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Case Report

Imaging features of primary dural lymphoma: A report of 3 cases ☆,☆☆

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ABSTRACT

Primary dural lymphoma (PDL) constitutes a rare subgroup within central nervous system lymphomas, defined by its exclusive confinement to the dura mater, without a concurrent brain or systemic lymphatic involvement. This distinctive localization presents diagnostic challenges. In this report, we present a series of 3 cases where initial radiological presentations resembled meningiomas. We meticulously analyze key differentiating imaging characteristics, in CT, morphological MRI, and spectroscopy imaging. And show that recognizing and understanding these nuanced features are pivotal in enabling accurate differentiation of PDL and facilitating timely clinical intervention.

Introduction

Primary central nervous system (CNS) lymphoma is an extranodal non-Hodgkin lymphoma that arises from the brain parenchyma, eyes, meninges, or spinal cord in the absence of systemic disease. The most common type of primary CNS lymphoma presents as a space-occupying lesion in the brain parenchyma. Lymphoma arising primarily from the meninges without brain or systemic involvement is rare [1]. The reported cases of primary dural lymphoma (PDL) in the literature are limited, with only 105 cases documented [2].

This lymphoma subtype accounts for less than 1% of all central nervous system lymphomas and less than 0.1% of all non-Hodgkin's lymphomas [3,4]. Given the uncommonness of

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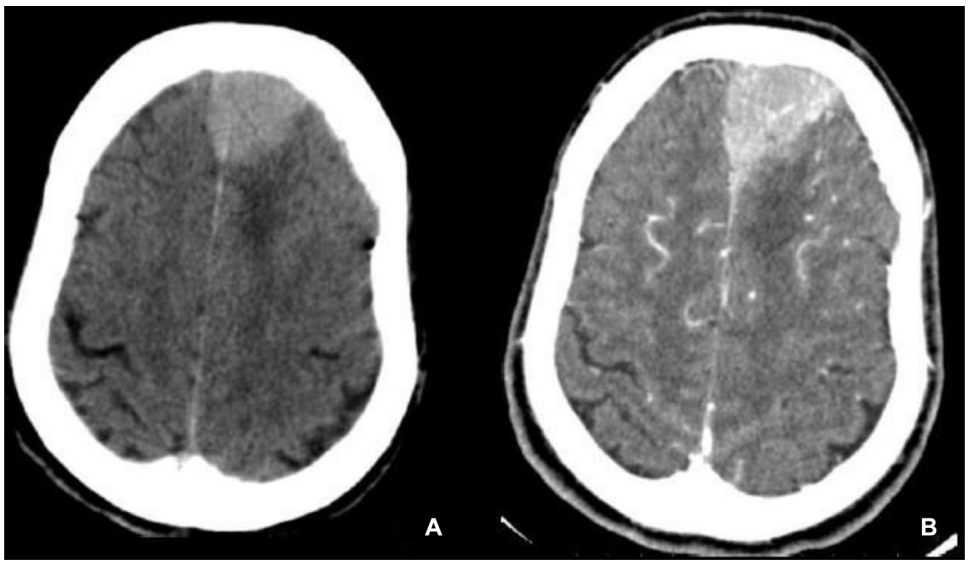

Fig. 1 – The precontrast CT scan (A) reveals the presence of an expansive extra-axial frontal median lesion that exhibits homogeneous hyperdensity relative to the cerebral cortex. The postcontrast CT slice (B) demonstrates intense and heterogeneous enhancement of the lesion.

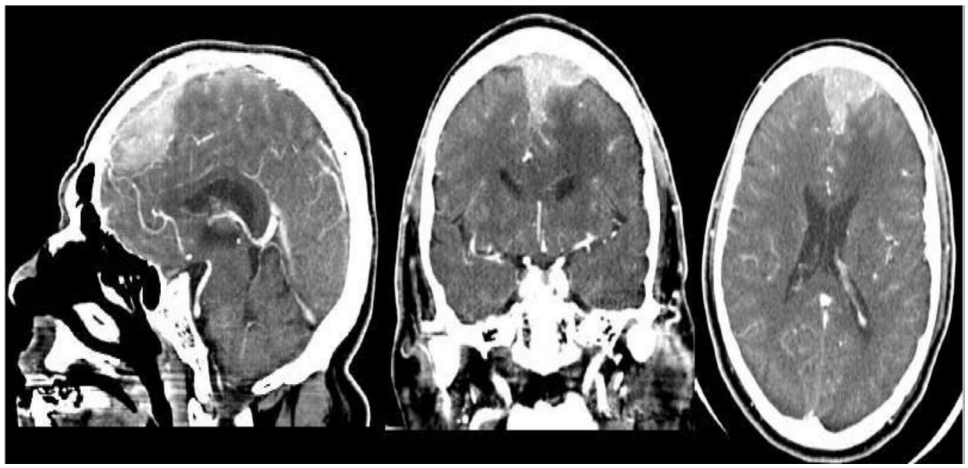

Fig. 2 – Cerebral CT scan after contrast agent injection: This tumor lesion has an irregular shape, polylobed margins, a rich vascular supply, and presents a broad base of implantation into the falx cerebri.

its symptoms, its discovery is usually incidental; otherwise, symptoms such as headache, seizure, focal sensory symptoms and visual disturbances can be found in some patients. PDL is often diagnosed as meningioma because of similar imaging findings, In O'Neil's series of 15 patients with PDL, a clinical and radiographic diagnosis of meningioma was made in 14 of 15 patients prior to histopathological diagnosis [5].

Magnetic resonance imaging (MRI) plays a major role in diagnosis that would affect therapeutic decisions which can be either surgical resection or postoperative focal radiotherapy. We report in our review 3 cases of extra PDL mimicking meningiomas.

Case reports

From 2012 to 2023, 3 patients with primary dural lymphoma were admitted to the Central Hospital of the Army of Algiers.

Patient 01

A male patient aged 51 years old, without any significant medical history or prior cranial trauma, arrived at the emergency department following a spontaneous, generalized seizure without fever. The subsequent neurological examination did

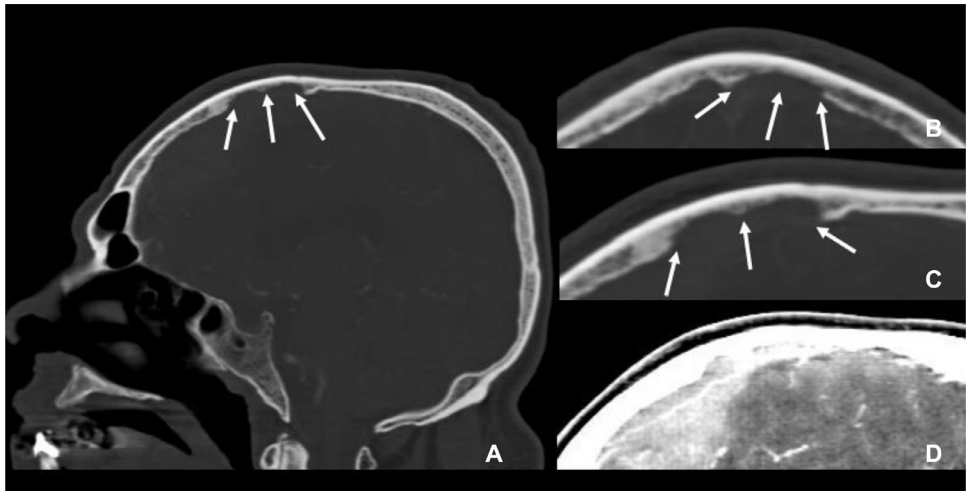

Fig. 3 – The CT scan with bone windows (A–C) reveals a lytic lesion of the cranial vault adjacent to the process (arrow), with irregularities predominantly observed at the level of the inner table. Following contrast agent injection (D), enhancement is observed adjacent to the inner tables.

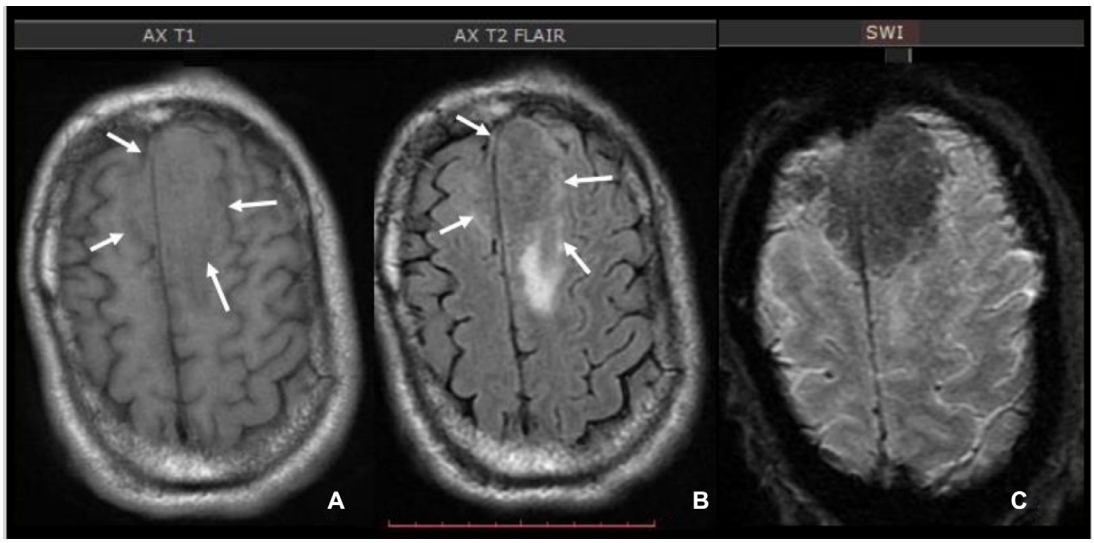

Fig. 4 – Axial T1-weighted image (A) Isointense signal, heterogeneous hypointense FLAIR surrounded by hyperintense signal (white arrows) (B) In T2*, there is a marked signal drop related to magnetic susceptibility (C).

not reveal any neurological deficits. As part of the investigation into the underlying cause.

A cranial computed tomography (CT) scan was ordered. The CT scan, performed using a 64 GE scanner before and after the administration of contrast agent, revealed the presence of an extra-axial mass of the left frontal lobe spontaneously hyperdense with a heterogeneous enhancement after injection of iodinated contrast media (Fig. 1). This lesion has an irregular shape, polylobate borders, rich vascularisation and a broad dural base (Fig. 2). Additionally, the CT scan showed evidence of bone destruction in the inner layer of the frontal bone (Fig. 3).

To further evaluate the mass, an MRI of the brain was performed. The MRI demonstrated that the mass exhibited an isointense signal on T1-weighted images (Fig. 4A), and on fluid-attenuated inversion recovery (FLAIR) imaging, it appeared heterogeneous and hypointense (Fig. 4B). The mass was surrounded by a thin layer of cerebrospinal fluid (CSF). On susceptibility-weighted imaging, the mass appeared hyposignal (Fig. 4C).

Notably, there was a significant presence of vasogenic edema surrounding the tumor, which manifested as a hyperintense signal on FLAIR images (Fig. 4B). Diffusion-weighted imaging revealed restricted diffusion within the mass, as evidenced by low apparent diffusion coefficient (ADC) values. Following the administration of a gadolinium-based contrast agent, the mass exhibited heterogeneous enhancement (Fig. 5).

Spectroscopic imaging of the mass revealed the presence of lipid peaks and the absence of an Alanine peak (Fig. 6).

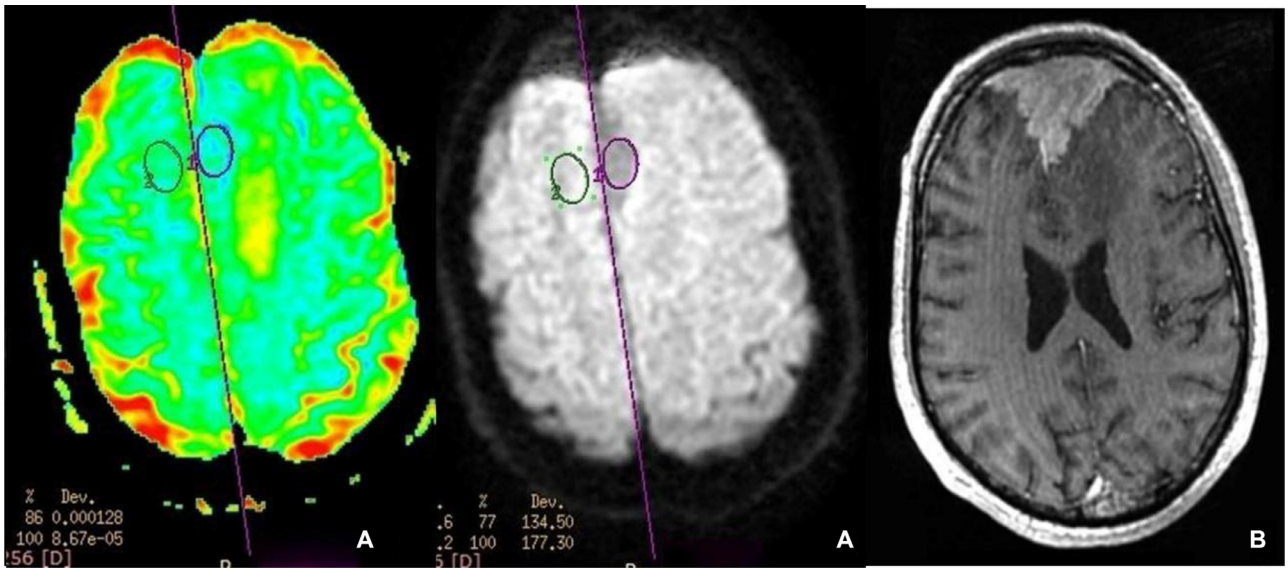

Fig. 5 – low ADC on diffusion-weighted imaging (A) Heterogeneous enhancement after injection of gadolinium (B).

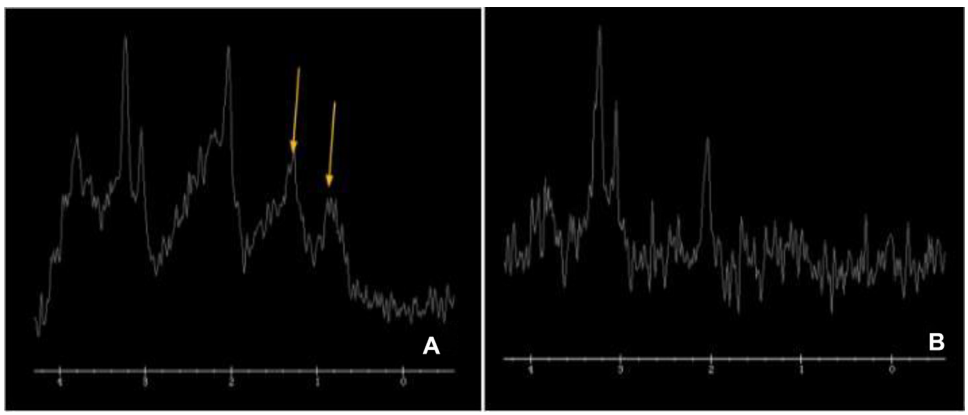

Fig. 6 – Spectroscopy imaging: Peak of lipids on short TE (yellow arrow) (A) long TE (B).

The neurosurgery team conducted a surgical excision of the tumor, which was subsequently sent for laboratory studies. The results confirmed the presence of PDL. Following the surgery, an MRI was performed, revealing a complete resolution of the mass (Fig. 7).

A scintigraphy and F18-FDG PET-CT were employed for remote extension assessment. No concerning metabolic activity was observed locally or distantly, indicating an absence of potential malignancy (Fig. 8).

Patient 02

A male patient, aged 68 years old, presented to the emergency room with a chronic unilateral headache. Upon examination, the patient had no significant medical history, and the physical examination was unremarkable.

A cerebral MRI was performed, revealing evidence of nodular pachymeningeal thickening in a plaque-like pattern in the right fronto-parieto-occipital region.

The thickening appeared isointense on T1-weighted images, and in the absence of T2-weighted imaging, diffusion-weighted imaging (DWI) was utilized, which also showed isointensity on T2-weighted and FLAIR images. Susceptibility-weighted imaging (SWI) revealed hypointense areas. Notably, there was a significant decrease in the ADC value. There was no evidence of vasogenic edema; subcortical hypersignal was observed (Fig. 10).

After the administration of a gadolinium-based contrast agent, the thickening exhibited intense and homogeneous enhancement (Fig. 11). The estimated thickness of the thickening was 15 mm, and there was associated regular leptomeningeal thickening. Additionally, close contact with the superior sagittal sinus was noted, which appeared to be invaded, allowing only a small passage with involvement of the superior anastomotic vein. Furthermore, there was no evidence of meningeal thickening elsewhere or other lesions. On T1-weighted post-contrast MRI sequence, a discontinuity of the T1 hypointensity of the cortical bone in the parietal region was observed, ac-

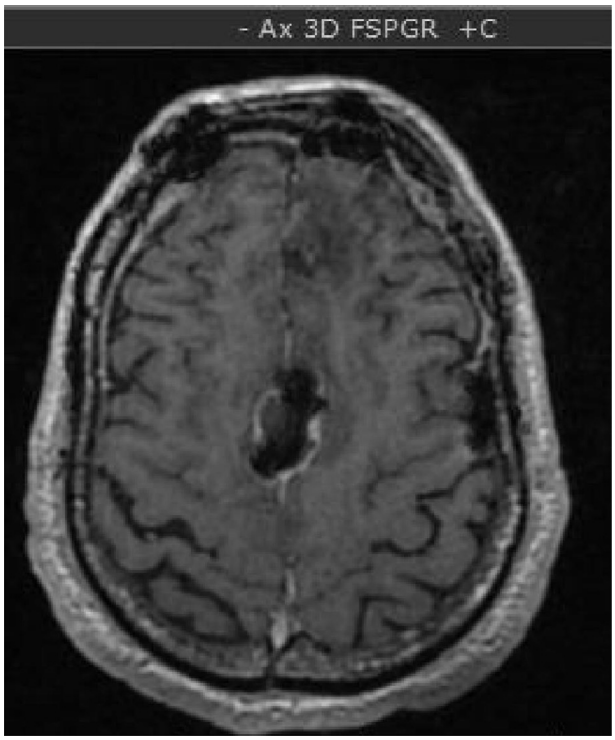

Fig. 7 – Complete resolution of the mass after surgery.

companied by thinning of the diploe involving the inner table (Fig. 9). Since bone involvement was not clearly appreciated on the MRI, and a CT scan was not initially performed, the patient underwent surgery and subsequently returned for a CT scan.

The CT scan clearly revealed bone involvement, showing an impression when compared to the contralateral side (Fig. 11). Biopsy with histopathological examination revealed a dural lymphoma. The patient was admitted to the hematology department where he received 8 sessions of chemotherapy with an R-CHOP regimen (cyclophosphamide, doxorubicin, prednisone, rituximab, and vincristine), an MRI was performed 7 later months, revealing total disappearance of the mass Fig. 12.

Patient 03

We report a case of a 29-year-old male patient suffering from persistent right otalgia and hypoacusis, priorly diagnosed with chronic otitis media in the right ear and consequently treated with antibiotics and corticosteroids. However, despite this treatment, the patient did not experience any improvement in symptoms. No noticeable biological abnormalities were found in his blood tests. A cerebral CT-Scan was performed, using a 64 GE scanner, and revealed the existence of an expansive intracranial extra-axial right temporal mass, with ill-defined limits, and a large dural base (Fig. 13). This mass was separated from the temporal bone by a fine CSF border, was isodense to the cerebral cortex and showed ho-

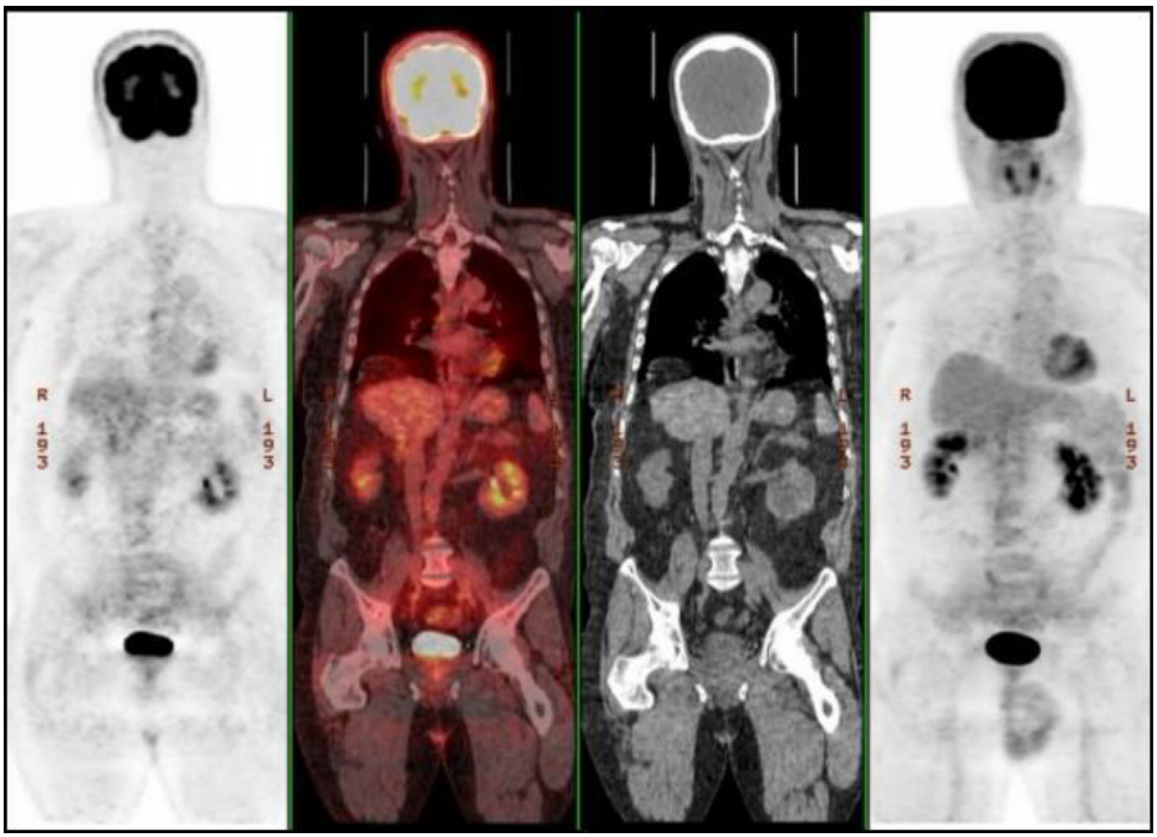

Fig. 8 – Scintigraphy and PET scan results: No concerning metabolic activity was observed locally or distantly.

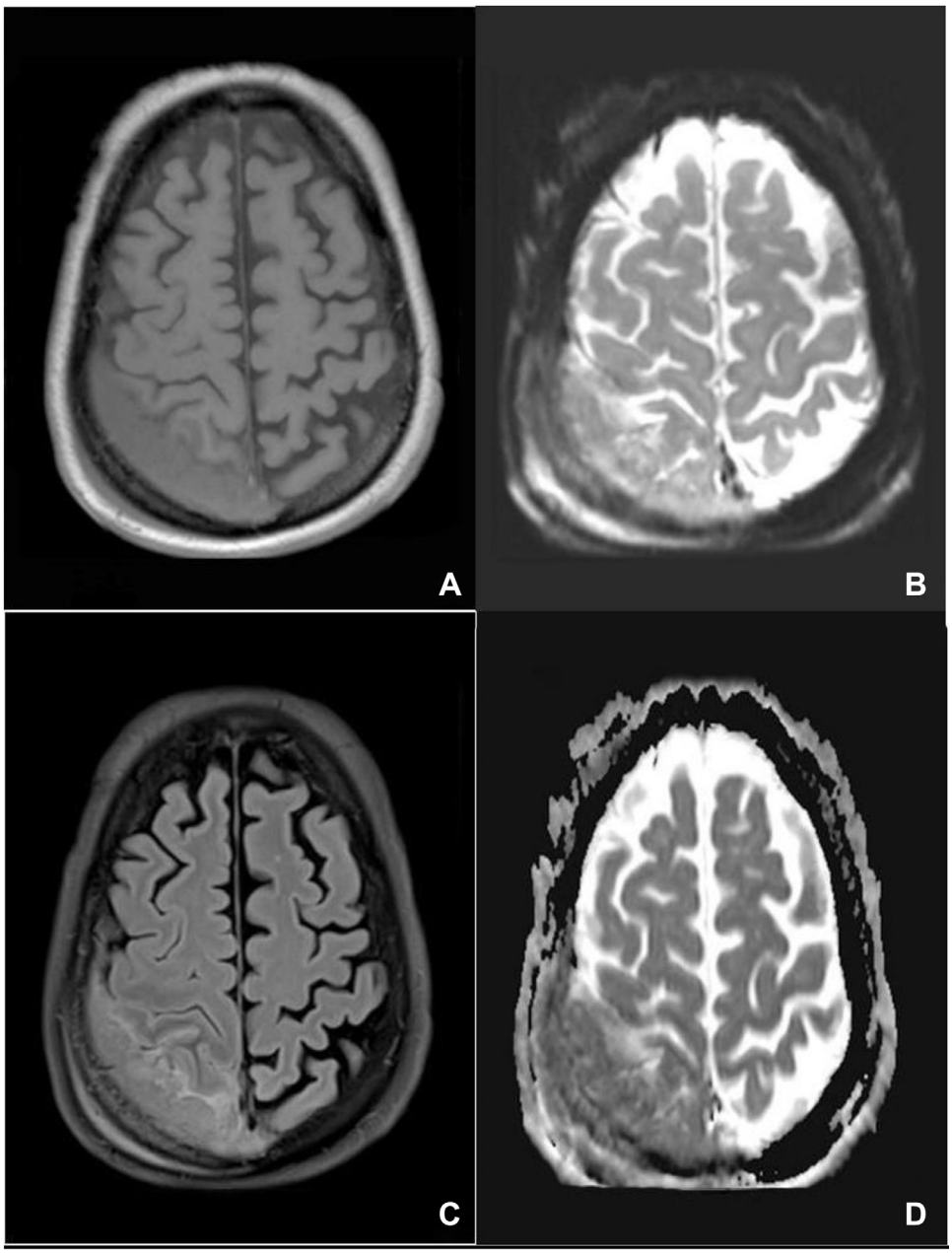

Fig. 9 – Nodular pachymeningeal thickening that appears as isointense on T1 (A), hyperintense on T2 (B) and FLAIR (C), significant decrease in ADC (D).

mogenous enhancement after iodine-based contrast agent injection (Fig. 13).

A large hypodense lesional area with no post-contrast enhancement was found surrounding the tumor, compatible with cerebral edema, which was responsible for a discrete mass effect on the right ventricle. Additionally, we noticed a filling of the mastoidian cells homolateral to the lesion. However, no venous sinus extension and no temporal bone invasion were evident. To broadly investigate this temporal expansive lesion, a MRI of the brain was performed. The MRI demonstrated a mass appearing iso-intense on the T1-weighted images, iso to hyperintense on T2-weighted images, and on DWI, the tumor appeared to be hyperintense with no restriction.

Following gadolinium injection, the tumor exhibited intense and homogenous enhancement (Fig. 13). A strong regular pachymeningeal enhancement was noted adjacent to the lesion. A large vasogenic edema area, hyperintense on FLAIR and DWI images with high ADC, was found surrounding the tumor (Fig. 14).

The MRI images provided valuable information, elucidating not only the infiltration of the pachymeninges (as pre-

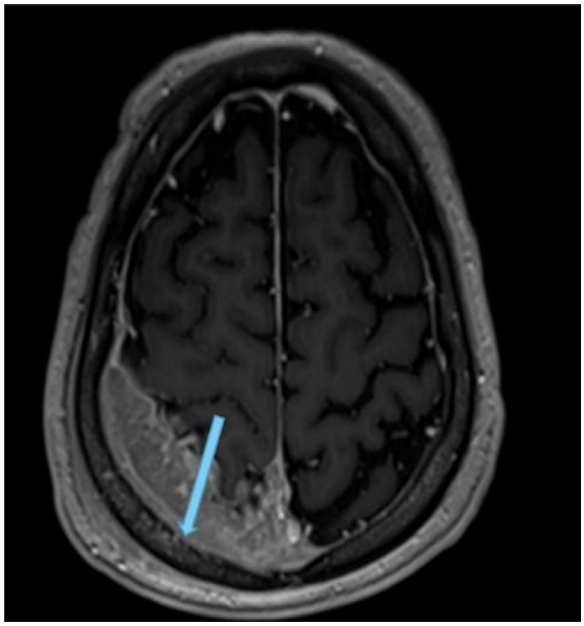

Fig. 10 – Intense and heterogeneous enhancement after the administration of gadolinium, discontinuity of the T1 hypointensity of the cortical bone in the parietal region (blue arrow). Note also the infiltration of the superior sagittal sinus (yellow arrow).

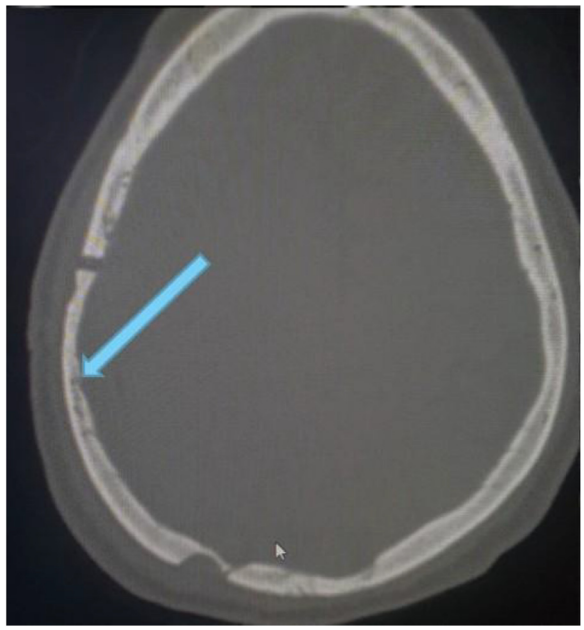

Fig. 11 – CT scan of the head shows the impression of inner table when comparing it to the contralateral side.

viously detected on the CT-Scan) in the right petrous apex and acoustico-facial bundle but also a distinct involvement of the hypoglossal canal extending into the post styloid space. Consequently, the tumor caused the anterior displacement of the internal carotid artery. Additionally, the infiltration of the mastoid cells was confirmed, along with the involvement of the geniculate ganglion, affecting the second (canalicular) and third (labyrinthine) segments of the right facial nerve (VII). The spectroscopy revealed the presence of a lipid peak, an increased Choline-to-Creatine ratio, and the absence of an Alanine peak (Fig. 15). After performing a surgical excision on the tumor. The findings from the laboratory studies conclusively verified the existence of PDL.

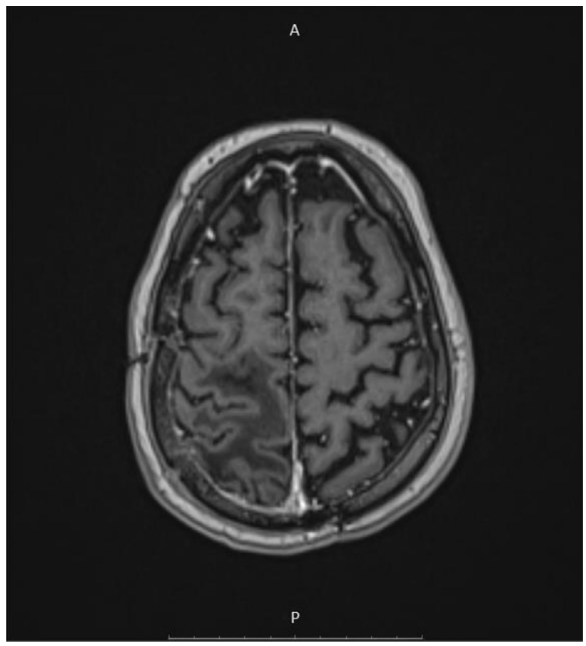

Fig. 12 – Axial T1 with gadolinium injection, showing complete resolution of the mass.

Discussion

Leptomeningeal lymphoma typically arises as a complication of systemic non-Hodgkin lymphoma. Individuals who are at a higher risk of leptomeningeal involvement often exhibit diffuse histologic characteristics, undergo a leukemic transformation, or have advanced disease (stage IVB) with bone marrow, visceral, or cutaneous involvement [6,7]. While meningeal disease may accompany the initial presentation of systemic lymphoma, it rarely occurs before the development of systemic lymphoma.

However, in cases where it does occur first, evidence of systemic disease usually follows within approximately 4 months. And although the expansion of primary CNS lymphoma into the leptomeninges is common, primary leptomeningeal lymphoma itself is uncommon [8], It generally presents as a low-grade malignancy, resulting in a favorable prognosis. Typically, it manifests as extra-axial tumors with diffuse enhancement following contrast agent administration, often resembling a meningioma and frequently leading to misdiagnosis.

In our 3 cases, the initial diagnosis involved confirming the extra-axial nature of the tumor, supported by several dis-

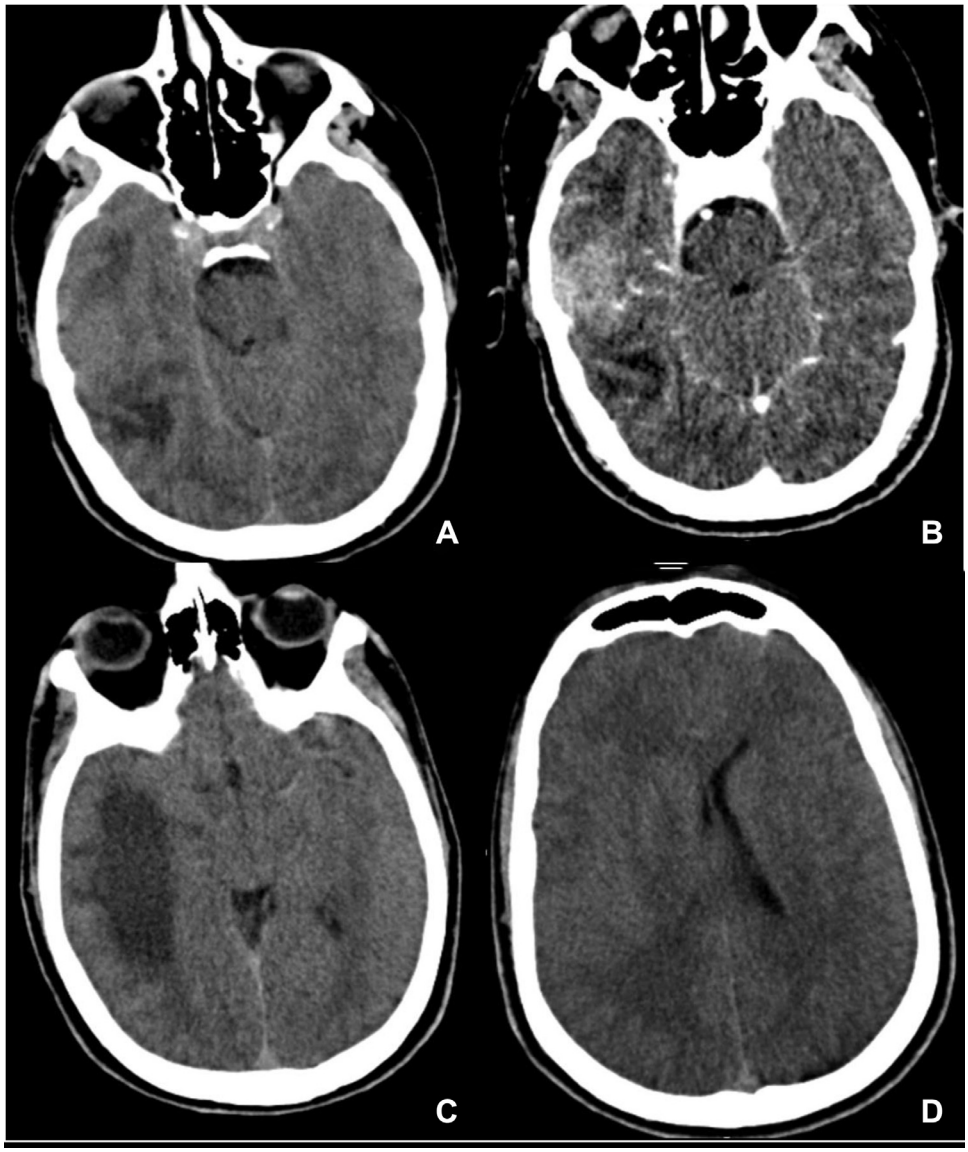

Fig. 13 – Cerebral CT-Scan: (A+B) extra-axial lesion with heterogeneous enhancement, (C) cerebral edema, (D) mass effect on the right ventricle.

cernible signs. The most apparent indicator is the prominent pachymeningeal thickening with the presence of a dural tail sign. Angles of connection between the lesion and adjacent structures are obtuse. The presence of the CSF cleft sign, clearly visualized on FLAIR imaging, indicates the presence of CSF serving as a barrier between the lesion and brain parenchyma. Its persistence on FLAIR can be attributed to the mixture of proteinaceous elements from the tumor. Evidence of bone involvement is also observed. Despite the tumor's considerable size, the absence of significant edema further supports its extra-axial localization. Intra-axial lesions typically exhibit significant edema. Additionally, there is an invasion of the venous sinus in 2 cases, confirming the extra-axial nature of the tumor.

In the context of extra-axial tumors, there are several potential differential diagnoses to consider. Meningioma is the most common with a prevalence of 53 per 100,000 people [9] and is characterised by various radiological features. It is typically isointense in T1 and T2-weighted imaging, shows variable diffusion without restriction, exhibits hypointense calcifications in all sequences, demonstrates intense and homogeneous enhancement after Gadolinium injection, hyperostosis of adjacent bone [10], higher grades of meningioma are more aggressive with unclear delimitation between the cortex and surrounding brain areas by Invasion of parenchyma, heterogeneous enhancement with areas of tumor necrosis, and osteolysis of adjacent bone can be seen in some cases [11]. Spectroscopic imaging also plays a crucial role by demonstrating a lipid peak [5] and the absence of alanine peak, the latter being characteristic of meningioma [12].

However, the presence of atypical features, such as clear demarcation with the adjacent cortex, heterogeneous en-

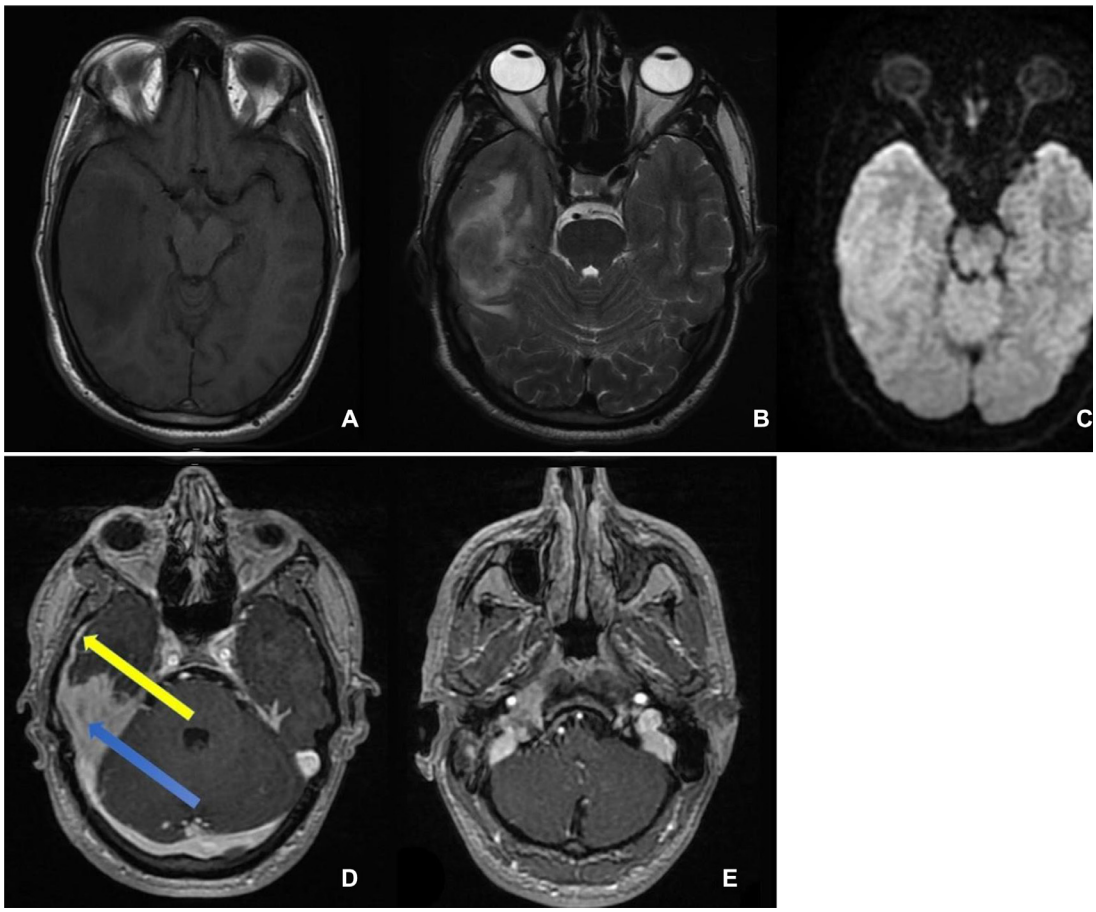

Fig. 14 – Brain MRI, isointense on T1 (A), isointense on T2 (B), no restriction of diffusion (C), heterogeneous enhancement (blue arrow), pachymeningeal enhancement (yellow arrow) (D), local extension (E).

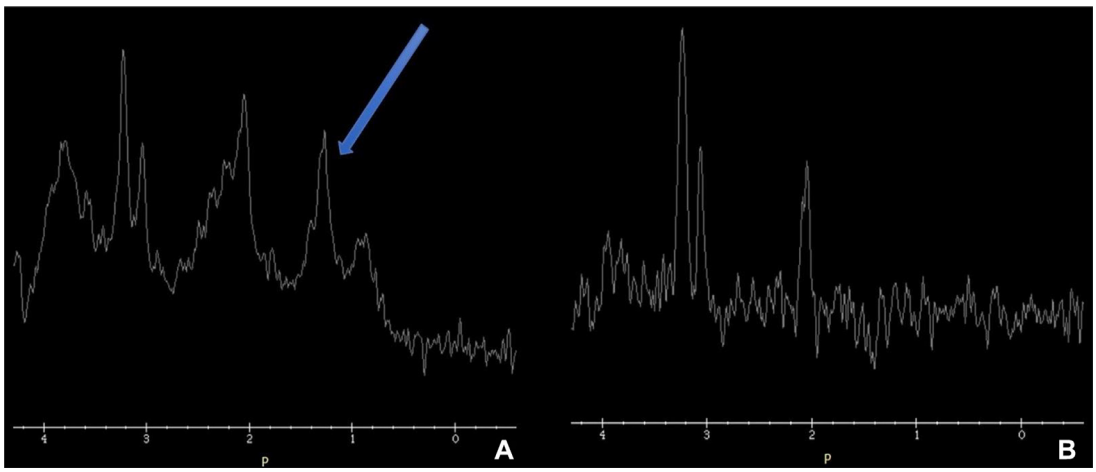

Fig. 15 – Spectroscopy imaging: Peak of lipids on short TE (blue arrow) (A), long TE (B).

hancement without necrotic areas, irregular contours, ADC restriction, increased lipid peak, and diploe lysis, diminishes the likelihood of meningioma as the underlying pathology in the 3 cases examined.

Metastasis originating from primary cancers, such as breast, lung, or prostate cancer, represents the second most frequent cause of extra-axial tumors. However, investigations conducted on the 3 male patients did not identify any primary malignancies capable of metastasizing to the dura.

In our 3 cases, none showed any indication of systemic involvement. There was no evidence of primary brain parenchymal participation. Brain imaging analysis revealed an extra-axial mass originating from the dura mater in all cases, suggesting a localised manifestation. Pathological examination confirmed the mass as lymphoma. Dural lymphoma exhibits noteworthy distinguishing imaging features, such as a blurred interface with the adjacent parenchyma, and bone lysis observed in both the first and second cases, instead of hyperostosis seen in meningioma. Additionally, hyperintensity on diffusion imaging with restricted ADC, suggestive of increased cellular density, is a characteristic finding, its absence in the third case is probably related to corticosteroids received by the patient [10]. However, it is essential to consider that a higher grade of meningioma may also manifest as a low ADC. Furthermore, dural lymphoma cases demonstrate potential involvement of the leptomeninges and a tendency to induce slightly more pronounced edema, as evident in the first and third patients.

These imaging distinctions contribute to the differential diagnosis of dural lymphoma and aid in distinguishing it from other intracranial pathologies, such as meningioma.

Conclusion

MRI plays an important role in the diagnostic guidance of extra-axial tumors, including distinguishing between dural lymphoma and meningioma. Namely, heterogeneous enhancement, hyperintensity with restricted ADC, and extension with bone infiltration resulting in osteolysis and dural venous sinus involvement leading to thrombosis. Spectroscopic imaging also plays a crucial role by demonstrating a lipid peak and the absence of an alanine peak, the latter being characteristic of meningioma. Biopsy and histopathological study remain the preferred examination to confirm the results, upon which the treatment depends.

Patient consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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