Primary CNS Lymphoma - atypical imaging findings. Think about Lymphomatosis cerebri.
Published on 25.06.2009
DOI: 10.1594/EURORAD/CASE.7371
ISSN: 1563-4086
Section: Neuroradiology
Case Type: Clinical Cases
Authors: Bastos Lima P1, Parreira T1, Pereira R2, Rio F1.1Neuroradiology, 2Neurosurgery, Coimbra University Hospitals, Portugal.
Patient: 65 years, female

Clinical History:

Complaints of dizziness, headaches and vomits since 3 moths.

Imaging Findings:

A 65 year old otherwise healthy woman presented with headaches, vomits and dizziness with gait disturbance since 3 moths and worsening. Neurological examination showed some confusion, ataxic gait and dysmetria. Initial lab and imaging studies were done. Lumbar punction was inconclusive in relation to presence of neoplastic cells. MRI showed a diffuse infratentorial involvement, with some areas isointense to gray matter on T1WI (figure 1) and hyperintense on TR long sequences (eg.figure 3), mainly involving cerebellum (hemispheres and vermis), posterior pons (near IV ventricle), midbrain and posterior thalami. Some mass effect is seen, in particular in the fourth ventricle and Sylvius aqueduct (axial and parasagittal images- fig. 1b and 4b). After gadolinium, there are some cerebellar areas of predominant enhancement, without enhancement in the brainstem (figure 4).

Biopsy was done and the diagnosis was of CNS Lymphoma, with diffuse large B-cells (non-Hodgkin type). She began treatment with dexamethasone.

FDG-PET scan showed only brain (posterior fossa) hypermetabolism, suggesting Primary Central Nervous System Lymphoma - PCNSL (figure 5 a/b).

Progressive worsening was noted during some months, at last also with confusion and tetraparesis (more pronounced at left side). CT scan was done and showed a new supratentorial lesion (on right parietal and occipital lobes).

In spite of treated with high-dose methotrexate and radiotherapy, she died some months after initial diagnosis (5 months).

We present a case of an immunocompetent patient and neuroimaging studies showed initial diffuse infratentorial involvement and, some months latter, a new supratentorial lesion.

Discussion:

MRI findings are atypical for PCNSL : multiple high signal intense foci in the posterior fossa on long TR images, seeming an infiltrative process with areas of strong enhance and others without enhancement after contrast.

DWI (not performed) would be useful for diagnosis, because PCNSL have restricted diffusion. Initial studies point that Perfusion-WI may be helpful in the diagnosis of any kind of CNS lymphoma showing low rCBV. Differential considerations includes other neoplasms, such as gliomas, metastases and "lymphomatosis cerebri" (LC) subtype of PCNSL. Initial histopathology showed an evidence of mass lesion and areas of scattered lymphoid cells in the perivascular region; another focal lesion later - the final diagnosis was PCNSL. In this case we thought about LC,
variant of PCNSL, an uncommon pattern of lymphoma cell spread analogous to a rare diffusely infiltrative type of glioma known as gliomatosis cerebri (scattered tumour cells in the matter without a discernable cohesive mass formation). The cases reported of LC (diagnosis by autopsy) are associated with progressive cognitive decline, showing widespread and diffuse tumour infiltration of individual lymphoma cells in the cerebral white matter. PCNSL is nowadays considered an important differential diagnosis of brain lesions. Although uncommon, its incidence has increased dramatically over the last decades as a result of its occurrence (near 6%) in AIDS patients, but also being more frequent in immunocompetents.

The origin of the PCNSL remains unknown. CNS doesn't have endogenous lymphoid tissue or lymphatic circulation. It has been postulated that it develops from reticulum cells (normally found in the meningeal and Virchow-Robin spaces at the brain periphery); others think that arises from microglial cells. The latter theory is topographically appealing because PCNSL predilection for the deep or midline regions of the brain, being different from metastatic forms of lymphoma, which invade the brain surface through the perivascular spaces of penetrating arterioles. Nearly all of PCNSL are of non-Hodgkin type.

The common initial presentation is at the sixth and seventh decades of life, with single or multiple focal intracerebral masses, with a higher incidence of multiple lesions in AIDS patients. Up to 75% of lymphomatous masses seem to be in contact with ependyma, meninges or both. In recurrent disease, the subarachnoid space is often involved. The supratentorial compartment is most commonly involved (75-85%), affecting periventricular white matter and deep gray matter, and mainly in the frontal or temporal lobes. Involvement of corpus callosum or the central gray matter (basal ganglia, thalamus, hypothalamus) is not rare and more frequently than other CNS neoplasms. The lesions do not calcify, and haemorrhage is uncommon.

The imaging findings are not specific for lymphoma. Secondary brain involvement by systemic lymphoma is indistinguishable from PCNSL on MRI, although parenchymal involvement is more common in PCNSL. MRI signal intensity characteristics may be quite variable, ranging from marked hyperintensity to marked hypointensity (high cellularity). Although typically PCNSL enhances solidly and avidly, in the setting of AIDS there is a spectrum of patterns of pathologic enhancement ranging from solid to ring-like.

**Differential Diagnosis List:**
- Primary CNS Lymphoma - Non-Hodgkin type; diffuse large B-cell Lymphoma

**Final Diagnosis:** Primary CNS Lymphoma - Non-Hodgkin type; diffuse large B-cell Lymphoma

**References:**


Figure 1

a

Description: It reveals only brain hypermetabolism and localized at posterior fossa (cerebellum).

Origin:

b

Description: Origin:
**Description:** FLAIR images better depicts hyperintense infiltrative foci at cerebellum (hemispheres and vermis), pons (tegmentum of pons and medial lemniscus), midbrain and posterior thalami. Some mass effect is better seen, in particular in the fourth ventricle and Sylvius aqueduct. **Origin:**
**Description:** It shows isointense infiltrating lesions in the posterior fossa, mainly in the cerebellar white matter (more at right), cerebellar vermis and brainstem (posteriorly, near superior IV ventricle). Some mass effect is seen in axial and parasagittal images. **Origin:**

**b**

**Description:** Parasagittal images show better the infiltrate/mass effect of lesions in cerebellar white matter. **Origin:**
**Figure 4**

*a*  
Description: TR long images reveals high signal intensity of the lesions, better seen on T2WI and FLAIR. Origin:

*b*  
Description: T2WI shows high signal intensity lesions involving the cerebellum and brainstem (middle pons - fig 2b; midbrain - fig 2c) Origin:
Description: Origin:
Figure 5

Description: Intense and irregular contrast-enhancing lesions in the cerebellum. Origin:

b

Description: Parasagittal images shows better the enhanced lesions, minly involving cerebellar white matter and inferiorly. At midline, the enhancement seems to design the cerebellar folia, involving the superior cerebellar peduncle and medial cerebellar nucleus (fastigial nucleus). Origin:
Description: Coronal planes show that one area has ring enhance, inferiorly. Origin:
Description: Another brain lesion: supratentorial, right medial parieto-occipital (near the ependyma), with mass effect (adjacent ventricular system) and strong enhance after contrast. Origin: