

with high local invasion, rapid growth, and early distant metastasis unless they are excised in a timely manner<sup>(2)</sup>. The most common locations for MPNST in neurofibromatosis patients are the extremities, head, and neck. Thoracic involvement, however, is remarkably rare, few cases having been reported<sup>(3)</sup>. According to the size and location of the intrathoracic tumor, compressive manifestations such as pain, dyspnea, dysphagia, and superior vena cava syndrome may be the presenting manifestations, as seen in our patient, who reported dyspnea as the sole symptom related to his MPNST<sup>(3,4)</sup>.

The identification of MPNST in neurofibromatosis patients may be troublesome for several reasons. First, the existence of multiple benign neurofibromas may delay the identification of changes in plexiform neurofibromas. In addition, because superficial cutaneous neurofibromas do not undergo malignant transformation, MPNSTs often remain undetected until they reach a moderate size or cause compressive symptoms. Furthermore, CT and magnetic resonance imaging might not be accurate enough to differentiate benign from malignant lesions with any degree of reliability in the very early stages, although advances have been made in the area of positron emission tomography<sup>(4-6)</sup>. Therefore, any suspicious lesions should generally prompt prompt histological sampling<sup>(7)</sup>.

Although the mainstay of successful treatment of an MPNST is surgical excision after disease staging, neoadjuvant chemotherapy may be employed in order to reduce its dimensions beforehand, especially in patients with lesions surrounding vital organs. Radiotherapy might also delay recurrence, although it has not been shown to improve survival in MPNST patients<sup>(8)</sup>.

## REFERENCES

- Hirbe AC, Gutmann DH. Neurofibromatosis type 1: a multidisciplinary approach to care. *Lancet Neurol*. 2014;13:834-43.
- Porter DE, Prasad V, Foster L, et al. Survival in malignant peripheral nerve sheath tumours: a comparison between sporadic and neurofibromatosis type 1-associated tumours. *Sarcoma*. 2009;2009:756395.
- Chao BH, Stogner-Underwood KA, Kiev J, et al. Intrathoracic malignant peripheral nerve sheath tumour in neurofibromatosis 1. *Journal of Clinical Oncology*. 2008;26:2216-8.
- Grimer R, Judson I, Peake D, et al. Guidelines for the management of soft tissue sarcomas. *Sarcoma*. 2010;2010:506182.
- Yap YS, McPherson JR, Ong CK, et al. The NF1 gene revisited – from bench to bedside. *Oncotarget*. 2014;5:5873-92.
- Salamon J, Veldhoen S, Apostolova I, et al. 18F-FDG PET/CT for detection of malignant peripheral nerve sheath tumours in neurofibromatosis type 1: tumour-to-liver ratio is superior to an SUVmax cut-off. *Eur Radiol*. 2014;24:405-12.
- Gutmann DH, Aylsworth A, Carey JC, et al. The diagnostic evaluation and multidisciplinary management of neurofibromatosis 1 and neurofibromatosis 2. *JAMA*. 1997;278:51-7.
- Brems H, Beert E, de Ravel T, et al. Mechanisms in the pathogenesis of malignant tumours in neurofibromatosis type 1. *Lancet Oncol*. 2009;10:508-15.

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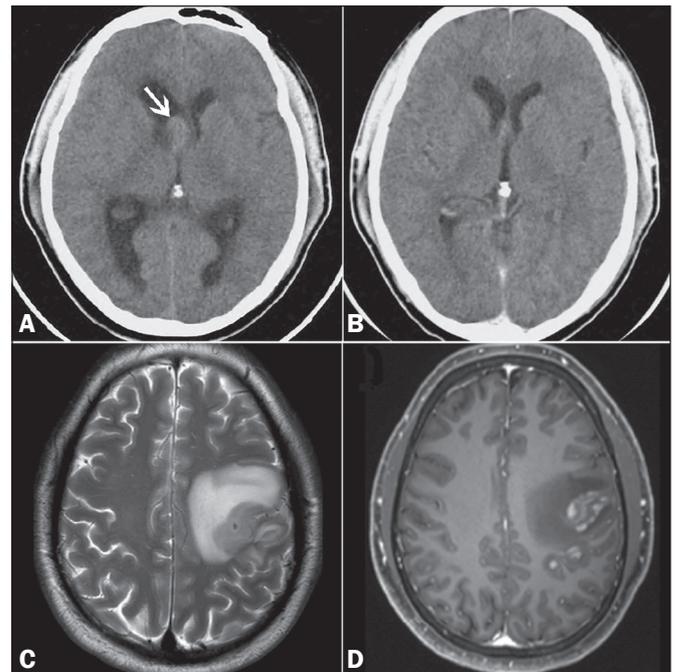
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### Burkitt-like lymphoma of the brain mimicking an intraventricular colloid cyst

Dear Editor,

A 32-year-old male sought treatment, complaining of headache. Computed tomography (CT) of the brain revealed hyperdense intraventricular nodule to the right of the foramen of Monro, highly suggestive of a colloid cyst (Figure 1A). The patient was using dexamethasone as pain therapy. In a CT scan of the brain obtained one month later, no nodules were observed (Figure 1B). Cervical and thoracoabdominal CT scans also showed no abnormalities. At two months, the patient presented with convulsions. Magnetic resonance imaging (MRI) of the brain showed a cerebral mass (Figures 1C and 1D). Histopathological and immunohistochemical analysis of a biopsy sample revealed Burkitt-like lymphoma, which is one of the non-Hodgkin lymphomas. Ancillary examinations ruled out systemic disease and viral infection.

Lymphomas are designated primary when they originate at and are confined to a given site<sup>(1-3)</sup>. Primary central nervous system (CNS) lymphomas account for up to 6% of brain neoplasms and 1-6% of extranodal lymphomas; approximately 90% of primary CNS lymphomas are non-Hodgkin lymphomas of the diffuse large B-cell subtype<sup>(1-6)</sup>. The incidence of CNS lymphoma is higher in the presence of certain immunodeficiencies, especially human immunodeficiency virus (HIV) infection<sup>(2)</sup>. Among immunocompetent individuals, the prevalence of CNS lymphoma is highest (60-67%) in men 45-75 years of age. In that group, CNS lymphomas present as a single homogeneous mass (in 62%), often in the supratentorial compartment (in 83%) and notably in the deep white matter (in 57%). The corpus callosum and regions surrounding the ventricles are typically affected. Perilesional



**Figure 1. A:** Non-contrast-enhanced CT scan of the brain, showing well-delineated, discretely hyperdense intraventricular nodule to the right of the foramen of Monro (arrow), promoting slight dilation of the lateral ventricles (obstructive hydrocephalus). **B:** Follow-up CT of the brain, obtained one month later, showing no such nodule. **C,D:** MRI of the brain after episodes of seizures, T2-weighted sequence (**C**) and paramagnetic contrast-enhanced T1-weighted sequence (**D**), showing an intra-axial frontoparietal mass in the left cerebral hemisphere, with intense perilesional vasogenic edema and heterogeneous enhancement.

edema is also common, being seen in 77–90% of cases<sup>(1,3–6)</sup>. On CT scans, CNS lymphomas are typically hyperdense, because they are hypercellular and have a high nucleus-cytoplasm ratio<sup>(1,3)</sup>. On MRI, they often demonstrate a hypointense or isointense signal in T1-weighted sequences and an isointense or hyperintense signal in T2-weighted sequences. After intravenous administration of contrast medium, they show homogeneous (90%) or, in rare cases, annular enhancement. They also exhibit signs of restricted water diffusion. Perfusion-weighted imaging shows less vascularization than that seen in other malignant brain tumors. On magnetic resonance spectroscopy, CNS lymphomas show elevated lipid and choline peaks, as well as a reduction in N-acetyl-aspartate levels<sup>(1,3–5)</sup>. The definitive diagnosis is made by biopsy<sup>(1,2,4,6)</sup>. Such lymphomas respond to chemotherapy and radiotherapy, the surgical option being used for tumor mass reduction<sup>(1,3–5)</sup>. Overall survival ranges from 15% to 80%, depending on the age of the patient, as well as on the characteristics and stage of the disease<sup>(2,4)</sup>.

The list of differential diagnoses of expansile CNS lesions in imaging studies is extensive, including glioma, acute ischemia, inflammatory processes, and infectious diseases<sup>(1,3–5,7–11)</sup>. When such lesions appear in an intraventricular location and are hyperdense on CT, they can be confused with colloid cysts, which are common at that site and exhibit similar density<sup>(4)</sup>.

Burkitt-like lymphomas are highly malignant, with cellular characteristics intermediate between those of diffuse non-Hodgkin large B-cell lymphoma and those of Burkitt lymphoma<sup>(12–14)</sup>. Burkitt-like lymphomas are typically associated with infection—HIV or the Epstein-Barr virus. They account for 2–3% of non-Hodgkin lymphomas in immunocompetent adults, being most common among the elderly<sup>(12–14)</sup>. Burkitt-like lymphomas can affect the brain, intestines, skin, ovaries, kidneys, liver, and bone marrow<sup>(12)</sup>. Chemotherapy is the most widely used treatment, although, even with treatment, survival is less than one year<sup>(13,14)</sup>.

The term “vanishing tumor” refers to a tumor that shows marked regression or disappears, with or without nonspecific therapy, and can recur or progress to new forms<sup>(2,4,15,16)</sup>. In the brain, lymphomas often occur after corticosteroid therapy, demyelinating diseases, or inflammatory disorders<sup>(15,16)</sup>.

## REFERENCES

- Mansour A, Qandeel M, Abdel-Razeq H, et al. MR imaging features of intracranial primary CNS lymphoma in immune competent patients. *Cancer Imaging*. 2014;14:22.
- Bellesso M, Bizzetto R, Pereira J, et al. Primary central nervous system lymphoma. *Rev Bras Hematol Hemoter*. 2008;30:54–60.
- Haldorsen IS, Espeland A, Larsson EM. Central nervous system lymphoma: characteristic findings on traditional and advanced imaging. *AJNR Am J Neuroradiol*. 2011;32:984–92.
- Sasani M, Bayhan M, Sasani H, et al. Primary central nervous system lymphoma presenting as a pure third ventricular lesion: a case report. *J Med Case Reports*. 2011;5:213.
- Reis F, Schwingel R, Nascimento FBP. Central nervous system lymphoma: iconographic essay. *Radiol Bras*. 2013;46:110–6.
- Alabdulsalam A, Zaidi SZA, Tailor I, et al. Primary Burkitt lymphoma of the fourth ventricle in an immunocompetent young patient. *Case Rep Pathol*. 2014;2014:630954.
- Castro FD, Reis F, Guerra JGG. Intraventricular mass lesions at magnetic resonance imaging: iconographic essay – part 1. *Radiol Bras*. 2014;47:176–81.
- Castro FD, Reis F, Guerra JGG. Intraventricular mass lesions at magnetic resonance imaging: iconographic essay – part 2. *Radiol Bras*. 2014;47:245–50.
- Destefani MH, Mello AS, Oliveira RS, et al. Chordoid glioma of the third ventricle. *Radiol Bras*. 2015;48:338–9.
- Schwingel R, Duarte SBL, Oshima MM, et al. Multiple hemangioblastomas, association with von Hippel-Lindau syndrome. *Radiol Bras*. 2015;48(2):xi–xiii.
- Dultra AHA, Noro F, Melo ASA, et al. Primary intercavernous lymphoma of the central nervous system. *Radiol Bras*. 2015;48:337–8.
- Simcock DE, O’Shaughnessy T, Balasanthiran A, et al. Pulmonary Burkitt’s-like lymphoma. *Respir Med Extra*. 2005;1:81–3.
- Re M, Di Massimo U, Romeo R, et al. Burkitt-like lymphoma of the sphenoid sinus: case report. *Acta Otorhinolaryngol Ital*. 2004;24:30–2.
- Johnson KA, Tung K, Mead G, et al. The imaging of Burkitt’s and Burkitt-like lymphoma. *Clin Radiol*. 1998;53:835–41.
- Okita Y, Narita Y, Miyakita Y, et al. Long-term follow-up of vanishing tumors in the brain: how should a lesion mimicking primary CNS lymphoma be managed? *Clin Neurol Neurosurg*. 2012;114:1217–21.
- Pohl P, Oberhuber G, Dietze O, et al. Steroid-induced complete remission in a case of primary cerebral non-Hodgkin’s lymphoma. *Clin Neurol Neurosurg*. 1989;91:247–50.

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## Giant cell tumor of the frontal sinus: a typical finding in an unlikely location

Dear Editor,

A 32-year-old female patient was admitted to the emergency room complaining of a knot on her forehead that had appeared 24 hours earlier. The patient underwent computed tomography (CT) of the skull, with and without intravenous administration of iodinated contrast medium. The CT scans revealed a dense, spontaneous, expansile extra-axial formation with its epicenter in the right frontal sinus, featuring an evident air-fluid level and well-defined borders (Figure 1A). On T2-weighted magnetic resonance imaging (MRI) sequences, the lesion also showed an air-fluid level (Figure 1B). A contrast-enhanced axial MRI scan showed peripheral enhancement (Figure 1C). The patient underwent surgery for complete resection of the lesion. The pathological examination demonstrated tumor-free margins, and im-

munochemistry showed that the lesion was characteristic of a giant cell tumor (GCT) of bone (Figure 1D).

GCT is one of the most common primary bone tumors, accounting for approximately 10% of all bone tumors and 25% of all benign bone tumors<sup>(1)</sup>. It mainly affects individuals 20–40 years of age and has an insidious onset, presenting with pain and a local increase in volume<sup>(1)</sup>. It is usually located in the epiphyses or metaphyses of the long bones, most commonly in the knees (distal femur or proximal tibia). Although it affects less than 1% of all bone sites within the skull (mainly the temporal and sphenoid bones), GCT tends to be more aggressive when it occurs at such sites<sup>(2–4)</sup>.

Based on the classical radiographic aspects, GCT of bone can be defined as a lytic, expansile lesion, resulting in thinning or erosion of the cortical bone<sup>(5)</sup>. CT is the best method to evaluate bone destruction and to identify pathological fractures. MRI can reveal soft tissue invasion and cystic areas (secondary