Case 11736

Cranial nerve palsy and lower limb weakness – An unusual presentation of occult testicular lymphoma.

Published on 16.04.2014

DOI: 10.1594/EURORAD/CASE.11736
ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Head and neck Neuroradiology spine
Genital / Reproductive system male Molecular imaging
Procedure: Imaging sequences
Procedure: Equipment
Procedure: Diagnostic procedure
Procedure: Image compression
Imaging Technique: MR
Imaging Technique: Ultrasound
Imaging Technique: PET-CT
Special Focus: Lymphoma Case Type: Clinical Cases
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Patient: 48 years, male

Clinical History:

A 48-year-old man presenting with progressive lower limb weakness and multiple cranial nerve palsies was referred for neuroimaging by the Neurology department. He was otherwise well with no other clinical abnormality and no relevant past medical history. He had no scrotal symptoms.

Imaging Findings:

MRI head and spine including gadolinium imaging showed pathological enhancement of cranial nerves (III, V and VIII), leptomeningeal surface of the cervical spine and the cauda equine [figures 1-5]. The differential diagnosis including inflammatory and neoplastic processes was considered but after correlation with lumbar puncture findings, malignancy was placed higher in the differential.

Whole body computed tomography (CT) was unremarkable. A positron emission tomography (PET-CT) scan was performed. This demonstrated high tracer uptake from spinal nerves, cord surface, cauda equina, left testis and adrenals glands, due to disseminated disease [figures 6 A-C, 7]. The highest uptake was noted to be within the left testicle [figure 7].

Ultrasound of the testes identified an ill-defined low echo poor focus in the left testis, corresponding to the PET findings [figure 8]. Core biopsy of the left testis allowed histologically proven non-Hodgkin’s lymphoma (NHL). A presumptive diagnosis of primary testicular NHL with leptomeningeal dissemination was made.
Discussion:

Neurolymphomatosis (NL) is the term applied to infiltration of the nervous system by lymphoma and non-tumour lymphocytes cells. [1] 90% of the cases of NL have been related to Non-Hodgkin’s Lymphoma (NHL). [2] Approximately 50% of the patients with central nervous system (CNS) metastases from NHL have progressive systemic lymphoma at the time of diagnosis of their CNS manifestation. The remaining 50% develop systemic disease within months. [3]

Primary testicular lymphoma is a rare but highly aggressive disease, representing 5-9% of all testicular tumours, with a 5-year survival rate ranging from 16-50%. [4, 5] Approximately two-thirds of the patients present with leptomeningeal spread and as shown in this case, leptomeningeal spread can often involve the cranial nerves, spinal cord surface, or spinal nerve roots and may present with cranial or spinal neuropathy.

Central nervous system lymphoma can be diagnosed with gadolinium-enhanced MRI with leptomeninges, ependyma, dura, or cranial nerves being the typical finding. There may be intrinsic superficial cerebral lesions and communicating hydrocephalus. It is recommended that any pre-emptive lumbar puncture is performed after MRI imaging of the CNS, to avoid false positive cranial or dural enhancement due to the lumbar puncture. [4]

MRI typically shows enhancement of affected nerves, and occasionally enlargement, as shown in this case, with the third nerve [figures 1] (arrow). MRI does not always provide ideal visualisation of lymphomatous involvement of peripheral nerves and therefore 18F-FDG PET/CT should be performed to complete the radiological staging. [6] Sensitivity and specificity of 18F-FDG PET/CT for the initial staging of NHL is reported to be 97 and 100%, respectively and is increasingly becoming a key part of the investigation of NHL. [3, 7]

As highlighted in this case, the presentation of neurolymphomatosis and staging of NHL, requires integrating information gained from MRI, 18F-FDG PET/CT, CSF cytology examination, but most importantly a high index of suspicion by the clinician and radiologist.

Differential Diagnosis List: Non-Hodgkin’s Lymphoma, Metastasis, Granulomatous meningitis/infection, Sarcoidosis

Final Diagnosis: Non-Hodgkin’s Lymphoma

References:

**Figure 1**

*Description:* Axial post contrast (gadolinium-enhanced) T1 weighted MRI showing bilateral abnormal enhancement and enlargement of the oculomotor nerves (arrow heads). *Origin:* Madani H, Department of Radiology, Royal Free Hospital.
Figure 2

Description: Pre (left) and post (right) gadolinium axial T1 weighted MRI images, showing abnormal enhancement of both trigeminal nerves (arrow heads). Origin: Madani H, Department of Radiology, Royal Free Hospital.
**Figure 3**

**Description:** Pre (left) and post (right) gadolinium axial T1 weighted MRI showing two abnormal enhancing foci involving the right vestibulocochlear nerve (arrow head). **Origin:** Madani H, Department of Radiology, Royal Free Hospital.
Figure 4

**Description:** Post contrast (gadolinium-enhanced) sagittal T1 weighted MR cervical spine images, with a magnified view on the right, showing leptomeningeal enhancement along the brainstem and spinal cord (arrow heads). **Origin:** Madani H, Department of Radiology, Royal Free Hospital.
Description: Sagittal T2 weighted (left) and post gadolinium (right) lumbar spine MRI showing subtle thickening of the cauda equina on the T2 image and enhancement of the cauda equina on the post gadolinium image (arrow heads). Origin: Madani H, Department of Radiology, Royal Free Hospital.
Description: 18F-FDG PET/CT showing increased activity of (A) the C8 dorsal root ganglia bilaterally, (B) adrenal glands, (C) lumbar nerves roots and cauda equina (arrow heads). Origin: Madani H, Department of Radiology, Royal Free Hospital.
Description: 18F-FDG PET/CT showing increased activity within the left testis (arrow head). Origin: Madani H, Department of Radiology, Royal Free Hospital.
Description: Single longitudinal view of ultrasound of the left testis, showing an abnormal poorly defined echo poor region due to lymphoma (arrow head). Origin: Madani H, Department of Radiology, Royal Free Hospital.