

Case Report

Central Neurocytoma

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Case

This is a previously healthy 20 year old female, with 3 months of worsening daily headaches. These headaches were diffuse, lasted for several hours, and mostly occurred in the morning. She was initially diagnosed and treated for migraines. In spite of being on migraine medications, however, her headaches worsened, and started to become associated with nausea, vomiting and blurry vision. She was brought to the emergency room, because she started to become progressively more lethargic over the course of several days.

On examination, in the emergency room, her vital signs were unremarkable, she was arousable to voice, but otherwise had no other focal neurological deficits.

Initial Neuroimaging Study

On axial FLAIR Magnetic Resonance Imaging (MRI) (Figure 1) there was a heterogeneously intense lesion that appeared to be centered, and arising from the septum pellucidum. There was also enlarged ventricular size with slight transpendymal edema (single black arrow). On T1 post-Contrast Coronal, (Figure 2) there was the partially enhancing lesion, once again centered on the septum pellucidum and obstructing both foramen of Monro. The Temporal horns of the lateral ventricles (white arrow) are enlarged while the 3d ventricle (black arrow) was normal in size signifying the obstructive nature to the hydrocephalus. Sagittal T1 without contrast (Figure 3) shows the heterogeneously enhancing, "soap-bubble" lesion obstructing the foramen of Monro. The hyperintense foci in the lesion either represent calcification or hemorrhage.

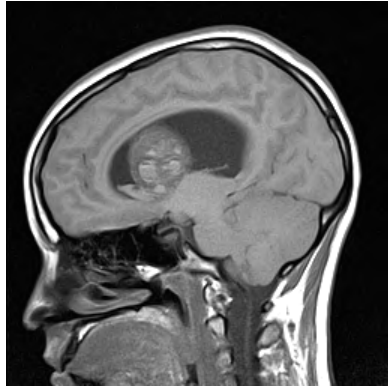


Figure 1: Sagittal T1 without contrast, showing the heterogeneously enhancing lesion obstructing the foramen of Monro. The hyperintense foci either represent calcification or hemorrhage.

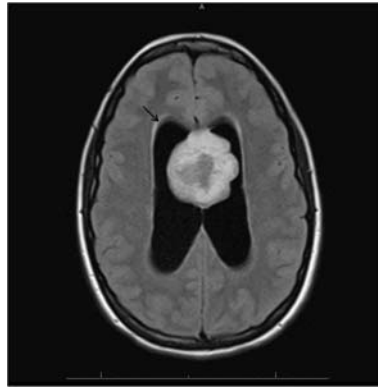


Figure 2: Axial FLAIR, showing enlarged ventricular size with slight transpendymal edema (single black arrow). The heterogeneously intense lesion appears to be centered, and arising from the septum pellucidum of the lateral ventricles.

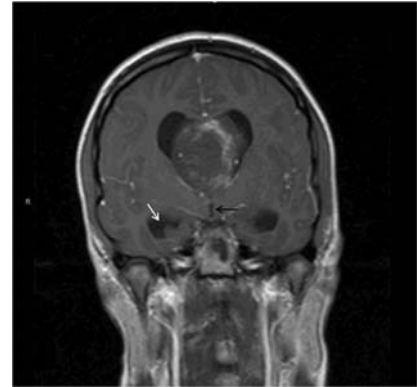


Figure 3: Coronal T1 post-contrast, showing the partially enhancing lesion once again centered on the septum pellucidum and obstructing both foramen of Monro. The Temporal horns of the lateral ventricles (white arrow) are enlarged while the 3d ventricle (black arrow) is normal in size signifying the obstructive nature to the hydrocephalus.

Surgery & Neuropathology

Because of the acuteness of the patient's course, she was brought emergently to the operating room where the lesion was resected via a bifrontal craniotomy, and interhemispheric, transcallosal approach. During the resection, the lesion was felt to be adherent to the septum pellucidum.

Histopathological examination showed a homogeneous collection of cells with clear cytoplasm, that looked very similar to an oligodendroglioma, but that stained positive for synaptophysin (Figure 4). This confirmed the diagnosis of central neurocytoma. The patient had no postoperative complications and was discharged several days later with resolution of her headaches and without any neurological deficits.

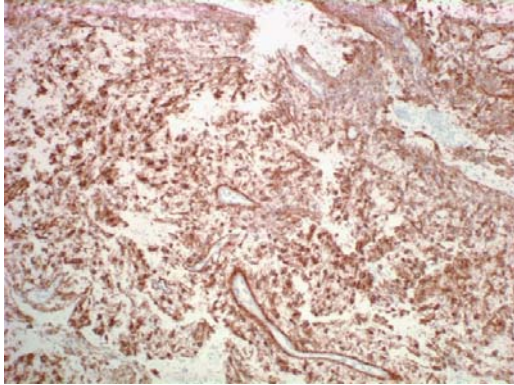


Figure 4: Synaptophysin immunostain showing strong positive staining, indicating the neuronal origin of the tumor

Discussion:

The differential diagnosis for lateral ventricular tumors depends on the specific part of the lateral ventricle that is involved and the age of the patient:¹

Table 1: Differential diagnosis according to age and location¹

Location	Adult	Pediatric
Foramen of Monro	<ul style="list-style-type: none"> • Central Neurocytoma • High Grade Astrocytoma • Subependymoma (Figure 5) • Colloid Cyst (Figure 7) 	<ul style="list-style-type: none"> • Oligodendroglioma • Subependymal giant cell astrocytoma (Figure 6) • Oligodendroglioma • Colloid Cyst
Body	<ul style="list-style-type: none"> • Oligodendroglioma • Central Neurocytoma • Astrocytoma • Subependymoma 	<ul style="list-style-type: none"> • Primitive Neuroectodermal Tumor • Astrocytoma
Trigone	<ul style="list-style-type: none"> • Meningioma • Metastases • Lymphoma 	<ul style="list-style-type: none"> • Ependymoma • Choroid Plexus Papilloma • Astrocytoma

In our 20-year old patient with a mass close to the foramen of Monro, central neurocytoma, high grade astrocytoma, oligodendroglioma, subependymoma and colloid cyst were the possibilities considered. MRI was helpful in refining the differential diagnosis.

- Central neurocytomas are usually T1 isointense (Figure 1), with variable intensity on T2 (Figure 2) and a heterogeneous enhancement pattern and globular calcifications imparting the classic “soap-bubble” appearing lateral ventricular lesion (Figure 3).² Another characteristic feature of central neurocytomas is that they usually appear to arise from either the septum pellucidum (our case) or foramen of Monro.³
- Oligodendrogliomas are T1 iso- or hypointense and T2 hyperintense tumors. Edema and gadolinium enhancement are minimal unless there are anaplastic features.
- Subependymomas (Figure 5) are non-enhancing masses that are T1 hypointense, T2 hyperintense and most commonly arise from the fourth rather than the lateral ventricle.²

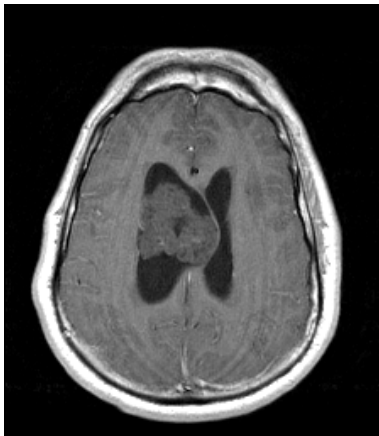


Figure 5: Axial T1 post-contrast, showing a subependymoma in the right lateral ventricle. These lesions are typically non-enhancing, and their most commonly found in the fourth ventricle

- Subependymal giant cell astrocytoma (Figure 6) in patients with tuberous sclerosis

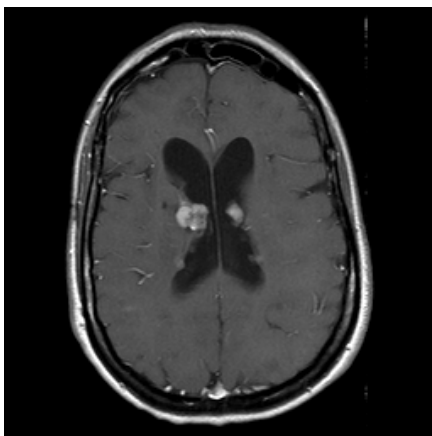


Figure 6: Axial T1 post-contrast depicting the contrast enhancing subependymal giant cell astrocytoma in a child with tuberous sclerosis. The most common location of subependymal giant cell astrocytomas is at the foramen of Monro

- Colloid cysts (Figure 7) arise from the roof of the 3rd ventricle obstructing the foramen of mono. On MRI, the signal intensity may vary from hypo- to hyperintense on T1 and T1 weighted images. The increase in signal intensity correlates with high cholesterol content. Frequently, colloid cysts are hyperintense on T1 and iso- or hypointense on T2. There may be a thin rim of gadolinium enhancement representing the cyst capsule.⁴



Figure 7: Axial non-contrast CT scan, depicts a classic colloid cyst as a well-circumscribed hyperdense lesion that arises from the roof of the 3rd ventricle

In our patient, the combination of soap bubble appearance, and relation to septum pellucidum made central neurocytoma the most likely diagnosis.

Central neurocytomas are slow-growing tumors that are of neuronal origin. Histologically they resemble oligodendrogliomas and were originally thought to be unusual intraventricular oligodendrogliomas until their neuronal character was established by electron microscopy. Currently, immunostaining for synaptophysin can confirm the neuronal nature (Figure 4).⁵ It is classified as WHO grade II.⁶ Treatment is with complete surgical resection, but radiation can be used for recurrence or incomplete resection.

Teaching Points

- 1) Central neurocytoma should be at the top of the differential in a young adult with a “soap-bubble” appearing intraventricular lesion that appears to arise from the septum pellucidum.
- 2) Complete surgical resection implies cure.

References

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