Imaging of Central Neurocytomas

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KEYWORDS

• Central neurocytomas • Neuroradiology • Computed tomography • Magnetic resonance imaging • Magnetic resonance spectroscopy • Brain tumor • Intraventricular tumor

KEY POINTS

- Central neurocytomas (CN) are classically found in the lateral or third ventricles, particularly in proximity to the septum pellucidum and the foramen of Monro; however, they can arise anywhere throughout the neuroaxis.
- On CT, CN appear as mixed density, partially calcified masses, and cysts and hemorrhage can occasionally be seen.
- On MRI, CN are isointense to brain on T1, have a "soap-bubble" multicystic appearance on T2, often exhibit fluid-attenuated inversion recovery hyperintensity, may have heterogeneous enhancement with gadolinium, and may have vascular flow voids; hemorrhage may be seen, but edema is usually minimal.
- A spectrum with an inverted alanine peak at 1.5 ppm, a notable glycine peak at 3.55 ppm, and the presence of N-acetyl aspartate (NAA) are most consistent with CN.
- Rarely, extraventricular or even spinal neurocytomas may be found. Similar characteristics of calcification, cystic components, associated hemorrhage, and vascular flow voids may be seen, although the characteristic cystic soap-bubble appearance is often absent.

INTRODUCTION: NATURE OF THE PROBLEM

Background/History

First documented in 1982 by Hassoun and colleagues,[1] central neurocytomas (CN) are formally classified as a benign World Health Organization (WHO) grade 2 neoplasms of neural origin,[2] arising from the glial cells lining the septum pellucidum and subependymal glial cells, particularly near the foramen of Monro. CN were initially considered to be an intraventricular variant of oligodendroglioma until pathologic analysis revealed cellular differences[1,3] and a population of immature neurons, usually without anaplasia and with a low MIB-1 labeling index.[2] Immunohistochemical analysis revealed that CN stained positive for synaptophysin, neuron specific enolase (NSE), and neuronal nuclear antigen,[4,5] unlike oligodendrogliomas, and later genetic analysis revealed genotypic differences (absence of 1p,19q codeletion,[6] and isocitrate dehydrogenase 1 [IDH1] mutations[7]). Further subtypes of neurocytomas have since been characterized, including extraventricular neurocytoma (EVN), which is officially recognized as distinct from CN by the WHO,[2] and reports of spinal neurocytoma.[8–10]

CN represent less than 0.5% of all cerebral neoplasms.[4,11] As of 2006, 438 cases had been reported,[5] and it is currently estimated that between 500 and 600 cases exist in the literature.[12] The peak prevalence of CN is in the third through fifth decades of life,[5,13] although cases in infants[14] and

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the elderly[5] have been described. Patients commonly present with symptoms of an intraventricular mass causing noncommunicating hydrocephalus, such as headaches (88%–90%) and visual disturbances (25%–38%).[6,13] Motor symptoms (19.8%) and altered mental status (10.9%) have also been commonly reported.[13] Rarely, seizures have been reported as the presenting symptom of EVN disease in the temporal lobe.[15,16] CN and EVN may present with either intratumoral,[17] intraparenchymal,[18] or intraventricular hemorrhage,[19–21] with all of the attendant sequelae of those processes. CN most commonly arise in the lateral and third ventricles, but reports of fourth ventricular,[17,22,23] pineal, and aqueductal[24] CN exist.

CN are a rare, but important consideration in the differential diagnosis of intraventricular lesions.[25] Because CN are benign lesions that often have an excellent prognosis with gross total surgical resection compared with difficult, infiltrative, or higher-grade lesions (and particularly oligodendrogliomas[11]), establishing the diagnosis early may guide surgical planning and patient management. CN may not require postoperative radiation therapy depending on the extent of resection, unlike most gliomas.[26] Fortunately, CN have several classical characteristics on CT, MRI, and magnetic resonance (MR) spectroscopy (MRS), which can assist clinicians in making the correct diagnosis.[25]

PREIMAGING PLANNING

Relevant Anatomy

CN typically arise in the lateral ventricles near the foramen of Monro (**Fig. 1**), but may arise from any cerebral spinal fluid (CSF) space including the third ventricle and the fourth ventricles. EVN may be found in any location within the parenchyma,[27] cranial nerves,[28] sella,[29] or even the skull base.[30,31] In several series, the median size of CN at diagnosis is reported as 4.6 to 5.2 cm.[25,32,33] CN range from subcentimeter asymptomatic lesions to up to 9 cm.[25,32,33] EVN have variable appearance on imaging but are largely similar to CN.[34] Although rare, spinal neurocytomas may be found with an associated syrinx or other sequelae of obstruction to CSF flow and may arise at any location within the spinal cord or nerve roots.[8] Accordingly, the treating physician should select imaging modalities that will permit visualization of the entire cranial vault (or entire neuroaxis if spinal disease is suspected).

Choice of Imaging Modality

Noncontrast computed tomography scan

Traditionally the initial imaging modality for the evaluation of any potential intracranial process,

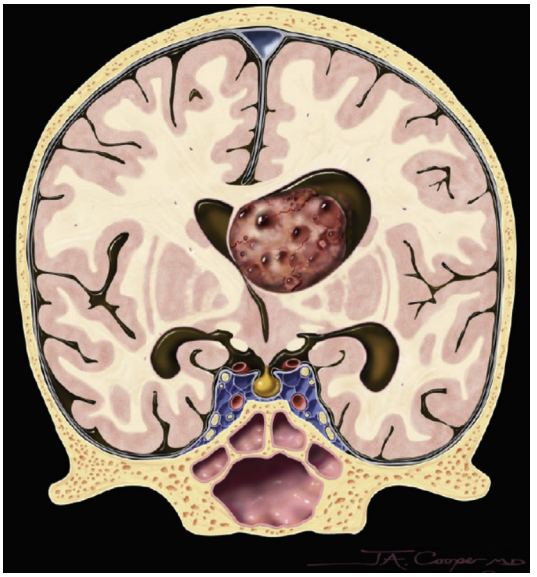

Fig. 1. An anatomic illustration in the coronal plane showing CN as a multicystic intraventricular lesion containing vessels and blood product. (*Courtesy of* Amirsys, Salt Lake City, UT; with permission.)

CT is often obtained during the initial assessment of altered mental status and is often ordered by a general practitioner or in the emergency department. CT is usually the most rapid and facile neuroimaging modality, at the cost of exposure to ionizing radiation. In the diagnosis of CN, CT is particularly valuable in rapidly evaluating the status of the ventricular system and in looking for calcification. CT can sometimes be useful in diagnosing the location of CN, although the solid component of CN is often isodense to brain and small CN can be missed on CT.[35]

Magnetic Resonance Imaging with and Without Gadolinium Contrast

MRI is typically the second step in the diagnostic algorithm for most intracranial lesions, unless a contraindication to CT scanning exists. Because the precise diagnosis will not be known at the time of ordering the MRI, it is wise to use a standard brain tumor protocol with specific sequences capable of evaluating midline and ventricular lesions. Most institutions have a standard brain tumor protocol that includes T1 precontrast and postcontrast, T2, and fluid-attenuated inversion recovery (FLAIR) sequences. Additional sequences such as gradient echo or proton density sequences can be helpful in diagnosing hemorrhage. Multiplanar imaging, particularly imaging in the sagittal plane, is necessary for midline