Primary dural lymphoma (PDL)

Clinical History:

A 55-year-old male patient was admitted to the hospital due to painless progressive swelling over the right frontal area for the last one year. The overlying skin was healthy. The patient referred occasional mild headaches. He had no history of fever, weight loss or night sweats.

Imaging Findings:

MR images showed a dural-based lesion that spread below the skull base. The tumour showed iso-signal intensity on T2 and FLAIR-weighted images (Fig. 1). DWI revealed restricted diffusion within the tumour with high signal intensity on DWI and corresponding low signal intensity on the ADC map (Fig. 2). T1- weighted image after contrast administration showed a homogenously enhancing extra-axial lesion with a dural tail sign (Fig. 3). CT scan with bone window showed right frontal bone sclerosis without bone destruction (Fig. 4).

A PET-CT scan of the whole body showed increased metabolic activity in a mediastinal lymph node and in the L5 vertebral body (Fig. 5). After chemotherapy treatment the increased metabolic activity disappeared (Fig. 6).

Histopathology of the resected dural mass showed a diffuse large B-cell lymphoma (Fig. 7).

Discussion:

Primary dural lymphoma (PDL) corresponds to an infrequent subtype of primary central nervous system lymphoma that carries a more favourable prognosis [1, 2]. The vast majority of PDL corresponds to low-grade lymphomas and few reported cases of other histologic subtypes have been described, including follicular, Hodgkin, and diffuse large B-cell [2-4]. Our reported case was diagnosed as diffuse large B-cell lymphoma (DLBCL) of the dura mater based on the histologic and immunohistochemistry features.

The dura is devoid of any lymphoid tissue, so pathogenesis of PDL remains unclear, and several hypotheses have been proposed, including chronic inflammation that could precede the malignant transformation of lymphoid cells [2, 3, 5].

Patients with PDL often complain of headaches, focal sensory or motor deficits, scalp swelling, meningeal irritations,
Diagnosis of PCNL with epidural or subdural involvement is based on imaging, and may result in a misdiagnosis of meningioma or other dural-based neoplasms [6]. Meningioma is the first differential diagnosis with major similarities in radiographic and clinical presentations [3]. Other possible differential diagnoses of PDL include epidural haematoma, haemangiopericytoma, meningeal metastases and meningeal sarcoma [6]. Morphological diagnosis of PDL relies on MRI examinations, but the confirmed diagnosis requires pathological examination. Dural lymphoma refers to a solid, confined mass, within or indistinguishable from the dura mater. The most common localisations are the cerebral convexities [2, 7]. The typical MRI appearance is an extra-axial mass attached to the dura, often with an enhancing dural tail, that enhance diffusely after contrast administration [1, 7]. Diffusion-weighted imaging (DWI) may also help to differentiate PDL from other dural masses because lymphomas are highly cellular tumours, so water diffusion is often restricted, making them appear hyperintense on DWI and hypointense on ADC maps. Scalp invasion without significant bone destruction is frequently seen because of the characteristic permeating growth pattern of lymphoma, with large soft tissue component and very little bone destruction [4, 7]. The presence of vasogenic oedema and parenchymal brain invasion is in favour of PDL more than meningiomas [1-3, 6].

Due to the paucity of cases, no standard treatment has been established. Various treatment combinations, including complete surgical resection, systemic chemotherapy and adjuvant radiation therapy have been attempted [3, 5, 6]. In the present case, despite the success of surgical resection, postoperative adjuvant chemotherapy was also employed. After treatment, follow-up with MRI is required owing to the high risk of systemic relapse.

**Differential Diagnosis List:** Diffuse large B-cell lymphoma in the dura with disseminated disease, Primary dural lymphoma (PDL), Invasive meningioma

**Final Diagnosis:** Diffuse large B-cell lymphoma in the dura with disseminated disease

**References:**


Lv ZW, Cheng KL, Tian HJ, Han XM. (2016) Primary diffuse large B-cell Lymphoma of the dura with scalp involvement; A case report and brief review of the literature. Oncology letters 11:3583-8

Figure 1

Description: Dura mater, bone and skeletal muscle infiltrated by neoplastic proliferation of high cell density, constituted by large cells, irregular nuclei and scarce cytoplasm (H&E, x4) Origin: Hospital 12 de Octubre. Department of Anatomical Pathology, Madrid 12 Spain.
**Description:** Dura mater, bone and skeletal muscle infiltrated by neoplastic proliferation of high cell density, constituted by large cells, irregular nuclei and scarce cytoplasm (H&E, x10)

**Origin:** Hospital 12 de Octubre. Department of Anatomical Pathology, Madrid 12 Spain.
Description: After paramagnetic contrast intravenous injection, the lesion demonstrates intense enhancement. Origin: Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
Description: Axial DWI high b-value. Diffusion restriction is demonstrated. Origin: Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
Description: ADC map displays low ADC. Origin: Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
Description: A PET-CT scan of the whole body demonstrates increased metabolic activity in a posterior mediastinal lymph node. Origin: Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
**Description:** A PET-CT scan of the whole body reveals increased metabolic activity in the L5 vertebral body. **Origin:** Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
Description: After chemotherapy treatment a PET-CT scan of the whole body shows that the increased metabolic activity had disappeared. Origin: Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
**Figure 6**

**Description:** The tumour shows iso-signal intensity on T2WI. **Origin:** Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
Description: The tumour shows iso-signal intensity on FLAIR. Origin: Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.
Description: CT scan with bone window shows right frontal bone sclerosis and involvement of both cranial tables with periosteal reaction. Note the absence of bone destruction. Origin: Hospital 12 de Octubre. Department of Radiology, Madrid, Spain.