Interesting images

Neurolymphomatosis as initial manifestation of recurrence in lymphoma

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We present the case of a 71-year-old woman diagnosed with diffuse large B-cell non-Hodgkin lymphoma, stage IV-B, IPI 3, in complete remission after 6 cycles of rituximab-CHOP.

Five months after completing treatment the patient went to the Emergency Department for a picture of headache and diplopy of one month of evolution. During admission she present lumbosciatalgia refractory to pharmacologic treatment. Lumbosacral MR was performed showing no signs of lymphomatous disease (Fig. 1). CSF study demonstrated relapse of lymphoma in the CNS and intrathecal therapy was initiated. At the end of the treatment 

¹¹F-FDG/PET-CT showed hypermetabolic foci in axillary, lumbar and pelvis regions compatible with nervous system tumoral involvement (neurolymphomatosis). In addition, hypermetabolic foci in the humeral head and sacrum were observed compatible with bone involvement by lymphoma (Fig. 2).

Lymphatic infiltration of the peripheral nervous system (neurolymphomatosis) is a rare manifestation of non-Hodgkin lymphoma. It is usually presented in patients with very extended non-Hodgkin lymphoma or as the first manifestation of relapse.

The onset of symptoms is generally progressive and insidious, presenting only radicular pain or associated to central or peripheral sensitivo-motor neuropathy.

Pain is the most common clinical presentation, generally being intense and described as relentless and, sometimes, burning pain. Neuropathy may present clinical symptoms similar to the Guillain-Barré syndrome, cauda equina syndrome (horse-tail) or quadriparesia, and differential diagnosis should include chemotherapy-induced neurotoxicity, infections, compression of a nerve root, radicular neuropathy and vasculitis.

Definitive diagnosis requires histological confirmation of the affected nerve, which, on occasions cannot be performed and is often not diagnostic since the affected zone may not be accessible to biopsy.

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Fig. 1. Sagittal T2-weighted sequence MR images of the lumbosacral spine showing multiple degenerative changes with no evidence of lymphomatous disease.
Fig. 2. 18F-FDG/PET-CT study: (a) MIP showing an increase in the 18F-FDG uptake in the left humeral head, sacrum and axillary and lumbar plexus. (b) PET, (c) CT and (d) fusion 18F-FDG/PET-CT coronal images showing lineal uptake corresponding with the left lumbar plexus suggestive of infiltration by lymphoma.

Fig. 3. Axial 18F-FDG/PET-CT images showing an increase in the 18F-FDG uptake in the right brachial plexus suggestive of lymphomatous infiltration.

MR is the imaging test presenting the best performance for the study of metastasis of the plexus and may show a slight mass adjacent to the plexus or identify metastatic infiltration. However, none of these characteristics is specific and, in some circumstances, the MR may be normal.

18F-FDG/PET-CT plays an essential role in staging, in the evaluation of response to treatment and in the detection of recurrence of lymphoma. Since the study assesses the entire whole body, it allows the restaging of the disease and the identification of tumor dissemination patterns which may be atypical. The use of hybrid images achieves precise localization (Fig. 3) and allows guidance of the biopsy to more accessible sites with greater metabolic activity in an attempt to reduce the rate of false negative results.

In our case, 18F-FDG/PET-CT confirmed the presence of tumor recurrence, demonstrating an increase in the uptake in the nervous structures which, together with the symptomatology referred, allowed the diagnostic suspicion of neurolymphomatosis and identified the site for performing the biopsy.

Conflict of interest

The authors have no conflicts of interest to declare.

References