical presentation and MRI images will enable better operative planning and optimal

management [2].

1)
Shafqat S, Hedley-Whyte ET, Henson JW. Age-dependent rate of anaplastic transformation in low-grade astrocytoma. Neurology. 1999 Mar 10;52(4):867-9. PubMed PMID: 10078745.
2)
Squires LA, Constantini S, Miller DC, Epstein F. Diffuse infiltrating astrocytoma of the cervicomedullary region: clinicopathologic entity. Pediatr Neurosurg. 1997 Sep;27(3):153-9. PubMed PMID: 9548526.