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Case Report

Intraventricular Soap Bubble Appearance: Central Neurocytomas from a Radiologist's Perspective

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Abstract

Central neurocytoma (CN) is a rare benign intraventricular neuroepithelial tumor, constituting only 0.25-0.50% of intracranial tumors. Typically affecting young adults, CN presents with increased intracranial pressure symptoms. We report a case of a 22-year-old woman with worsening headaches, vomiting, and blurry vision. Imaging revealed a soap-bubble-like mass in the left lateral ventricle, confirming CN after resection. CN is distinguished by its location, commonly in the lateral ventricle near the foramen of Monro, and its bubble soap appearance on imaging. Surgery remains the primary therapeutic option, providing the best long-term prognosis for these slow-growing tumors in young adults.

Keywords: *Intraventricular Neoplasm; Central Neurocytoma; MRI; Diagnostic Radiology*

Introduction

Central neurocytoma (CN) is a rare benign intraventricular neuroepithelial tumour, first described by Hassoun and colleagues in 1982 [1]. It accounts for only 0.25-0.50% of all intracranial tumours and affects young adults of both sexes at an equal rate [2]. Key to the diagnosis is its location, typically in the lateral ventricle near the foramen of Monro and with a distinctive attachment to the septum pellucidum [2]. In this report, we present a case of a 22-year-old female with a central neurocytoma and discuss the radiological aspect and differentials of this tumour.

Case Report

Miss G.T, 22 years old, was a previously healthy young women, who experienced diffuse headaches for several months, that recently started to become associated with vomiting and blurry vision which brought her to the emergency room.

Her vital signs were unremarkable on clinical examination, and she had no other focal neurological deficit. Also, her laboratory findings were within normal limits.

CT scan revealed a hydrocephaly associated to a large, well circumscribed mass, with a broad attachment to the left lateral ventricle, near the foramen of Monro. The mass was lobulated and had two components: a solid one and a cystic one (Figure 1).

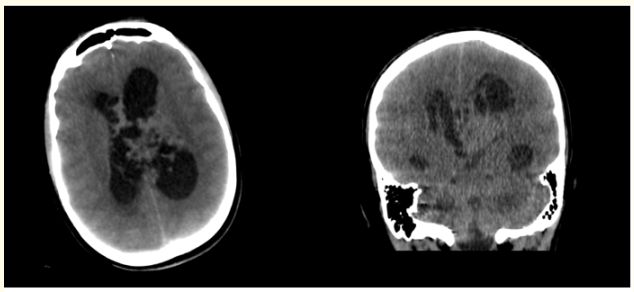

Figure 1: *Axial (a) and coronal (b) views of a CT showing a lobulated, intraventricular mass with two components: a solid isoattenuating one and a cystic one, associated with hydrocephalus. The tumor has a broad attachment to the superior and lateral wall of the left ventricle.*

On the MRI, the mass was heterogenous isointense to grey matter on T1 and T2 weighted imaging, and contained multiple cystic areas, giving it a soap-bubble like appearance. And on post contrast imaging, there was moderate and inhomogeneous enhancement of the mass lesion (Figure 2).

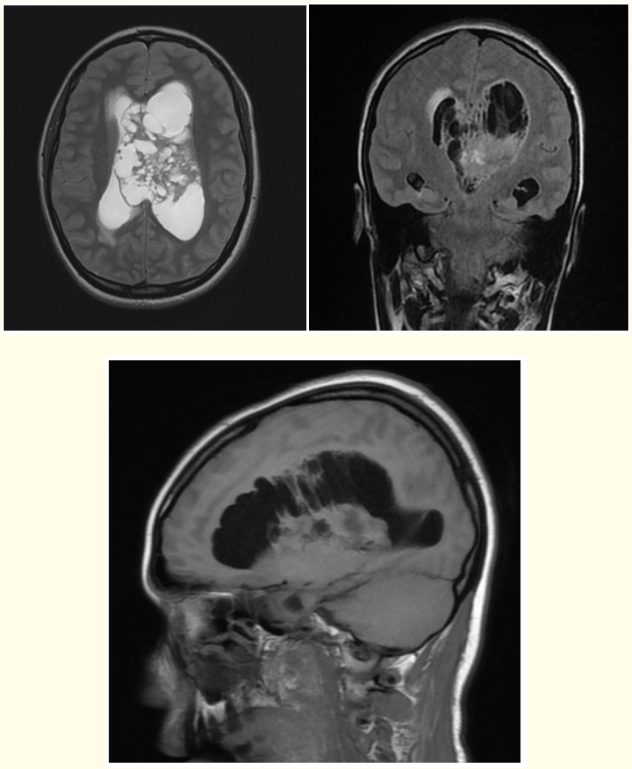

Figure 2: *(a) T2 axial (b) FLAIR coronal and (c) T1 sagittal MR images showing a lobulated, heterogenous, iso to hyperintense, intra ventricular mass with broad attachment showing multiple intratumoral and peritumoral cystic areas (soap-bubble appearance).*

Imaging characteristic and location of the lesion were highly suggestive of central neurocytoma. After resection, pathological diagnosis of tumor was central neurocytoma.

Discussion

Central neurocytoma was described for the first time by Hassoun., *et al*. in 1982, and is a pathology of young adults around the third decade, affecting both sexes similarly [1,2]. Initially, they were classified as WHO grade I lesions, however in 1993, this was updated to WHO grade II, as it was observed that at least some of these lesions showed a more aggressive behaviour [2]. Indeed, they can be derived from bipotential precursor cells of the periventricular germinal matrix capable of both neuronal and glial differentiation.

Patients present with signs and symptoms of increased intracranial pressure due to obstructive hydrocephalus. According to Schild., *et al*. study, those symptoms were most frequently headaches, and visual changes with insidious onset. Sometimes, patients may present with acute symptoms such as nausea or vomiting, related to sudden ventricular obstruction [2,4]. Finally, less commonly, the complaint is paraesthesia, lethargy, balance problems and tinnitus [4].

CN are thought to arise from septal nuclei, however the septum pellucidum is not seen well on imaging studies. Therefor the mass is typically centred in the midline near the foramen of Monro and has a broad-based attachment to the superior and lateral wall of the ventricle [2-4]. The obstruction of foramen of Monro is usually responsible for hydrocephalus. Tumor can spread through CSF and infiltrate the cerebellum, the brainstem and the peritoneum in patients with ventriculoperitoneal shunt. Their extra ventricular location is extremely rare, but some CNs have been previously described in the thalamus, cerebellum, pons, medulla oblongata and spinal cord [5].

Computed tomography depicts CN as an oval shaped masses, with sharply demarcated, polylobulated margins, isoattenuating or slightly hyperattenuating, within the body of the lateral ventricles with most frequently moderate heterogenous enhancement. Calcifications are found in approximately 51% of the cases, and they might be clumped, amorphous or globular [2,4,6].

MRI is more helpful in delimiting the tumor, specifying its insertion, but also analysing its different components. It usually demonstrates a heterogenous mass that is isointense on T1 weighted images, and iso to hyper-intense on T2-weighted images. In some cases, MRI might reveal areas of low signal intensity or absent signal on both T1- and T2 weighted images, representing hemorrhage, calcifications or tumor vessels. Cystic component is present more often than not, and is hyperintense on T2 WI [2,4]. The solid component of the tumor shows variable patterns of enhancement after gadolinium injection.

Xiando., *et al.* [7] brought together and described in the largest cohort of CN, six MRI features allowing to improve preoperative diagnostic accuracy. The "scalloping sign" consists in the presence of tumoral spicules attached to an undulated ventricular wall, better depicted on sagittal views, demonstrates a high specificity. CN's are thought to derive from the small grey nuclei of the septum pellucidum or bipotential progenitor cells in the periventricular matrix, which results in "the broad-based sign": the tumor usually lies alongside the septum pellucidum or the walls of the lateral ventricles with a broad base. This image feature has a high sensitivity, but might be present in ependymomas and astrocytomas in their intraventricular form as well. Therefore, it is interesting to analyse tumor margins for further clarity, as they are conspicuous in CNs while ependymomas and astrocytomas tend to invade the surrounding parenchyma and show a more aggressive behaviour. Furthermore, up to 85% of CN's have numerous cysts, giving the tumor a "soap bubble appearance", a sign with an excellent sensitivity. While cysts tend to occupy the center of other tumors because that is the most common site of ischemia and necrosis, they are found in the periphery of CNs. The "periphery cyst sign" is one of the three new signs described by Xiando., *et al*. that has a high specificity. The other signs are the "gemstone sign" consisting in the presence of one or multiple strikingly enhanced areas within a non enhanced or mildly enhanced surroundings in CN. It is thought to be caused by the existence of capillaries with a peculiar structure or some cavernous haemangioma-like changes. Finally, le "fluid-fluid level sign" usually results from haemorrhage [7].

Other exploration techniques such as proton MR spectroscopy make it possible to search for the specific metabolic characteristics of the neurocytoma. A glycine peak is deemed to be a valuable indicator of CNs, along with high choline and low NAA peaks [8,9]. It should be noted that high choline is not necessarily synonymous with proliferative potential; in fact, the majority of CNs have exhibited low proliferative potential. The presence of NAA indicates that CNs are of neuronal origin, but the metabolite's low level is likely due to the immaturity of the neurons of the tumor. Lactate peak is inconsistent in CNs. Chuang., *et al*. reported the presence of Alanine in CNs using 3-T MR systems, as an inverted doublet at 1.5 ppm with a TE of 135ms which is also characteristically observed in meningiomas, although the analyse of other mentioned metabolites should help differentiate both tumors [10]. Moreover, CNs seem to have higher NAA/Cr and Cho/Cr ratios and lower NAA/Cho compared to other intraventricular tumors [11,12].

On imaging, the differential diagnosis can be also approached depending on the exact location of the tumor and the age of the patient. Hence, intraventricular tumors in young adults include ependymomas, subependymomas, subependymal giant cell astrocytoma, choroid plexus papilloma, choroid plexus carcinoma, meningioma and intra ventricular metastasis. The typical CN has an affinity for the anterior half of the lateral ventricle, and some series even report a higher involvement of the left lateral ventricle. An extension to the third ventricle might occur in some cases, although isolated third or fourth intraventricular neurocytomas are rare [4]. Ependymomas mostly occur in the fourth ventricle and are more common in children. The main feature of such tumors is signal heterogeneity. On imaging, they typically show calcifications as well as haemorrhage, and a heterogenous enhancement of the tumor is observed on contrast enhanced studies. Subependymomas affects older adults compared to CN although they both have an affinity for the anterior half of the lateral ventricle. Subependymal giant cell astrocytomas are always located near the foramen of Monro, calcifications and intense enhancement are frequently observed features, and the diagnosis is made easier when other stigmata seen in tuberous sclerosis are found [2]. Choroid plexus papilloma, choroid plexus carcinoma, meningioma, or metastasis, are highly vascular tumors, and should be suspected when the tumor is centered on the choroid plexus [2,4,6].

The most efficient treatment is complete resection of the tumor, and CN have good prognosis with an overall survival rate at 5 years follow up ranging between 65.1% and 92.3% [13]. Patients might benefit from radiotherapy if the excision is incomplete.

Conclusion

CN are benign, slow-growing intraventricular tumours that predominantly affect young adults in their third decade of life. By utilising clinical, demographic, and imaging findings, the differential diagnosis for many common intraventricular tumours can be significantly narrowed. Surgery remains the most important therapeutic option. A safe maximal resection confers the best long-term outcome.

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