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Infratentorial and Intraparenchymal Subependymoma in the Cerebellum: Case Report

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Subependymomas are rare benign tumors located in the ventricular system. Intraparenchymal subependymoma is extremely rare; only 6 cases have been reported, and all were located in the supratentorial region. We describe a case of infratentorial, intraparenchymal subependymoma in a 28-year-old man with intermittent headache. Imaging revealed a well-demarcated cystic and solid cerebellar mass near the fourth ventricle. The mass had a microcystic component and calcification without contrast enhancement. Complete surgical excision was performed, and histopathology confirmed a subependymoma.

Index terms: *Subependymoma; Intraparenchymal; Cerebellum*

INTRODUCTION

Subependymoma is a rare, benign and slow-growing glial tumor that accounts for just 0.2-0.7% of all intracranial tumors and approximately 8% of ependymal neoplasms (1-3). Matsumura et al. (4) reported that the frequency of intracerebral subependymomas was 0.4% in 1000 serial routine autopsies. Although these tumors can occur in all age groups and both sexes, they tend to be more common in middle-aged to older individuals with a slight male predilection. Most are clinically silent, and the symptoms depend on tumor location and size. In some cases, especially when the tumors are larger, the symptoms are the result of increased intracranial pressure due to obstructive hydrocephalus (1, 5). Subependymomas most frequently arise in the fourth ventricle (50-60%), followed by the lateral ventricle (30-40%), and less frequently in the septum pellucidum and spinal cord (3, 6). Intraparenchymal subependymomas are extremely rare; only 6 cases have been reported in English literature. All of them were located in the supratentorial region (3, 7-9), and there has been no report of infratentorial subependymoma. Here, we describe the clinical and radiological feature of the first known case of infratentorial and intraparenchymal subependymoma arising in the cerebellum.

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CASE REPORT

A 28-year-old man was referred to our institution for a brain tumor that was detected on brain computed tomography (CT) performed at an outside hospital for intermittent headache. He had other non-neurologic symptoms, other than intermittent headache, for the past 1 year. The patient was previously healthy with unremarkable past medical history. A neurological examination revealed no

abnormal features, and initial lab findings were normal. Pre-contrast CT revealed a well-defined cystic and solid intra-axial mass with internal calcifications in the cerebellum, which measured 4.3 x 3.5 cm (Fig. 1A). Magnetic resonance image (MRI) revealed a mural nodule within the cystic mass. The mural nodule showed iso-to-hypointensity (relative to normal white matter) with multiple nodular high-signal intensities, suggesting calcifications on the T1-weighted image and hyper intensity on the T2-weighted image (Fig. 1B, C). There was no evidence of peritumoral edema around the mass and no enhancement of the mural nodule or cyst wall on the gadolinium-enhanced T1-weighted image. Diffusion-weighted image showed iso- or low-signal intensity without diffusion restriction while the same lesion was increased ADC relative to brain parenchyma (Fig. 1D, E). The mass was located near and displaced the fourth ventricle; however, it did not communicate with the ventricle, and normal parenchyma was clearly identified between the mass and the fourth ventricle on the thin-section three-dimensional T1-weighted images (Fig. 1F, G). Cerebral angiograms demonstrated a rather hypovascular mass; neither feeding vessels nor tumor staining was visualized (Fig. 1H). The patient underwent suboccipital craniotomy. The cerebellum showed mild bulging, and a yellowish-colored mass was identified via a supracerebellar approach. During the dissection of the mass from the cerebellum, fluid gushed out from patchy gray mass, which was composed of a gelatinous material. Within the surgical field, there was a sharp demarcation between the mass and the surrounding normal tissue, and there was no evidence that the mass was connected with the fourth ventricle.

Histopathological analysis revealed a clustered cellular neoplastic proliferation with islands of high nuclear density and dense and abundant fibrillary matrices. The tumor cell nuclei were round and isomorphic, but active mitosis was not observed (Fig. 1I). Occasional vascular pseudorosettes and calcifications were also seen. There was no evidence of atypia, vascular endothelial proliferation or necrosis. Immunohistochemical analyses were positive for glial fibrillary acidic protein and epithelial membrane antigen and negative for Ki-67, which supported the diagnosis of subependymoma.

The postoperative follow-up MRI revealed a total removal of the tumor resection; the normal cerebellar tissue remained well between the fourth ventricle and the post-operative parenchymal defect. This observation supports our hypothesis that the tumor did not contact the fourth ventricle. The patient was discharged without neurologic sequelae 10 days after surgery.

DISCUSSION

Subependymomas are at the benign end of the tumor spectrum and account for just 0.2-0.7% of all intracranial tumors (1). The World Health Organization classification system recognizes 4 types of ependymal neoplasms, including subependymoma and myxopapillary ependymoma (Grade 1), ependymoma and its variants (cellular, papillary, clear cell and tanycytic subtypes) (Grade 2) and anaplastic ependymoma (Grade 3) (9).

Subependymomas are usually located within or adjacent to ependymal-lined spaces. These tumors arise most frequently in the fourth ventricle (50-60%), followed by the lateral ventricle (30-40%); less frequent sites of origin are in the third ventricle, septum pellucidum and spinal cord, which is understandable given their proposed cellular origins (3). Numerous cell types have been proposed to be the cell of origin for subependymomas, including ependymal-glial precursor cells or bipotential subependymal cells, with the ability to differentiate into either ependymal cells or astrocytes, astrocytes of the subependymal plate or a mixture of astrocytes and ependymal cells. Other potential hypotheses propose that subependymomas arise from hamartomatous lesions or as a reaction to chronic ependymitis (3, 4).

A review of the English literature identified only 6 case reports of subependymomas, all of which were in intraparenchymal locations (Table 1). Specifically, all 6 were supratentorial subependymomas: 2 parietal (8, 9), 1 parietooccipital (3), 1 temporal (3) and 1 frontal (7). Location information was not available for 1 case (9). An infratentorial, extraventricular and intraparenchymal subependymoma has not been previously reported.

The pathogenesis of extraventricular ependymal tumors remains unclear. Ependymal cell rests, which are frequently found at the angle of the ventricles where ependymal cells extend deep into the adjacent white matter, have been hypothesized as the origin of these neoplasms (9, 10). However, even if the fourth ventricle is the most common location of subependymomas, the fact that there is no cerebellar subependymoma adjacent to the fourth ventricle suggests other causes. We speculate that the tumor may have arisen from ependymal-glial precursor or bipotential subependymal cells around the fourth ventricle, and that

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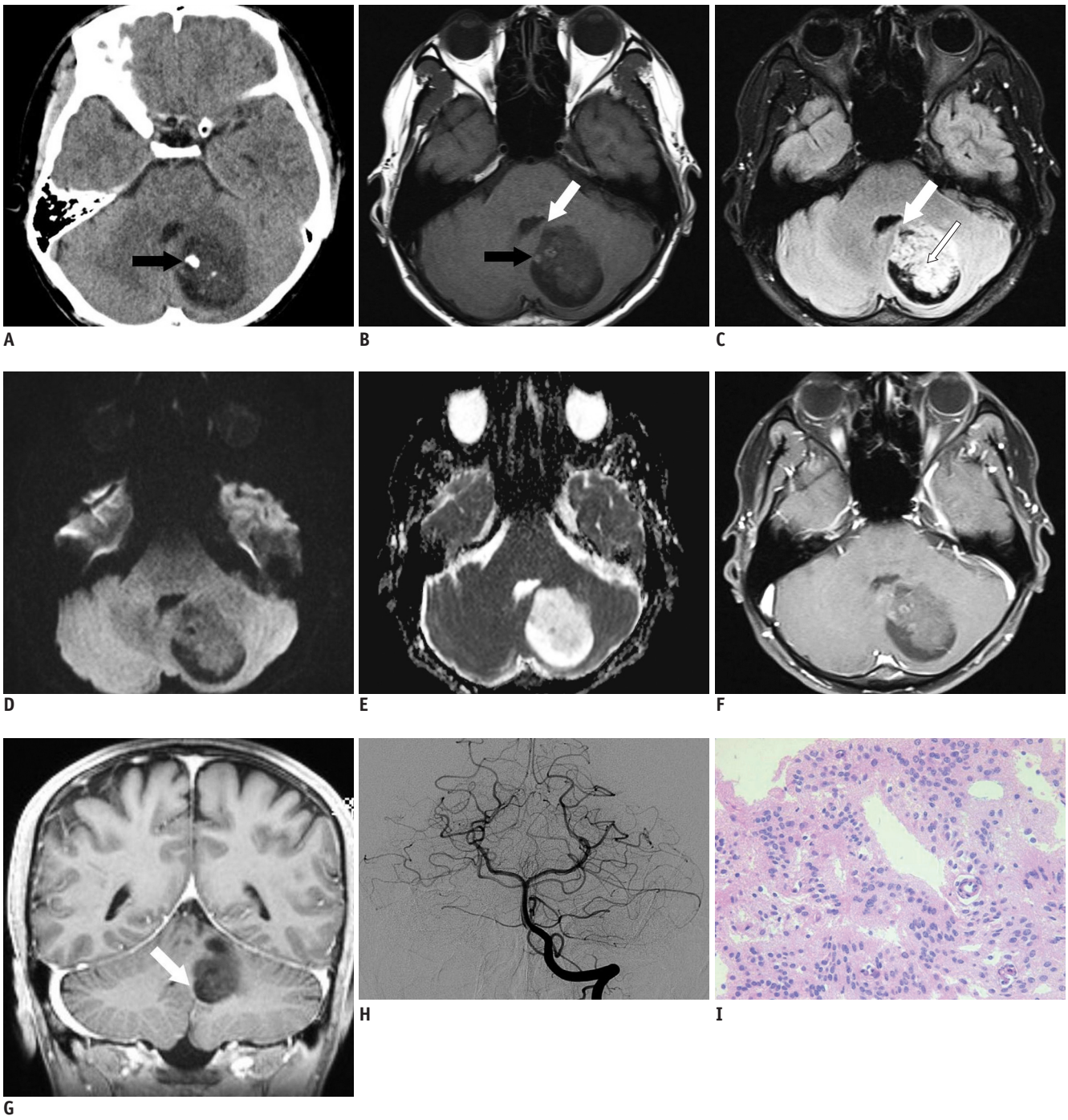

Fig. 1. Imaging features of cerebellar subependymoma in 28-year-old man.
A. Unenhanced axial CT scan demonstrates well-defined cystic and solid mass with multiple calcifications (black arrow) in left cerebellum. **B-E.** Mass exhibits iso- to hypointensity (relative to normal gray matter) on axial T1-weighted MRI and hyperintensity on axial T2-FLAIR. There were multiple, small, high-signal intensity lesions (black arrow) suggesting calcifications on T1-weighted imaging, and numerous dot-like low-signal intensities suggesting microcysts (thin white arrow) on T2-FLAIR. On DWI, mass exhibited hypointense signal (relative to gray matter) on DWI without diffusion restriction and increased ADC relative to brain parenchyma. There is no evidence of peritumoral edema, mass effect or intratumoral hemorrhage. Mass was near fourth ventricle, but clearly separated from it. Isointense area (thick white arrows) suggesting normal cerebellar white matter was observed between mass and fourth ventricle. **F, G.** On axial and coronal contrast-enhanced MR images, there was no enhancement in solid portion or peripheral wall, and normal parenchyma was clearly identified between mass and fourth ventricle (white arrow). **H.** Cerebral angiograms did not reveal feeding vessel or tumor staining. **I.** Microscopic images showing clustered cellular neoplastic proliferation with islands of high nuclear density with dense, abundant fibrillary matrices. Round and isomorphic nuclei were observed, but mitosis was not seen (HE; original magnification, x 40). FLAIR = fluid attenuated inversion recovery, DWI = diffusion-weighted image, ADC = apparent diffusion coefficient, HE = hematoxylin and eosin

Table 1. Literature Reviews of Intraparenchymal Subependymoma

References	Age/Sex	Location	Relationship with Ventricle	MRI	CT
Ragel et al. (3)	34/M	Rt. temporal	Adjacent to temporal horn	3.0 cm, T1 mixed, T2 hyper, moderate enhancement	N/A
Ragel et al. (3)	57/M	Rt. parietooccipital	Postero-lateral to Rt. lateral ventricle	2.5 cm, T1 hypo, T2 hyper, no enhancement	N/A
Natrella et al. (7)	45/M	Lt. frontal lobe	Far off the ventricle	Cystic extraaxial mass with mural nodule, minimal enhancement	N/A
Hankey et al. (8)	21/F	Rt. parietal lobe	N/A	N/A	Hemorrhagic cyst
Shuangshoti et al. (9)	N/A	Parietal lobe	N/A	N/A	N/A
Shuangshoti et al. (9)	N/A (follow up loss)				
Current case	28/M	Cerebellum	Adjacent to fourth ventricle	4.3 cm, T1 iso, T2 hyper, calcification, no enhancement	Well defined cystic and solid mass with internal cacifications

Note.— N/A = not available

the relatively scanty white matter supported exophytic growth in the cerebellum.

Common imaging findings of subependymomas include observations of a well-demarcated, lobulated mass, which are usually composed of solid or occasionally mixed components, including cystic or microcystic degeneration. Most subependymomas appear as hypo- or isodense masses on unenhanced CT, hypo- to isointense on T1-weighted MRI and hyperintense on T2-weighted MRI (3). Calcification is noted in 32-50% of cases, and can be a differentiating feature between the lateral and fourth ventricular subependymomas. In many reports, intratumoral calcifications have been reported to be more a frequent finding in infratentorial subependymomas, whereas it is an unusual finding in the lateral ventricle subependymomas (11). Contrast-enhanced CT or MRI usually show minimal or no enhancement and infrequently reveal a heterogeneous enhancement. Angiography demonstrates an avascular or hypovascular mass that may cause a mass effect on the surrounding cerebrovasculature. Therefore, peritumoral edema, mass effect, dense enhancement, high vascularity and intratumoral hemorrhage are rare findings in subependymomas (3, 11). The involvement of the cerebellar hemisphere presenting radiologically as a solid and cystic mass, pilocytic astrocytoma, hemagioma, hemangioblastoma, ganglioglioma, ependymoma and subependymoma should be included in differential diagnosis. Slight or lack of enhancement on Ct or MRI should arouse the suspicion of subependymoma. In our case, MRI showed a cystic and solid mass that included multiple dot-like low-signal intensities on T1-weighted and FLAIR images, which was suggestive of a microcystic portion within the solid component. Some punctate calcifications and no enhancement were also observed on CT and MRI. There was no evidence of mass effect, obstructive hydrocephalus, peritumoral edema or intratumoral hemorrhage. Thus, with the exception of the intraparenchymal location, all imaging findings corresponded with the diagnosis of subependymoma.

The treatment of choice for symptomatic subependymoma is a complete surgical excision. The purposes of the surgery are tumor resection, decompression of neural elements, establishment of a pathological diagnosis and restoration of normal cerebrospinal fluid pathways; however, the extent of the resection does not appear to influence the survival rates. Although postoperative morbidity is rare, reported complications include hydrocephalus, meningitis and sepsis. Routine postoperative irradiation is not recommended (1, 3). Patient age is the only variable associated with survival; those older than 50 years have a worse prognosis (1).

Conclusion

Intraparenchymal subependymoma is extremely rare; yet, if imaging features are suggestive of subependymoma in a cerebellar tumor, we should not exclude the possibility of subependymoma. It has distinguishing clinical and radiological features, but they are not pathognomic. Although making a preoperative diagnosis was difficult, preoperative suspicion of subependymoma is very important for surgical planning, given its benign nature and good prognosis.

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