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Images

An unusual cause of spinal cord compression

Idoia Lacoste [a], Sylvain Vergnet [b], Thomas Tourdias [c,d], Lidia Afonso Delgado [b], Gwendal Le Masson [a], Fanny Duval [a], Stéphane Mathis [a,*]

[a] Department of Neurology, Nerve-Muscle Unit, CHU Bordeaux (Pellegrin Hospital), place Amélie Raba-Léon, 33000 Bordeaux, France
[b] Department of Neurology, Movement Disorders Unit, CHU Bordeaux (Pellegrin Hospital), place Amélie Raba-Léon, 33000 Bordeaux, France
[c] Department of Neuroradiology, CHU Bordeaux (Pellegrin Hospital), place Amélie Raba-Léon, 33000 Bordeaux, France
[d] INSERM U1215, University of Bordeaux, France

1. Clinical background

A 54 year-old man complained of back pain for weeks. MRI showed bone destruction of the T2 thoracic vertebral body (without spinal cord compression nor any abnormal signal or enhancement of the spinal cord or nerve roots) and 20 bone lesions on the whole body. The diagnosis of multiple myeloma (MM) was establish: IgG lambda gammopathy (IgG peak = 40.6 g/L, normal <12 g/L); bone marrow biopsy showed 86% pleomorphic plasma cells. MM was graded ISS (International Staging System) 1, with poor prognostic factors because of the t(4;14) chromosomal aberration and the 17p13 deletion. He was treated by VTD (Velcade-Dectancyl-Thalidomide), Daratumumab, Melphalan and bone marrow transplantation. Pain progressively decreased after treatment. After six months, the serum IgG paraprotein band decreased (1.6 g/L), but he developed (within a week) gait disturbance, global weakness of lower limbs, bilateral sciatalgia and urinary/fecal dysfunctions. Deep tendon reflexes were absent in the lower limbs and weak in the upper limbs. Plantar reflex was indifferent in both sides. The electromyography revealed predominant bilateral L4-L5-S1 radiculopathy; nerve conduction studies showed mild axonal sensorimotor polyneuropathy (only affecting the lower limbs). Spine MRI showed bilateral nodular gadolinium-enhanced lesions of many lumbosacral nerve roots (Fig. 1) causing intradural compression of the conus medullaris. Brain MRI was normal.

2. What is the most likely diagnosis?

(a) CIDP (chronic inflammatory demyelinating polyneuropathy)
(b) Leptomeningeal myelomatosis
(c) Neurolymphomatosis
(d) Elsberg syndrome

3. Answer: Leptomeningeal myelomatosis

In our patient, CSF analysis showed high protein levels (500 mg/dL), normal glucose levels and 282 nucleated cells/ml (87% of atypical malignant plasma cells), consistent with the diagnosis of leptomeningeal myelomatosis (LMM). The patient died 5 months later despite radiotherapy (spine), intrathecal chemotherapy (cytarabine) and standard VAD (vincristine, doxorubicin, plus high-dose dexamethasone) chemotherapy.

Neurological symptoms are not uncommon in MM (due to hyperviscosity, hypercalcemia, spine compression, or paraprotein-related neuropathy), but direct involvement with accumulation of myeloma cells in CNS or meninges (LMM) is rare [1,2]: its incidence is estimated between 0.8 and 1.1% [3]. In LMM, contrast-enhanced MRI may reveal diffuse or focal spinal or cranial leptomeningeal contrast enhancement, as well as leptomeningeal-based mass lesions [2]. Because its sensitivity is much lower for hematological malignancies than for solid tumors, some cases of LMM with neurological involvement have been reported with normal craniospinal MRI [4]. Finally, CSF cytology examination is an effective method to detect malignant cells [4].

There is no standardized treatment for LMM, usually based on systemic treatment of MM and intrathecal chemotherapy (such as methotrexate). Treating such patients remains challenging: MM-directed therapy has only modest activity (6-month neurologic disease-free progression rate: 7%; median survival: 4 months) [3]. LMM usually occurs more frequently as a late complication of MM, its incidence increasing possibly related to modern chemotherapies altering the tumor microenvironment [5]. LMM usually presents with nonspecific multifocal symptoms or signs (cranial nerve palsy, encephalopathy or radiculopathy) [3]; consequently, the median interval from diagnosis of MM to detection of LMM is 6–18 months [4], usually associated with poor prognosis (median survival: 3 months) [2]. Finally, direct LMM spinal cord compression is rare but has to be known by neurologists.

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Competing interests statement

None.

* Corresponding author.
E-mail address: stephane.mathis@chu-bordeaux.fr (S. Mathis).

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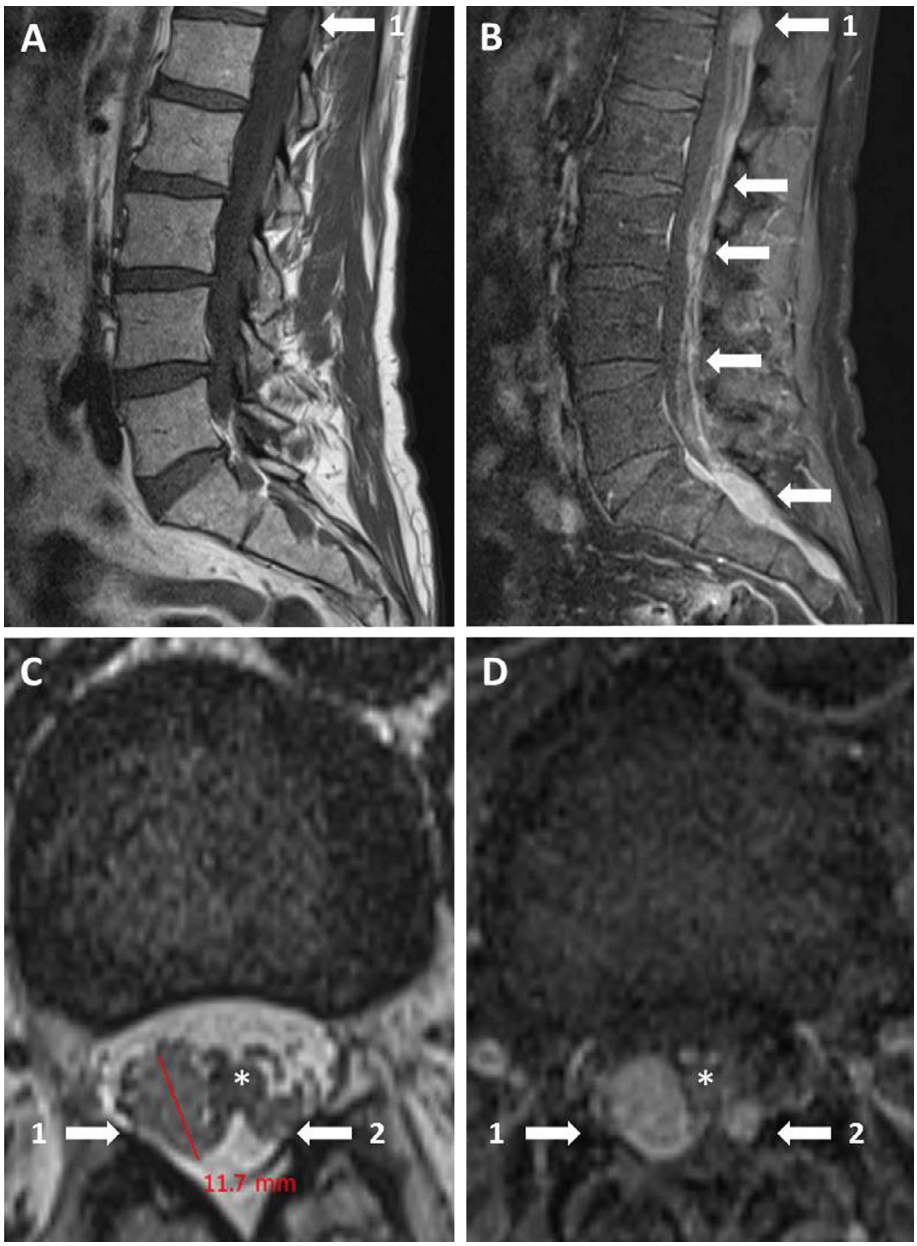

Fig. 1. Nodular extra-medullary lesions along lumbosacral nerve roots (arrows) are enhanced by gadolinium injection (A: T1-weighted MRI; B: post-contrast T1-weighted-fat saturated MRI). The largest nodular lesion (1) causes direct compression of the conus medullaris (*), with another smaller contralateral nodular lesion (2) (C: axial lumbar T2-weighted MRI; D: axial lumbar post-contrast T1-weighted-fat saturated MRI).

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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