CASO CLÍNICO/CASE REPORT

Central Neurocytoma Mimicking a Hemorrhagic Ependymal Cyst

Neurocitoma Central que Mimetizando um Quisto Ependímico Hemorrágico

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Abstract

Central neurocytoma (CN) is a rare intraventricular tumor, predominantly affecting young adults without sex predilection. Differential diagnosis includes other intraventricular tumors, emphasizing the need for accurate preoperative diagnosis. Surgical resection remains the cornerstone of treatment, with favorable long-term survival outcomes.

We present a case of CN in a 65-year-old male presenting with diplopia and headache. Magnetic resonance imaging (MRI) revealed a multicystic right ventricular lesion without solid components, asymmetric ventriculomegaly, and evidence of prior hemorrhage. Histopathological examination confirmed CN. This case of an atypical appearing central neurocytoma, lacking solid components or enhancement and with evidence of prior hemorrhage highlights the importance of keeping central neurocytoma on the differential for an intraventricular mass, even when the typical findings are absent. Initial differential considerations were led with more benign entities, like hemorrhagic complications of a choroid or ependymal cyst.

Resumo

O neurocitoma central (CN) é um tumor intraventricular raro, que afeta predominantemente adultos jovens com igual prevalência em ambos os sexos. O diagnóstico diferencial inclui outros tumores intraventriculares, enfatizando a necessidade de um diagnóstico pré-operatório preciso. A ressecção cirúrgica permanece como o pilar do tratamento, com resultados favoráveis de sobrevivência a longo prazo. Apresentamos um caso de CN num homem de 65 anos com diplopia e cefaleia. A ressonância magnética revelou uma lesão ventricular direita multiquística sem componentes sólidos, ventriculomegalia assimétrica e evidência de hemorragia prévia. O exame histopatológico confirmou CN.

Este caso de CN com aparência atípica, sem componentes sólidos ou realce e com evidência de hemorragia prévia, destaca a importância de considerar o CN no diagnóstico diferencial de uma massa intraventricular, mesmo quando os achados típicos estão ausentes. Considerações diferenciais iniciais foram orientadas para entidades mais benignas, como complicações hemorrágicas de quistos do plexo coróide ou ependimários.

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Introduction

Central neurocytoma (CN) is a rare intraventricular tumor arranged as grade 2 by the latest World Health Organization (WHO) classification.[1,2] It is a neuronal-glial tumor with an incidence of 0.25%–0.5% of all intracranial tumors and a slowly growing intraventricular tumor occurring in young adults generally between 20 and 40 years with no sex predilection.[3] The most common location is the anterior portion of the lateral ventricles, often with extension into the third ventricle or bilaterally, and is usually attached to the septum pellucidum near the foramen of Monro.[2]

The symptoms caused by this tumor are non-specific and arise from increased intracranial pressure due to the obstruction of cerebrospinal fluid drainage.[2] Other clinical features can include behavioral disorders and seizures.[3] Although CN usually exhibits benign histology, in some cases, it may display more aggressive histologic features and can rarely demonstrate malignant behavior with craniospinal dissemination, including infiltration into the cerebellum, brainstem, and peritoneum in patients with a ventriculoperitoneal shunt.[2]

Case Report

A 65-year-old man presented with 3 months of worsening diplopia and a new onset headache. On physical exam, a 6th nerve palsy was demonstrated. Subsequently, the patient underwent magnetic resonance imaging (MRI), which revealed a non-enhancing multicystic right ventricular lesion without solid components, asymmetric ventriculomegaly, and evidence of prior hemorrhage (**Fig. 1**). The patient underwent endoscopic ventricular biopsy and partial mass resection. Within the right lateral ventricle, a white webbed lesion was encountered overlying the foramen of Monroe, wherein adhesions were noted. Samples were sent to pathology, which demonstrated a monomorphic proliferation of cells with round nuclei, a stippled "salt and pepper" chromatin distribution, and variably present nucleoli. Some cells have perinuclear halos imparting an oligodendroglioma-like morphology. Immunohistochemistry revealed that the neoplastic cells are diffusely positive for synaptophysin and show nuclear positivity for NeuN. GFAP was negative and OLIG2 highlighted a few scattered neoplastic cells (**Fig. 2**). The findings are consistent with central neurocytoma.

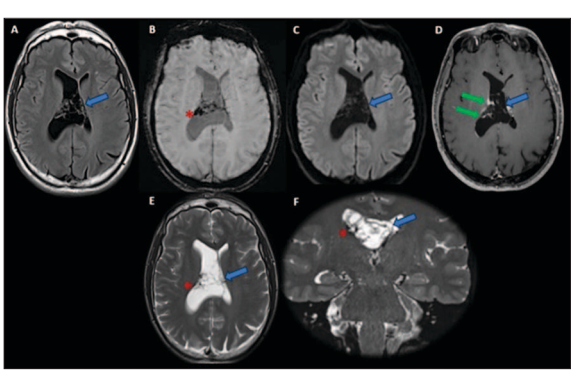

Figure 1. Brain magnetic resonance imaging (MRI) on axial FLAIR (A), SWI (B), diffusion (C), T1 post-contrast (D), and axial (E) and coronal (F) T2-weighted images. Large multiseptated cystic lesion (blue arrows) centered in the right lateral ventricle body and pushing away the septum pellucidum and the choroid plexus. There were signs of prior hemorrhage with multiple hemosiderin foci within the lesion and lateral ependymal wall (red asterisks). The only detected enhancement was related to the choroid plexus, and internal septations (green arrows).

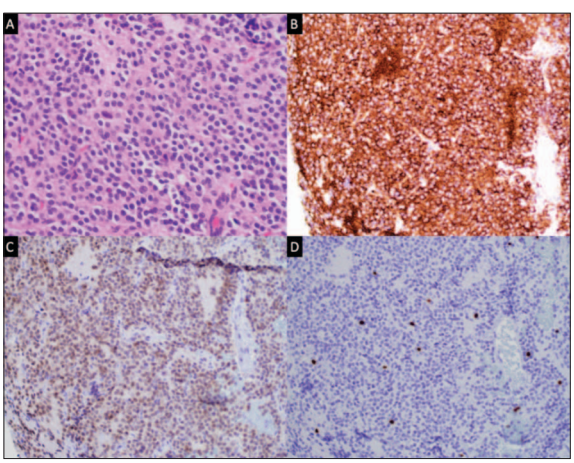

Figure 2. H&E stain showing a monomorphic proliferation of cells with round nuclei, a stippled "salt and pepper" chromatin distribution, and variably present nucleoli. Some cells have perinuclear halos imparting an oligodendroglioma-like morphology. (A; H&E; 200X magnification). Immunohistochemistry revealed that the neoplastic cells are diffusely positive for synaptophysin (B; 100X magnification) and show nuclear positivity for NeuN (C; 100X magnification). OLIG2 is mostly negative, with only rare stained nuclei (D; 100X magnification). GFAP (not shown) was negative.

Discussion

Diagnosis relies on radiologic and histologic findings.[4] On computed tomography (CT), central neurocytoma (CN) typically appears as a well-demarcated intraventricular mass that is isoattenuating or slightly hyperattenuating. Calcification is present in about half of the cases, and cystic changes are also reported. The key diagnostic feature is the tumor's location, usually attached to the foramen of Monro or the septum pellucidum with a broad-based attachment. However, other intraventricular tumors in adults should be considered in the differential diagnosis.[3]

MRI is the preferred imaging modality for accurately diagnosing central neurocytoma by providing a detailed analysis of the tumor and its various components. MRI is especially useful for distinguishing central neurocytoma from other diagnoses, with characteristic features such as a "spongy appearance" and "broad-based attachment."[3] Central neurocytoma is heterogeneous, appearing as an isointense mass on T1-weighted images and iso- to hyperintense on T2-weighted images. Calcifications, vessels, cysts, or hemorrhage, when present, display hyposignal or a signal on both T1 and T2 sequences, except for cysts which are hyperintense on T2-weighted images. Although cysts are usually present and have some characteristic features, they may not always be visible on imaging. Upon gadolinium injection, a degree of enhancement is typically observed.[3]

Magnetic resonance spectroscopy (MRS) is useful for differentiating central neurocytoma from meningioma and other intraventricular tumors. The spectroscopic profile of central neurocytoma is characterized by high levels of choline and glycine, low levels of N-acetylaspartate (NAA), and absent or low levels of alanine (Ala) compared to intraventricular meningioma. The choline/creatine and NAA/creatine ratios are significantly higher in central neurocytoma compared to other intraventricular tumors, such as oligodendroglioma, astrocytoma, subependymoma, and glioblastoma multiforme.[3] Dynamic susceptibility contrast-enhanced MRI perfusion shows that central neurocytoma has intermediate vascularity—greater than that of ependymomas and subependymomas but less than that of highly vascularized tumors like meningiomas, papillomas, and renal carcinoma metastases.[3]

Histologically, neurocytomas are composed of small, round cells with minimal variation in shape and size, but they exhibit variable architecture. Rosette-like areas or perinuclear halos can mimic ependymoma, pineocytoma, or oligodendroglioma. Immunostaining typically shows positivity for neuronal antigens and rarely for glial antigens. Electron microscopy reveals a high degree of neuronal differentiation.[5]

The primary treatment for central neurocytoma remains surgical resection, with the extent of resection playing a crucial role in prognosis. According to the 5th Edition of the WHO Classification of Central Nervous System Tumors (2021), the estimated 10-year overall survival rate is 82%, emphasizing the importance of complete resection whenever feasible.[5] Prognostic factors include the extent of resection and the Ki-67 labeling index, which, when elevated, is associated with a more aggressive disease course and may indicate the need for adjuvant therapy.[5] Tumors with a Ki-67 index above a certain threshold are often managed with additional therapies, such as radiotherapy, to reduce recurrence risk.[2]

This case of an atypical appearing central neurocytoma, lacking solid components or enhancement and with evidence of prior hemorrhage, highlights the importance of considering central neurocytoma in the differential diagnosis for intraventricular masses, even when typical findings are absent. ■

Contributorship Statement / Declaração de Contribuição

JK, SA, OL, OLN, MLD: Conception and design. Data analysis and interpretation. Writing and critical review of an important part of its intellectual content.

All authors approved the final version to be published.

JK, SA, OL, OLN, MLD: Conceção e desenho. Análise e interpretação dos dados. Redação e revisão crítica de parte importante do seu conteúdo intelectual.

Todos os autores aprovaram a versão final a ser publicada.

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