Fourth ventricular subependymoma presenting as worsening headache

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Subependymomas are rare, slow-growing benign neoplasms. Although most are asymptomatic, they can present with symptoms related to increased intracranial pressure and hydrocephalus. We describe a 47-year-old man with worsening headaches who was found to have a subependymoma, with a focus on the imaging findings, differential diagnoses, pathology, and treatment.

CASE REPORT

A 47-year-old man, who previously had undergone gastric bypass for morbid obesity, presented to his neurologist complaining of headaches for 3 years, with major worsening of their frequency and severity for 2 weeks. The headaches now involved his entire head and were only transiently relieved with over-the-counter analgesics.

Magnetic resonance imaging (MRI) of his brain was obtained and revealed an extraaxial mass in the inferior fourth ventricle extending through the foramen magnum and impressing upon the pontomedullary junction and medulla with posterior compression of the cerebellar tonsils. The mass had similar signal intensity to brain parenchyma on T1-weighted images and showed scattered heterogeneous enhancement (*Figure 1*). The T2-weighted images revealed increased signal intensity relative to brain parenchyma and lack of adjacent edema (*Figure 2*). A gradient echo sequence showed scattered foci of hypointensity indicating T_2^* susceptibility favoring the presence of calcific deposits and/or blood (*Figure 3*).

The patient underwent a craniotomy with complete resection of the fourth ventricular mass (*Figure 4*). Histologic evaluation of the lesion confirmed the diagnosis of subependymoma (*Figure 5*). The patient was subsequently discharged from the hospital after an uneventful postoperative course.

DISCUSSION

Subependymomas account for about 1% of all intracranial neoplasms (1). They are slow growing and well circumscribed. While they usually arise in the fourth ventricle in adults (60%),

Figure 1. (a) Sagittal and (b) coronal T1-weighted postcontrast MRI reveals a heterogeneously enhancing fourth ventricular mass (arrows).

they have also been described in the lateral and third ventricles as well as in the spinal cord (2). Imaging characteristics commonly include a well-defined solid or less frequently mixed solid and cystic intraventricular mass. The solid component is isodense on computed tomography, isointense on T1-weighted images, and hyperintense on T2-weighted images. Peritumoral edema is usually not present. Lesions are typically 1 to 2 cm in diameter; however, tumors >5 cm have been described (2). Enhancement characteristics are variable: they are most often absent, but mild to moderate and even intense enhancement has been described. Calcific deposits can be seen with larger tumors and are frequently a constituent of fourth ventricular subependymomas. Hemorrhage is rare (2–4). The differential diagnosis of a subependymoma is an ependymoma, choroid plexus papilloma, central neurocytoma, subependymal giant-cell astrocytoma, hemangioblastoma, cavernous malformation, and metastasis. In an individual of this age, and in the location of this mass with the given MRI features outlined above, the

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Figure 2. Axial T2-weighted MRI. The mass (arrow) is seen insinuated between the posterior aspect of the medulla and the ventral aspect of the cerebellar tonsils without evidence of vasogenic edema.

Figure 3. Axial multiple-echo recalled gradient echo (MERGE) MRI. The mass (arrow) has scattered foci of hypointensity representing areas of calcification and/or hemorrhage, although calcification would be favored in a subependymoma.

diagnosis of exclusion was a subependymoma with a less likely consideration being an ependymoma.

On gross examination, subependymomas have a white to gray color, are well circumscribed with a firm texture, and are

Figure 4. Intraoperative image demonstrates a subependymoma (arrow) inferior to the cerebellar tonsils prior to removal (asterisks: cerebellar hemispheres).

Figure 5. Hematoxylin and eosin–stained section of the fourth ventricular subependymoma showing characteristic clustering of nuclei in a well-differentiated fibrillary background. Original magnification $\times 100$.

usually avascular with an attachment to the ventricular wall by a narrow pedicle. These lesions are thought to arise from subependymal glial cells, although other considerations include astrocytes from the subependymal plate, ependymal cells, and mixed ependymal and astrocytic cells (2, 5). On histological examination, clusters of nuclei on a fibrillary background are seen. Immunohistochemical staining for glial fibrillary acidic protein is highly positive. Mitotic activity is typically minimal or absent, corresponding to a World Health Organization grade 1 lesion (2, 5).

The treatment of choice for subependymomas is complete surgical resection, which yields an excellent prognosis with rare recurrences (2, 6). A complete surgical resection is more commonly possible with lesions located in the lateral ventricles,

with fourth ventricular locations often treated with subtotal resections. There is little documented additional benefit from chemotherapy or radiotherapy (2, 6).

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Avocations

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