Extraosseous epidural multiple myeloma: Rare disease, no consensus, our experience about a case

Myélome multiple épidural extraosseux : Maladie rare, pas de consensus, notre expérience à propos d’un cas

H Khay*, A Abdellah, M Khoulali, N Raouzi, N Oulali, F Moufid

Abstract
Multiple myeloma is a malignant, multi-centric hemopathy. Bone involvement is common with a vertebral predilection. Extraosseous epidural localization with spinal compression is exceptional and globally, it has a poor prognosis. No consensus for the therapeutic decision has been established. To our knowledge only fifteen cases have been reported in the literature.

We report the case of a 50-year-old woman, admitted to the emergency room for T11 spinal cord compression, classified Fränkel D, the radiological assessment having shown T11/T12 epiduritis, compressive on the spinal cord. The diagnosis of Multiple Myeloma was made. The patient underwent emergency laminectomy and epiduritis removal. Additional chemotherapy was administered. Progression was favorable with complete recovery of the neurological deficit. The patient is a candidate for autologous bone marrow transplantation.

The purpose of our case report is to discuss our findings in light of those in the literature in order to contribute to the improvement of the therapeutic management and prognosis of this rare disease.

Keywords: case report, Epidural space, Multiple myeloma, Spinal cord compression.

Résumé
Le myélome multiple est une hémopathie maligne et multicentrique. L’atteinte osseuse est fréquente avec une prédilection vertébrale. La localisation épidurale extra osseuse avec compression médullaire est exceptionnelle et globalement, elle est de mauvais pronostic. Aucun consensus pour la décision thérapeutique n’a été établie. À notre connaissance Seulement quinze cas ont été rapportés dans la littérature.

Nous rapportons le cas d’une patiente de 50 ans, admise aux urgences pour une compression médullaire T11, classée Fränkel D, le bilan radiologique ayant montré une épidurite T11/T12, compressive sur le cordon médullaire. Le diagnostic de Myélome multiple est posé. La patiente a bénéficié en urgences d’une laminectomie et d’une exérèse de l’épidurite. Une chimiothérapie complémentaire a été administrée. L’évolution a été favorable avec récupération complète du déficit neurologique. La patiente est candidate à l’autogreffe de la moelle.

Notre rapport de cas a pour but, une discussion de nos résultats à la lumière de ceux de la littérature, afin de participer à l’amélioration de la prise en charge thérapeutique et du pronostic de cette maladie rare.

Mots-clés : Rapport de cas, Espace épidural, Myélome multiple, Compression médullaire.
Introduction

Multiple myeloma (MM) is a hematologic malignancy, which accounts for approximately 10% of hematologic cancers and is defined by clonal multiplication of tumor plasma cells invading the hematopoietic bone marrow and secondarily other extraosseous localizations. The diagnosis of MM is based on CRAB (Calcemia-Renal-Anemia-Bone) criteria. In addition, other clinical signs may be present, depending on the location of the tumour tissue. Spinal bone involvement is the most common. However, epidural localization is very rare. To our knowledge, the world literature counts 15 cases, our case is the 16th. We report, through one case, our diagnostic and therapeutic experience of this rare localization of MM.

Clinical case

We report, the case of a 50-year-old woman with no particular history. She was admitted to the emergency room for heaviness and paresthesias of both lower limbs, as well as back pain of progressive installation over 3 months, without vesico-sphincteric disorders. Neurological examination found spastic paraparesis allowing walking with assistance, sharp osteotendinous reflexes in the lower limbs, hyposthesia with a T11 level. General examination was normal.

The patient benefited from a spinal MRI (Magnetic Resonance Imaging) and a spinal CT scan, having shown an extradural tumour process, opposite T11-12, invading and infiltrating the post arch opposite with extension towards the neural foramens, more marked on the right. The lesion represses and compresses the medullary cone. The lesion is isointense at T1 and T2. It contrasts homogeneously and intensely. The CT scan shows minimal osteolytic involvement of the posterior arch of T11 (Figure 1). The rest of the radiological workup showed puncture lesions of the iliac bones on the pelvic radiograph. Biologically, sedimentation rate, serum calcium and creatinine were normal. The CBC (complete blood count) showed anemia at 10 g/dl normocytic normochromic. Protein electrophoresis (PEP) and serum and urinary immunofixation favoured a mono-clonal immunoglobulin (Ig)A-type gammopathy with lambda light chains (Figure 2). The myelo-gram found 32% bone marrow invasion by dystrophic plasma cells. Osteomedullary biopsy (OMB) showed 90% plasma cells.

The patient underwent emergency T11-12 laminectomy and macroscopically complete removal of an epidural hemorrhagic lesion. The histological study of the epiduritis came back in favor of MM (Figure 3). The evolution was favourable, with complete recovery of the neurological deficit after one week. The patient received four courses of VCD protocol chemotherapy (Velcade, Cyclophosphamide and Dexamethasone). The patient did not present any particular complication. At 6 months follow-up, the patient remained asymptomatic and in good general condition. The patient is a candidate for autologous transplantation.

<table>
<thead>
<tr>
<th>Author</th>
<th>Country</th>
<th>Year</th>
<th>Age</th>
<th>sex</th>
<th>MM type</th>
<th>Epidural level</th>
<th>Clinical signs</th>
<th>Treatment</th>
<th>Follow-up (months)</th>
<th>Outcome</th>
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<tr>
<td>Hino et al.</td>
<td>Japan</td>
<td>1992</td>
<td>62</td>
<td>M</td>
<td>IgD lambda</td>
<td>NA</td>
<td>Bone, Sequard syndrome</td>
<td>CT</td>
<td>14</td>
<td>NED</td>
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<td>Mawson et al.</td>
<td>Japan</td>
<td>1992</td>
<td>55</td>
<td>M</td>
<td>IgD lambda</td>
<td>T3</td>
<td>Back pain, paresthesia</td>
<td>CT</td>
<td>19</td>
<td>DOD</td>
</tr>
<tr>
<td>Lohf et al.</td>
<td>China</td>
<td>1994</td>
<td>55</td>
<td>F</td>
<td>IgD lambda</td>
<td>T4-5</td>
<td>Back pain</td>
<td>CT/surgery</td>
<td>7</td>
<td>DOD</td>
</tr>
<tr>
<td>Pabstb et al.</td>
<td>Germany</td>
<td>1996</td>
<td>60</td>
<td>F</td>
<td>IgA kappa</td>
<td>C7-L2-L3</td>
<td>Back pain, paraplegia T10,</td>
<td>RTCT/RP/</td>
<td>3</td>
<td>DOD</td>
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<tr>
<td>Watanabe et al.</td>
<td>Japan</td>
<td>2000</td>
<td>83</td>
<td>M</td>
<td>IgD lambda</td>
<td>C7-T2</td>
<td>Paraplegia T8,</td>
<td>CT</td>
<td>NA</td>
<td>NED</td>
</tr>
<tr>
<td>Hu et al.</td>
<td>Taiwan</td>
<td>2004</td>
<td>45</td>
<td>F</td>
<td>IgG kappa</td>
<td>T11-L2</td>
<td>Back pain, L5/S1 radiculopathy,</td>
<td>Surgery/CT</td>
<td>5</td>
<td>DOD</td>
</tr>
</tbody>
</table>

Table I: the literature cases of epidural extraosseous myeloma

### Discussion

MM accounts for about 1% of malignancies and 10-15% of hematological malignancies. It is defined by malignant plasma cell proliferation. The preferred localization of this disease is bone, with a multifocal character and a predilection for the spine, with a prevalence of 60% in diagnosed cases of MM. It is a discrete male-dominated disease.

According to the International Myeloma Working Group, MM is defined by the presence of clonal spinal cord plasmacytosis ≥10% and at least one of the following criteria:

- **CRAB criteria**: Calcium-Renal-Anemia-Bone
- **Hypercalcemia**: Serum calcium > 0.25 mmol/L (>1 mg/dL) above the upper limit of normal or > 2.75 mmol/L (>11 mg/dL),
- **Renal impairment**: creatinine clearance < 40 mL/min or serum creatinine > 177 mmol/L (>2 mg/dL),
- **Anemia**: Hemoglobin < 10 g/dL or > 2 g/dL below the lower limit of normal,
- **Bone lesions**: At least one osteolytic lesion present on standard skeletal X-rays, Scanner or PET

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<table>
<thead>
<tr>
<th>Authors</th>
<th>Country</th>
<th>Year</th>
<th>Gender</th>
<th>Ig Type</th>
<th>Localization</th>
<th>Symptoms</th>
<th>Treatment</th>
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<tr>
<td>Naama et al.</td>
<td>Morocco</td>
<td>2008</td>
<td>M</td>
<td>Ig G kappa</td>
<td>T4-T</td>
<td>Back pain, paraplegia T4, Vesiico-sphincter disorders</td>
<td>CT/RT</td>
<td>NA</td>
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<tr>
<td>Aziz et al.</td>
<td>USA</td>
<td>2009</td>
<td>M</td>
<td>Ig kappa (type not specified)</td>
<td>T1</td>
<td>Paraparesis T2, Vesico-sphincter disorders</td>
<td>Surgery/CT</td>
<td>NA</td>
</tr>
<tr>
<td>Avadhani et al.</td>
<td>India</td>
<td>2010</td>
<td>F</td>
<td>Ig kappa (type not specified)</td>
<td>T5-T7</td>
<td>Back pain, paraplegia T8</td>
<td>Surgery/RT/CT</td>
<td>NA</td>
</tr>
<tr>
<td>Jung et al.</td>
<td>South Korea</td>
<td>2012</td>
<td>M</td>
<td>Ig G kappa</td>
<td>T4-T6</td>
<td>Back pain, Paraparesis T8</td>
<td>Surgery/CT</td>
<td>NA</td>
</tr>
<tr>
<td>Ha et al.</td>
<td>South Korea</td>
<td>2013</td>
<td>F</td>
<td>Ig G kappa</td>
<td>C7-T2</td>
<td>Back pain, paraplegia, Vesico-sphincter disorders</td>
<td>CT/RT</td>
<td>NA</td>
</tr>
<tr>
<td>Ha et al.</td>
<td>South Korea</td>
<td>2013</td>
<td>F</td>
<td>Ig G lambda</td>
<td>C7-T2</td>
<td>Back pain</td>
<td>Surgery/CT</td>
<td>NA</td>
</tr>
<tr>
<td>Ha et al.</td>
<td>South Korea</td>
<td>2013</td>
<td>F</td>
<td>Ig G lambda</td>
<td>T3-T5</td>
<td>Back pain, paraparesis</td>
<td>Surgery/CT</td>
<td>NA</td>
</tr>
<tr>
<td>Durcal et al.</td>
<td>France</td>
<td>2014</td>
<td>F</td>
<td>Ig lambda (type not specified)</td>
<td>T9</td>
<td>Back pain, asthenia, weight loss</td>
<td>NA</td>
<td>NA</td>
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<tr>
<td>Turki M. et al.</td>
<td>Tunisia</td>
<td>2018</td>
<td>M</td>
<td>Ig D lambda</td>
<td>C4-6 &amp; T9</td>
<td>Paraparesis T12</td>
<td>Surgery/RT/CT</td>
<td>NA</td>
</tr>
<tr>
<td>Present case</td>
<td>Morocco</td>
<td>2020</td>
<td>F</td>
<td>Ig A lambda</td>
<td>T11-12</td>
<td>Paraparesis T11</td>
<td>Surgery/CT</td>
<td>6</td>
</tr>
</tbody>
</table>


*Country: the country where the cases were reported and studied.*
Three new criteria have been added since 2014:

- Spinal cord plasma cell count $\geq 60\%$,
- Free light chain ratio of serum $\geq 100$ (defined by the Binding Site test, with the affected free light chain to be $\geq 100$ mg/L),
- $> 1$ focal lesion on Magnetic Resonance Imaging (MRI) (at least 5 mm in size).

Our patient meets the diagnostic criteria for MM. The proliferation of plasma cells is first intramedullary at the bone level, then these malignant cells will cross the cortex to invade adjacent tissues. Pathologic fractures of vertebral bodies with tumor invasion in the epidural space on contact is a common complication of MM. Epidural MM can usually originate from epidural or paraspinal lymphoid tissue. Extraosseous MM is a less common form with a predilection for mucous membranes of the upper airways, gastrointestinal tract, pleura and central nervous system. Neurological impairment has been reported in less than 5% of patients. It usually occurs as a result of vertebral collapse or epidural extension from an adjacent diseased vertebra. Isolated epidural involvement without local bone disease is extremely rare and is considered a poor prognostic factor. To our knowledge, only fifteen cases of epidural myeloma without bone involvement associated with spinal cord compression have been reported in the literature, our current case is the sixteenth.

The differential diagnosis of the epidural form of MM can be made with any epidural process, including meningioma, neurinoma, lymphoma, hemorrhagic vascular process, metastases, and epidural abscess. Epidural MM is a medical-surgical emergency, requiring emergency decompression and further medical treatment of the disease. For the management of epidural MM, no clear consensus is currently established due to the limited number of cases reported in the literature. A review of the series of cases reported in the literature indicates that:

- Predominance of back injury with a rate of 80%,
- The minimal predominance of the female sex is 56.25% in cases of extraosseous epidural MM.
- The average age of patients on admission was 52.87 years with extremes of 39 and 85 years.
- More than two thirds, or 75% of cases, were admitted to the paraplegia or paraparesis stage.
- Initial improvement after decompression surgery was noted in 53% of cases.
- The prognosis is pejorative, it is that MM added the complications of spinal cord compression.

The therapeutic alternatives vary according to the authors (Table 1) and include decompression surgery, focal radiotherapy and chemotherapy. The combined treatment has been shown to give better results than chemotherapy alone. The evolution was unfavourable for the majority of patients. It is characterized either by an initial neurological deterioration or by an initial neurological improvement, usually partial, followed by a relapse. The lack of long-term follow-up data makes it impossible to draw relevant conclusions on the outcome of all patients.

In our patient’s case, a combination of Protocol VCD chemotherapy and emergency surgery resulted in a complete recovery of the neurological deficit after one week. After 6 months of treatment, the patient still remains in good general condition and is free of neurological deficit. Evolution is unpredictable in MM. Urgent diagnostic; therapeutic and complication management may explain the improved therapeutic and prognostic outcomes of some patients with epidural MM. But the overall prognosis remains poor.

**Conclusion**

MM is a malignant haemopathy, the diagnosis of which is based on clinical, biological and radiological criteria. The extraosseous epidural
localization of MM is exceptional. Therapeutic management is multimodal with best results in combined treatment. Urgent management improves the functional prognosis while the overall prognosis is poor. At present, the limited number of cases does not allow a clear consensus to be established for effective and unified management of ex-traosseous epidural MM cases. We report in this manuscript our experience and a summary of cases from the literature, in order to further clarify this disease.

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References


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