Case 13596

Eurorad

A rare cause of acute paraplegia: granulocytic sarcoma

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Section: Neuroradiology
Area of Interest: Neuroradiology spine
Procedure: Diagnostic procedure
Imaging Technique: MR
Special Focus: Pathology Case Type: Clinical Cases
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Patient: 20 years, male

Clinical History:

A 20-year-old man, without antecedent, especially myeloid leukaemia, presented with dorsal spinal pain, asthenia in the legs, urinary and anal incontinence. Clinical examination found a flaccid paraplegia. The blood analysis showed a bicytopenia and leukocytosis consisting of 30% blasts. The bone marrow aspiration showed the presence of myeloid precursors.

Imaging Findings:

Emergency spinal MRI was performed, showing posterior epidural process extending from D4 to D10, isointense (similar signal intensity to muscle) on T1-weighted and T2-weighted images, compressing the spinal cord and associated to diffuse low signal intensity of the vertebral bone marrow (tumoral infiltration).
The patient was admitted to the operating room for a possible decompression. The surgery was limited to a D6 laminectomy with a biopsy of the posterior epidural process, the intraoperative appearance was very infiltrative, not releasable to the dura. The histological examination confirmed the diagnosis of myeloid leukaemia.
MRI performed after surgery on T1 and T2-weighted images showed a regression of compression process with stigmata of laminectomy D6.

Discussion:

Granulocytic sarcoma, also known as chloroma or extramedullary myeloblastoma, is a rare solid tumour composed of primitive precursors of the granulocytic series of WBC that include myeloblasts, promyelocytes, and myelocytes [1].This tumour was called chloroma by King in 1853 because of a greenish appearance, which is caused by the presence of myeloperoxidase granules in the malignant myeloid cells.
All organs can be affected, but most commonly the bones, lymph nodes and skin. The central nervous system is rare with an incidence of 3.25% [2]. Spinal location is exceptional with a predilection for the dorsal segment [3]. Clinically, the epidural localization can manifest as a spinal syndrome, root or sensorimotor deficits, with rapidly progressive evolution towards a spinal cord compression syndrome. This is a condition that affects the paediatric population and young adults, often male [4].
In about 1/4 of cases granulocytic sarcoma occurs outside the context of haematological pathology, however, myeloid sarcoma may precede acute myelogenous leukaemia with an average of six months. [5]
The mechanism for this location is epidural infiltration of the adventitia epidural vessels by malignant cells. This observation is also particular in its presentation, it supports the pathophysiological hypothesis that the spread to the nervous system would occur through blood.
On MRI, the tumour appears as an infiltrative epidural mass, isointense signal on T1 weighted images and

intermediate signal on T2 weighted images, homogeneous or heterogeneous, enhanced after gadolinium injection. The presence of signal abnormality of the bone marrow of the vertebrae may suggest the diagnosis [1]. The epidural lesion is often spread over 2-3 levels. In our patient the lesion affected seven levels of D4 to D10.
The histological appearance of this tumour is very variable. Immunohistochemistry is used to support the diagnosis: the anti-lysozyme is positive in 79% of cases and anti-myeloperoxidase (MPO) in 93% of cases [6, 7].
Treatment of granulocytic sarcoma is identical to that of acute myelogenous leukaemia (chemotherapy). Surgery is indicated in emergency in case of spinal cord compression. [8]
The epidural spinal location of leukaemia is very rare.
Our patient received high-dose chemotherapy (based on anthracycline, cytosine and arabinoside) with a transfusion support.
The outcome was favourable with complete haematologic and cytogenetic remission and partial neurological recovery.
Our case report illustrates the rarity of the epidural localization of granulocytic sarcoma as the main revealing symptom of acute myeloid leukaemia, and it shows the importance of an early diagnosis with appropriate treatment.
Differential Diagnosis List: Granulocytic sarcoma of the spine, Lymphoma, Leptomeningeal metastasis, Plasmocytoma, Inflammatory lesion

Final Diagnosis: Granulocytic sarcoma of the spine

References:

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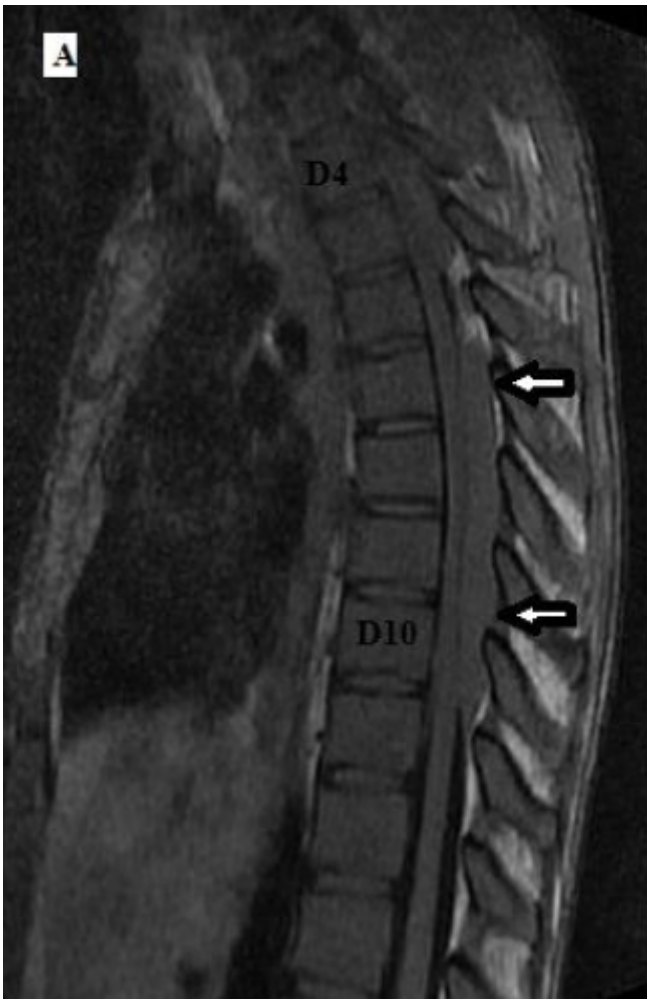
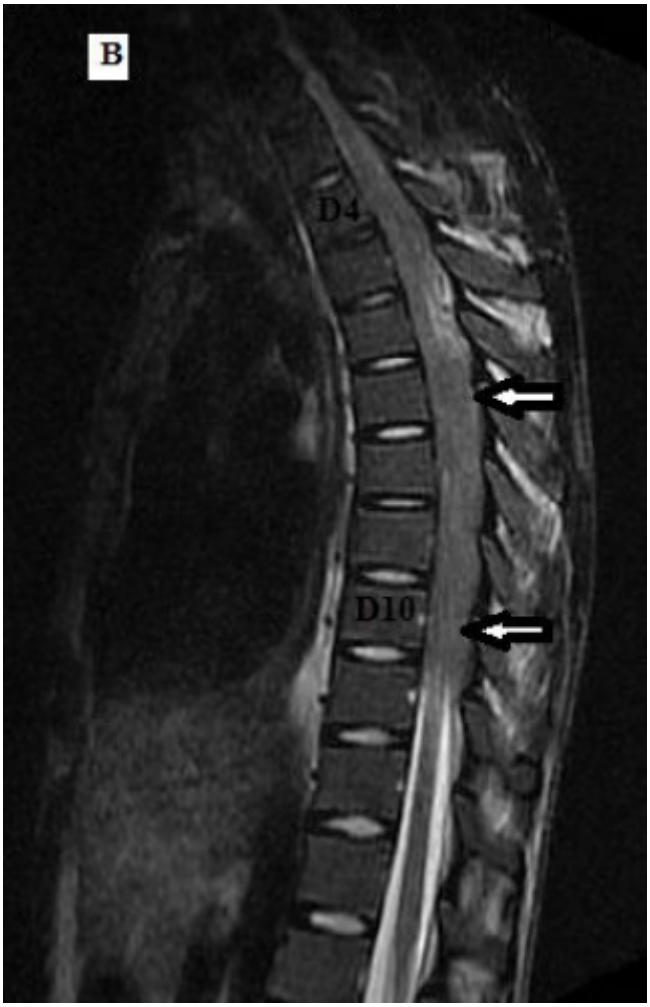
Figure 1

a



Description: T1-weighted sagittal MRI showed: Extensive epidural lesion extending from D4 to D10, isointense, compressing the spinal cord, associated to a diffuse low signal intensity of the vertebral bone marrow (tumoral infiltration). **Origin:** department of radiology; CHU Ibn Sina, Rabat

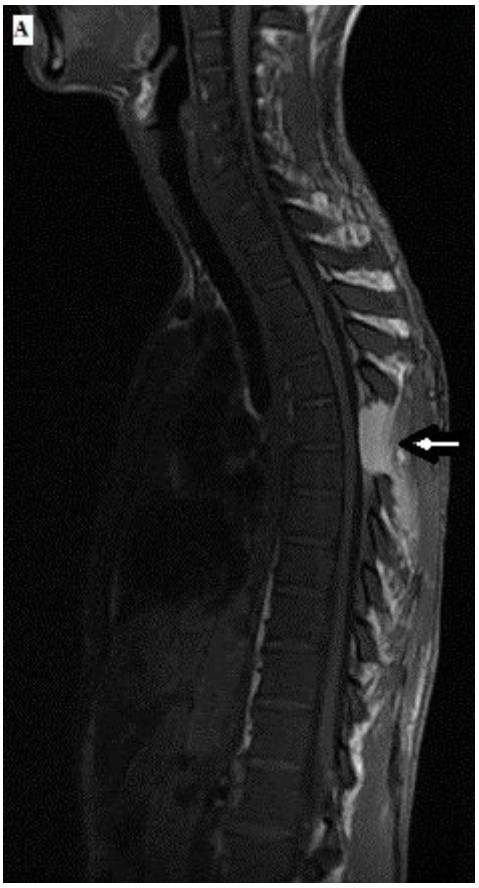
b



Description: T2-weighted sagittal MRI showed: Extensive epidural lesion extending from D4 to D10, isointense, compressing the spinal cord. **Origin:** department of radiology; CHU Ibn Sina, Rabat

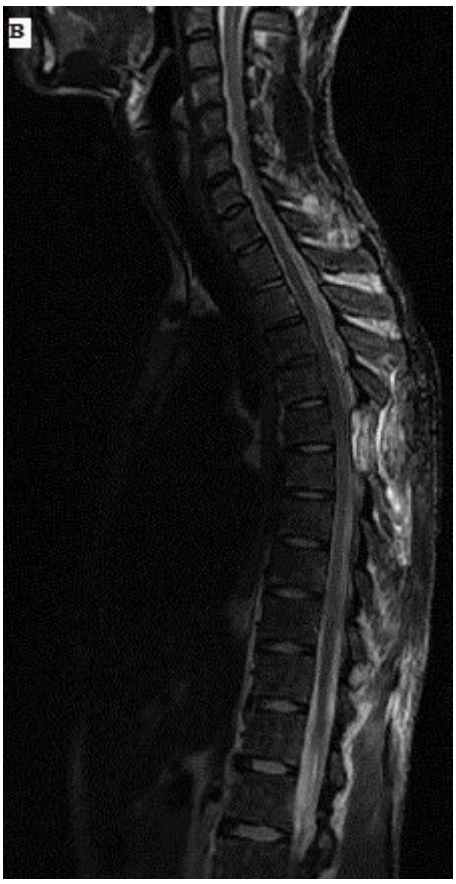
Figure 2

a



Description: Sagittal spinal cord MRI T1: complete regression of compression process (the epidural mass was completely resected) with stigmata of laminectomy D6 (arrow) **Origin:** department of radiology; CHU Ibn Sina, Rabat

b



Description: Sagittal spinal cord MRI T2: complete regression of compression process (the epidural mass was completely resected)
with stigmata of laminectomy D6. **Origin:** department of radiology; CHU Ibn Sina, Rabat