

## Case Report

# Subependymoma with extensive microcystic transformation: a case report

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**Abstract:** Subependymomas are benign tumors that occur predominantly in the ventricular system. We described a case of a 44-year-old female with a large subtentorial mass which compressed the brain stem and fourth ventricle. Complete surgical excision of the tumor was achieved. The tumor composed of collections of cytologically bland cells with round to oval nuclei set within wide expanses of a delicate fibrillar matrix, and extensive microcystic changes were found. The tumor demonstrated positive staining with GFAP and S-100 protein, and did not stain with antibodies to Neu-N or Progesterone receptor, patchy expression of Epithelial Membrane Antigen. An MIB-1 labeling was lower than 1%. The tumor was totally resected and didn't recur after the initial surgery.

**Keywords:** Subependymoma, microcystic transformation

### Introduction

Subependymomas are small, discrete tumors of adults lying most often at the foramen of Monro or the fourth ventricle. It is composed of clusters of ependymal and astrocyte-like cells in a dense fibrillary stroma. It is typically attached to a ventricular wall and the most common site is the fourth ventricle. Most of the intraventricular subependymomas are subclinical. Histologically, the tumor may be either compact or microcystic, but extensive microcystic change is rarely reported. We describe histopathologic features of a subependymoma with extensive microcystic transformation and review the literatures.

### Case report

A 44-year-old female was being evaluated because of occasional headache and instability for approximate 4 years. Magnetic resonance imaging (MRI) study revealed a well-demarcated nonenhancing subtentorial mass. The mass was oval solid, which showed hypodense compared to gray matter on non-enhanced CT. Magnetic resonance imaging revealed the mass was smooth margins and was well demarcated from surrounding tissue. The signal of the mass was hyperintense on T1-weighted

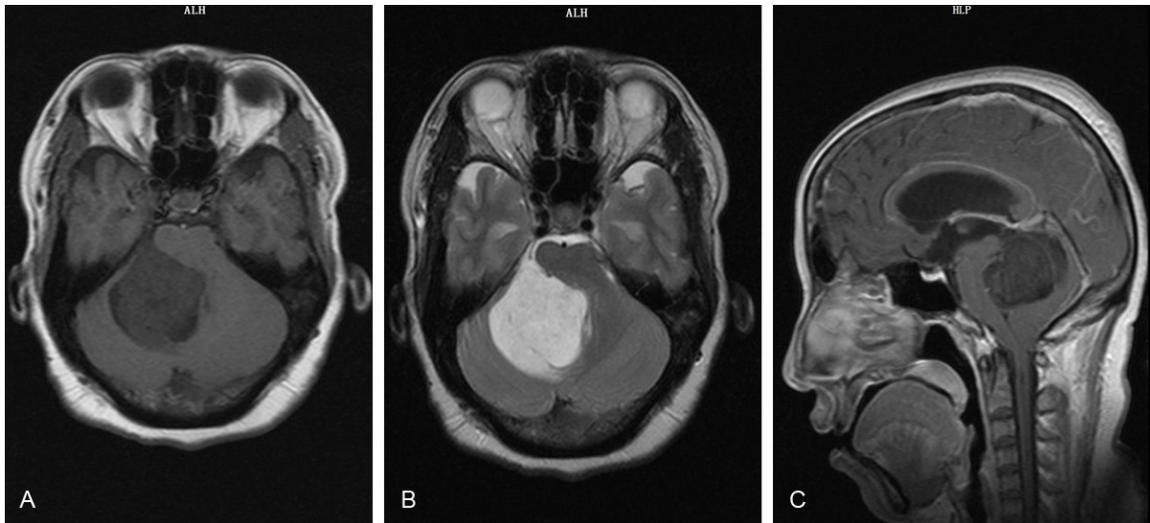
image (**Figure 1A, 1C**), and hyperintense on T2-weighted image (**Figure 1B**). No contrast enhancement was found in the mass. The mass was measuring 4.7 cm×5.8 cm. The mass compressed the brain stem and fourth ventricle, but there was only mild ventricular enlargement. The fourth ventricle was compressed to move towards left side by the mass. Supratentorial hydrocephalus was also found. On operation view, the tumors were solid mass with clear borders. There were cystic changes in the tumor. The tumor appeared soft and did not adhere to tentorium of cerebellum, and located in deep cerebellum, about 0.5 cm distance to the surface of cerebellum.

During surgery, a gray to white and soft, well-defined mass beneath cerebellum was submitted for intraoperative pathologic consultation. After operation, the patient recovered fully with no recurrence of his previous symptoms. The postoperative MRI revealed total removal of the mass.

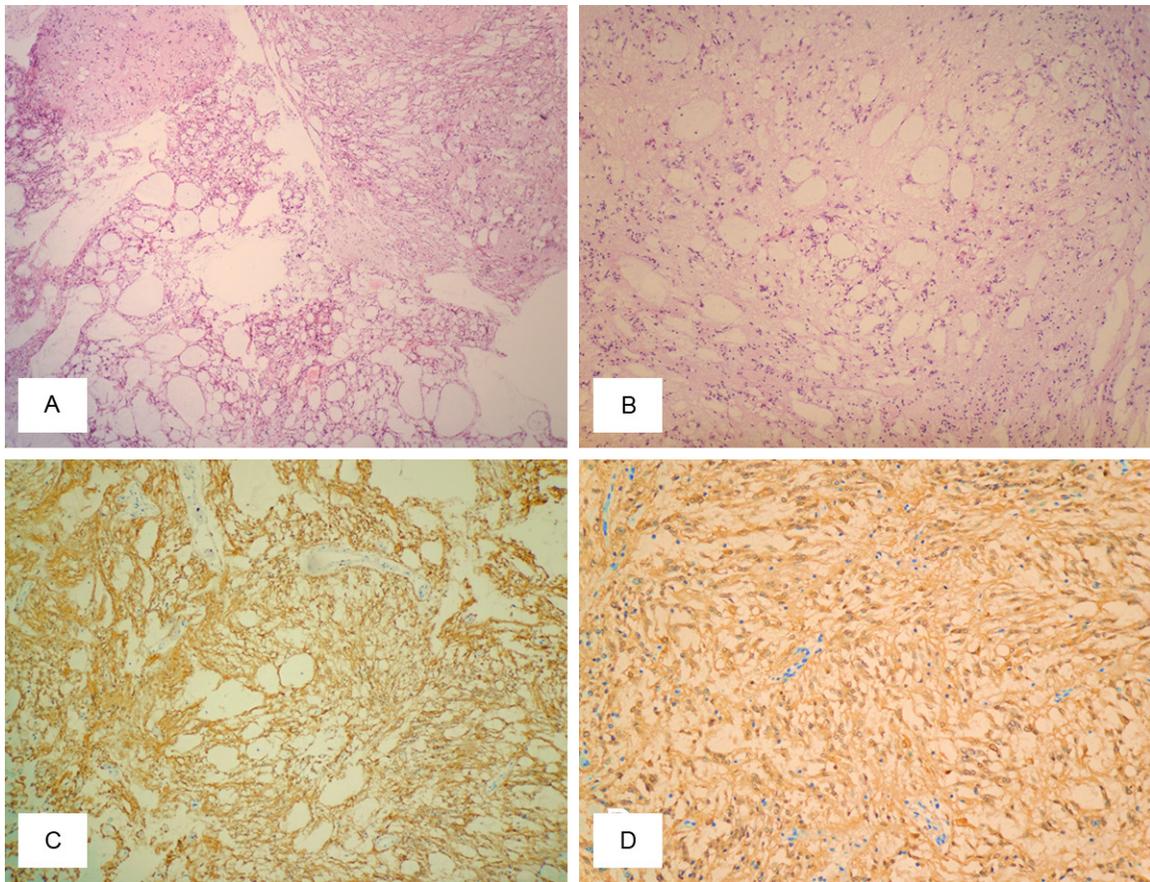
### Pathological findings

Microscopically, the mass showed delicately fibrillar stroma with prominent microcystic changes, abundant and extensive cystic changes were also found (**Figures 2A, 2B**). Nuclei

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**Figure 1.** A: Axial T1-weighted image demonstrates a hypointense mass. B: The mass was hyperintense on T2-weighted images. C: The mass shows long T1 signals on sagittal T1-weight image, which was well demarcated from surrounding tissue.



**Figure 2.** A, B: The mass showed delicately fibrillar stroma with prominent microcystic changes, abundant and extensive cystic changes were also found. Cellular density was low and occasional nuclear clusters in a dense fibrillar matrix were seen. There was no evidence of atypia, vascular endothelial proliferation or necrosis. C: Immunohistochemistry revealed intense expression of glial fibrillary acidic protein. D: S100 protein was positive.

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were round to oval. Mitosis was not observed. Cellular density was low and occasional nuclear clusters in a dense fibrillar matrix were seen. There was no evidence of atypia, vascular endothelial proliferation or necrosis. Immunohistochemistry revealed intense expression of glial fibrillary acidic protein (**Figure 2C**) and S100 protein (**Figure 2D**), weakly positive expression of oligo-2, and patchy expression of epithelial membrane antigen. No expression of Neu-N or Progesterone receptor was present. MIB-1 proliferation index was very low.

### Discussion

Subependymomas rarely encountered in the central nervous system [1], which are likely to remain asymptomatic throughout life and some were found by autopsy. If symptomatic, tumor location and size are critical factors for presentation. Subependymomas are indolent, benign gliomas, which occur in middle to late adulthood. They typically appear as a pedunculated intraventricular mass most commonly in the fourth ventricle, followed by the lateral ventricles, less commonly in spinal cord [2-4]. Subsequent reports have been in isolation or as small series, except for a few larger case series [5]. Many supratentorial subependymomas appear to be centered in the cortex or subcortical white matter. Therefore, a lack of ventricular involvement does not exclude subependymoma from the differential diagnosis. The histogenesis of subependymomas is still a matter of debate, with candidates including subependymal glia, astrocytes, ependymal cells, or some mixture of these cells. A recent theory hypothesizes that they originate from tanycytes, which are cells normally located in the subependymal zone [6]. The histological features of subependymomas are characteristic by loose clusters of bland, rather monomorphic cells with nuclear features of ependymal differentiation in a rich fibrillary background. Microcystic transformation is common. True ependymal rosettes, ependymal canals, and perivascular pseudorosettes are lacking. Immunohistochemically, subependymomas usually show strong immunopositivity for GFAP and S-100 antigens [7]. Subependymomas have the lowest rate of MIB-1 proliferation index [8]. The main problem encountered in histological diagnosis is represented by the possible coexistence of an additional ependymoma

which gives the tumor the dignity of grade II [7]. Subependymoma with extensive microcystic changes should differentiate with other tumors with microcystic transformation, such as pilocytic astrocytomas, oligodendrogliomas, well-differentiated astrocytomas, chordoid gliomas, myxoid meningiomas, and myxopapillary ependymomas. Ependymomas have complex cytogenetic and molecular genetic alterations that are not observed in astrocytomas [9]. The surgical aims are the maximal safe tumoral resection, the decompression of neural elements, and establishment of a pathological diagnosis and the restoration of normal CSF pathways. As subependymomas are low-grade lesions with low rates of cell proliferation and a benign clinical course, complete surgical removal is usually curative.

### Disclosure of conflict of interest

None.

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