WOMAN AGED 24 YEARS WITH FOURTH VENTRICULAR MASS

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CLINICAL HISTORY

A woman aged 24 years presented with ataxia, visual obscurations, and headaches over 4 weeks. She had no significant past medical history, and was taking no medication. MR imaging revealed an enhancing mass in the fourth ventricle, which was removed at neurosurgery. Macroscopic examination revealed soft gray tissue measuring 10 × 5 × 5 mm and several fragments of beige/gray tissue measuring 5 × 5 × 4 mm were submitted for histopathological examination.

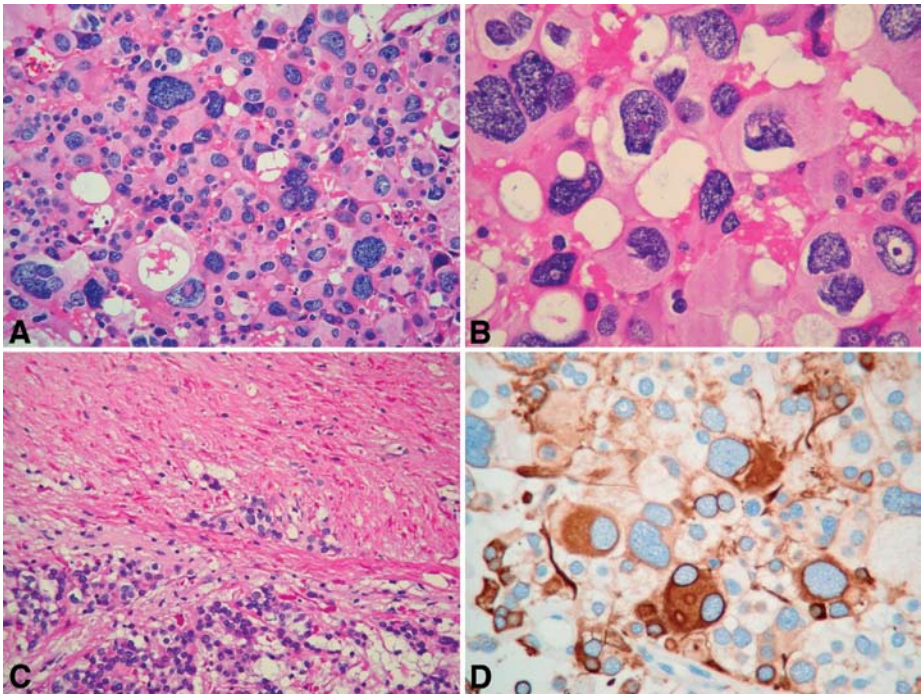

Figure 2.

MICROSCOPIC PATHOLOGY

Microscopy showed a tumor consisting of sheets of cells interrupted by a network of capillaries and one focus of necrosis. Additional architectural features were scattered rosettes and canalicular structures (Arrow, Figure 1), but there were no convincing pseudorosettes. Cytologically, there was a striking dimorphism manifesting either as groups of uniform cells with round or oval nuclei and indistinct cytoplasmic borders (Figure 1), or as areas where giant tumor cells exhibited gross pleomorphism and well-defined cytoplasmic borders (Figure 2A, B). Both areas were occasionally crossed by fascicles of spindle-shaped cells with fibrillary processes, around which were scattered Rosenthal fibers (Figure 2c). Reticulin was sparse.

Immunohistochemistry demonstrated that most tumor cells were GFAP-positive (Figure 2d). No reactivity was detected with neuronal antibodies, anti-cytokeratin antibodies, or antibodies to epithelial membrane antigen. The maximum Ki-67 labeling index was less than 10%.

Ultrastructural examination revealed microvilli and cilia, as well as the presence of complex intercellular zipper-like junctions (Figure 3).

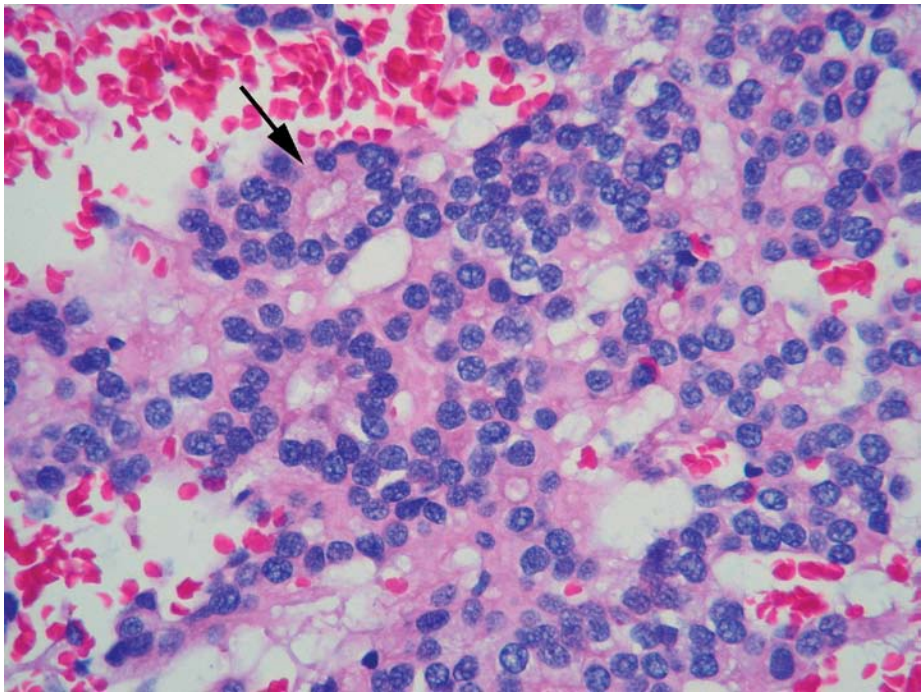

Figure 1.

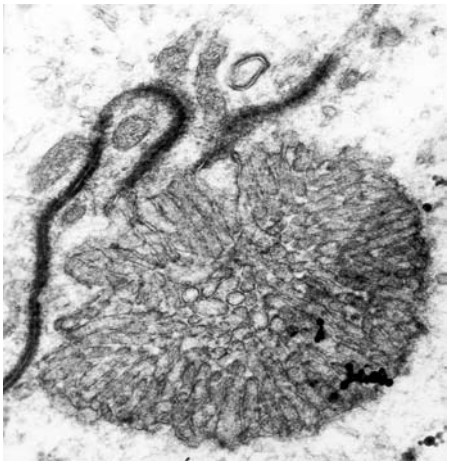

Figure 3.

DIAGNOSIS

Giant cell ependymoma.

DISCUSSION

The World Health Organization classification of CNS tumors recognizes a range of ependymal neoplasms, including clear cell, papillary, tanycytic, and perispinal myxopapillary variants (4, 5, 7, 9). Typically, giant cells characterize none of these; most examples of ependymoma contain isomorphic cells, and even moderate cytologic pleomorphism tends only to be found in anaplastic tumors. However, several features of this tumor would place it among the ependymomas; it is well demarcated, contains foci of uniform cells that are associated with rosettes and canals, and electron microscopy has demonstrated cilia, microvilli and complex intercellular junctions, all of which are typical of this group of tumors (4). Some uncommon gliomas, such as the pleomorphic xanthoastrocytoma (PXA) and giant cell glioblastoma, contain gigantic cells among a strikingly pleomorphic tumor cell population (3, 6), but the characteristics of the present tumor do not fit with these diagnoses, or the histopathologic features of a subependymal giant cell astrocytoma (SEGA). The SEGA, PXA and giant cell glioblastoma do not contain rosettes and canals. Other characteristics of the PXA, such as lipidized giant cells and a reticulin-rich architecture, were absent from this tumor. Also absent were the anaplastic features that would be expected in a giant cell glioblastoma.

Giant cell ependymomas have been reported, but are very rare (2, 8, 10). Two have occurred in the filum terminale, one with features of a myxopapillary ependymoma and foci of giant cell formation, and one that was composed entirely of giant cells (10). Two supratentorial examples have also been described, one intraventricular with features of anaplasia (2), and one extraventricular intraparenchymal example (8). Both of these tumors contained pseudorosettes and cytological features that clearly identified them as ependymomas. We are unaware of any report of a giant cell ependymoma in the posterior fossa.

The biologic behavior of such a rare variant of ependymoma is difficult to predict. The relatively good outcome associated with the PXA suggests that the presence of grossly atypical cells in gliomas is not necessarily associated with a poor prognosis (6). Ki-67 labeling index appears to be a prognostic indicator for ependymomas (1), and the relatively low Ki-67 LI in this case has so far (12 months) been associated with no disease progression.

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