Rapid expansion of a previously asymptomatic subependymoma

Case report

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This 39-year-old man presented with a 6-month history of occipital headaches. Magnetic resonance imaging revealed an irregularly shaped fourth ventricle mass. One month after his initial presentation, he was admitted to the hospital with significant tumor expansion and clinical deterioration. A posterior fossa craniectomy was performed and the mass was resected. Histopathological analysis of this tumor showed central necrosis with associated edema in an otherwise typical and benign-appearing subependymoma. To the authors’ knowledge, this is the first reported case of rapid, nonhemorrhagic expansion associated with necrosis in a previously asymptomatic subependymoma.

Key Words • subependymoma • tumor expansion • magnetic resonance imaging

F irst described in 1945 by Scheinker, subependymomas are rare, benign, slow-growing tumors typically located in the fourth ventricle, the lateral ventricles, or, occasionally, the cervicothoracic spine. These tumors are well-circumscribed noninvasive masses composed of hypocellular clusters of small uniform nuclei within a dense fibrillary matrix. Often only identified at autopsy examination, subependymomas have been estimated to comprise fewer than 10% of ependymal tumors. Because of their slow growth, fewer than 50% of subependymomas ever become symptomatic. In this report, we present a case of rapid, nonhemorrhagic expansion associated with necrosis in a previously asymptomatic subependymoma.

Case Report

Presentation and Examination. This 39-year-old man presented with a 6-month history of occipital headaches, hypertension, and fatigue, as well as occasional blurry peripheral vision and lower-extremity paresthesias. His neurological status was normal. Magnetic resonance imaging revealed a 3.5 × 3 × 2.7–cm midline fourth ventricle irregularly shaped, variably enhancing mass extending through the foramen of Magendie and displacing the medulla and cerebellum. There was no associated hydrocephalus. The neuroimaging characteristics of the mass were consistent with a subependymoma, ependymoma, or choroid plexus tumor. Because the patient was tolerating his relatively mild symptoms well, the decision was made to undertake serial neuroimaging to observe the mass.

One month later, the patient presented to the emergency department with severe headache and nausea. His neurological status was otherwise normal. Computerized tomography scanning revealed ventriculomegaly and enlargement of the fourth ventricle mass, raising concerns that the tumor was malignant. Given the lesion’s expansion and the patient’s clinical deterioration, we chose to excise the tumor.

Operation. A suboccipital craniectomy and C-1 laminectomy were completed. Once the dural edges were retracted, the tumor could be seen protruding between the cerebellar tonsils. The tumor had a heterogeneous appearance. Its inferior portion was yellow-gray, firm, and lobulated whereas its superior aspect was yellow-brown, soft, and possessed elements that appeared necrotic. No hemorrhage was seen. The central core of the tumor was debulked using an ultra-
Sonic aspirator. The tumor walls were then dissected away from the surrounding brain and most of the mass was removed except for a thin layer of cells on the floor of the fourth ventricle. Complete hemostasis was ensured and the surgical site was closed in the usual fashion. The patient tolerated the procedure well and was taken to our step-down unit for observation overnight.

**Histopathological Evaluation.** Grossly, the tissue had a pink-tan appearance. On microscopic examination, the tissue was clearly a tumor with low cellularity (Fig. 2). The tumor cells had small uniform nuclei. They were arranged in clusters separated by fibrillary cellular processes. These microscopic characteristics clearly identified the tumor as a subependymoma. Within the tumor mass, a large central necrotic cavity was present. Small foci of hemosiderin-laden macrophages suggestive of an old hemorrhage were also seen. It is important to note, however, that there was no large acute hemorrhage to account for the rapid expansion of the tumor. Around the periphery of the necrotic core, microvascular thrombosis and edematous tumor tissue were present. Surrounding this was typical subependymoma tissue and no microvascular thrombosis or edema. It is likely that the microvascular thrombosis led to vascular congestion, central infarction, subsequent necrosis, and intratumoral edema. The rapid expansion of this subependymoma is, therefore, most likely attributable to intratumoral microvascular thrombosis leading to necrosis and edema. This association is well reported in relation to malignant brain tumors such as glioblastoma multiforme, but it has not been previously reported in subependymomas. The MIB-1 labeling index of this tumor was less than 1%, which is typical of subependymomas.

**Postoperative Course.** The patient was discharged from the hospital 3 days after the operation. He had no headache and was neurologically intact. He remains well 1 year postoperatively.

**Discussion**

Most subependymomas remain asymptomatic and are therefore generally associated with a benign clinical course. Nevertheless, the mortality rate in cases of symptomatic

![Fig. 1. A and B: Sagittal T₁-weighted Gd-enhanced MR images obtained at initial presentation (A) and 1 month later (B) at admission following tumor expansion. C and D: Coronal T₁-weighted Gd-enhanced MR images at initial presentation (C) and 1 month later (D) at admission following tumor expansion. Significant tumor expansion and changes in Gd enhancement are evident.](image-url)
subependymomas was reported to be as high as 51% before neurosurgical microscopes were used, and is now approximately 9% in the microneurosurgical era.\textsuperscript{7,9} The mortality and morbidity rates are higher in cases involving tumors in the floor and lateral recesses of the fourth ventricle.\textsuperscript{9} Because of the potential morbidity associated with subependymomas and their treatment, it is important to recognize factors that might be associated with a less benign clinical course. The two most commonly identified poor prognostic factors associated with subependymomas are tumor location and size.\textsuperscript{6,9}

The fourth ventricle is the most common location for subependymomas.\textsuperscript{9} Of the fourth ventricle subependymomas, those most likely to become symptomatic originate in the region of the inferior floor.\textsuperscript{9} In addition to location, tumor size also helps to predict whether a subependymoma will become symptomatic.\textsuperscript{5} In reviewing a large series of subependymomas, Scheithauer\textsuperscript{9} found that symptomatic fourth ventricle tumors averaged 4 cm in their largest dimension, whereas asymptomatic subependymomas averaged 0.8 cm. Furthermore, large subependymomas are more likely to include cystic elements, focal calcification, vessel hyalinization, hemorrhage, and necrosis. This type of degenerative change may be seen in as many as one third of subependymomas.\textsuperscript{6} Of these degenerative changes, necrosis is exceedingly uncommon.\textsuperscript{7,9} Mixed-type subependymomas with ependymomatous elements tend to have a worse prognosis. The MIB-1 index is generally low with subependymomas, and the rare cases of increased MIB-1 labeling have not been reliably associated with an increased aggressive clinical behavior.\textsuperscript{7} In our case, in addition to degenerative change and location in the inferior floor of the fourth ventricle, the most important negative prognostic factor was the microvascular thrombosis with necrosis and edema that led to rapid expansion.

On neuroimaging, subependymomas generally appear to be discrete, well-demarcated, noninvasive intraventricular masses in which little or no surrounding edema is present.\textsuperscript{2} Subependymomas are typically iso- or hypointense on T\textsubscript{1}-weighted MR images, exhibit mild to moderate hyperintensity on T\textsubscript{2}-weighted MR images, and demonstrate no or variably heterogeneous enhancement after Gd administration on T\textsubscript{1}-weighted MR images.\textsuperscript{2} The neuroimaging characteristics of subependymomas are suggestive but are not specific enough to allow for a definitive diagnosis. Neuroimaging also allows clinicians to monitor growth rate. The rapid nonhemorrhage-associated growth and the lesion’s peritumoral ring enhancement in our case is highly unusual for subependymomas. If rapid growth and clinical deterioration are identified, as in our case, prompt surgical intervention is recommended.

Resection of subependymomas is generally reserved for symptomatic tumors and is often curative.\textsuperscript{7} Even subtotal resections have been associated with good long-term outcomes.\textsuperscript{3} Postoperative radiotherapy is therefore usually not recommended for subependymomas.\textsuperscript{5} In the setting of the rarely reported recurrent subependymoma, some authors have recommended radiation therapy, but this is controversial.\textsuperscript{1,3,5,11}

**Conclusions**

To our knowledge, this is the first reported case of a subependymoma featuring a large area of central necrosis associated with edema, rapid tumor expansion, and no hemorrhage. Aside from the presence of necrosis, the tumor displayed the classic histopathological characteristics of subependymoma, including hypocellular clusters of isomorphic nuclei separated by dense bundles of glial processes. The tumor’s microvascular thrombosis causing vascular congestion and central infarction, which led to necrosis and edema, likely accounted for the rapid and significant tumor expansion in our case. The tumor’s dramatic growth during a 1-month period was associated with a marked deterioration in the patient’s clinical condition. Resection of the tumor relieved the patient’s symptoms and he remains well 1 year postoperatively.

**References**

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