MULTIPLE MYELOMA ASSOCIATED WITH EXTRAMEDULLARY PLASMACYTOMA CAUSING NERVE ROOT COMPRESSION: A CASE REPORT

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Abstract: Multiple myeloma associated with extramedullary plasmacytoma at initial presentation is rare. We describe a 45-year-old female patient with an initial presentation of low back pain and right side L5, S1 radiculopathy. There was no evidence of vertebral involvement but an epidural tumor was found later during neurosurgical intervention. The final diagnosis was immunoglobulin G, kappa multiple myeloma complicated with spinal root compression by an extramedullary plasmacytoma. No osteolytic lesion was noted over the length of the spine. Pathology revealed high-grade plasmablastic myeloma. During the clinical course, the patient was refractory to induction chemotherapy, and there was progressive deterioration of renal function. Urinary tract infection by Morganella morganii and pulmonary infection of unknown cause developed 5 months later, and the patient died.

Case Report

A 45-year-old woman was in good health until she experienced right buttock pain radiating to the lateral aspect of the lower limb, which was associated with numbness and a tingling sensation over the distal part that progressed during a 2-month period. The pain was exacerbated by walking uphill or lying on her right side. She had no bowel or bladder complaints. She denied any history of trauma, weight loss, or fever.

She went to the orthopedic department for evaluation and treatment. Roentgenography of the lumbar spine showed mild lumbar spondylosis. Analgesics did not alleviate her pain, and she was referred to the rehabilitation department for further management.

At the initial work-up, she was found to have mild anemia (hemoglobin concentration, 96 g/L). There was no organomegaly and no lymphadenopathy. Laboratory studies revealed proteinuria and increased blood urea nitrogen (BUN; 13.5 mmol/L) and creatinine concentrations (221 μmol/L).

Neurologic examination revealed no tenderness or knocking pain over the lumbar spine. The straight leg raising test was positive on the right side at approximately 45°, and the crossed straight leg raising test was positive on the left side.

Multiple myeloma is a malignant neoplasm of plasma cells involving mainly bone and bone marrow, which can present as myelomatosis, solitary myeloma of bone, and/or extramedullary plasmacytoma [1]. It is commonly associated with marked destructive osteolytic bony lesions. Bony destruction is usually responsible for the major clinical features of multiple myeloma, which include intractable bone pain and pathologic fracture [2]. The spine is one of the most common sites of lesion development. The lower thoracic and lumbar spine are most frequently involved [3], and these patients typically manifest low back pain as the initial presentation [4].

Spinal cord or nerve root compression is the most common neurologic complication of multiple myeloma. Frequently, these patients develop marked osteolytic bony lesions in the vertebral body [5, 6].

Here, we report an unusual case of multiple myeloma that initially presented with low back pain and progressive radicular pain over the right lower leg. Later, these manifestations were found to be due to a large epidural extramedullary plasmacytoma without accompanying adjacent osteolytic bony lesions. Simultaneously, the patient was found to have multiple myeloma.
Muscle strength on right ankle dorsiflexion and big toe extension was grade 4. Sensory examination found normal results to pinprick, light touch, and position sensation on the right side. The ankle reflex was absent on the right side. Electrophysiologic examination, including nerve conduction study and electromyography, revealed right side L5, S1 radiculopathy.

Initial tentative diagnosis was lumbar disc herniation, and magnetic resonance (MR) imaging demonstrated a large epidural tumor with marked compression and anterior displacement of the thecal sac from T11 to L2 (Fig. 1). A metastatic tumor survey, including technetium-99m whole body bone scan, abdominal echo, tumor marker assay, and chest roentgenogram, was negative.

The patient underwent neurosurgical intervention with T11–L2 laminectomy to excise an extradural, grayish, friable tumor. The pathologic examination revealed plasma cell neoplasm compatible with plasmacytoma. The patient was referred to the hematology department for further management. At that time, the laboratory work-up showed hemoglobin 87 g/L, normochromic normocytic anemia, white blood cell count 6.4 x 10^9/L (with a normal differential count), and platelet count 145 x 10^9/L. The serum total protein concentration was 63 g/L; albumin concentration, 25 g/L; globulin, 37 g/L; alkaline phosphatase, 31 U/L; calcium, 2.84 mmol/L (normal, 2.2–2.6 mmol/L); uric acid, 874.3 µmol/L (normal, 150–480 µmol/L); BUN, 11.8 mmol/L (normal, 0.2–8.9 mmol/L); creatinine, 221 µmol/L (normal, 35–130 µmol/L); lactate dehydrogenase (LDH), 3.53 µkat/L (normal, 1.7–3.2 µkat/L); β2-microglobulin, 20,285 µg/L (normal, < 2,700 µg/L). The results of liver function tests and electrolyte assays were within normal limits. Serum protein electrophoresis identified a spike in the gamma fraction and the urine protein electrophoresis also identified a spike in the beta fraction. Immunoelectrophoresis of serum and urine confirmed immunoglobulin G (IgG) (kappa) monoclonal gammopathy with free monoclonal kappa light chain. The serum concentrations of immunoglobulins were as follows: IgG 17.8 g/L, IgM 0.34 g/L, IgA 0.15 g/L, and IgE 0.005 U/L. Bone marrow biopsy showed diffuse infiltration of plasmablastic cells and the normal hemopoietic component was barely seen (< 5%) (Fig. 2). The bone survey demonstrated osteolytic lesions in the femur, humerus, and pelvis. The spine was intact (Fig. 3). The final diagnosis was multiple myeloma, IgG kappa, stage IIb, associated with an extramedullary plasmacytoma over the spinal cord at the T11–L2 level.

The patient underwent two courses of chemotherapy with oncovin (0.4 mg/day continuous infusion for 4 days), epirubicin (13 mg/m^2 continuous infusion for 4 days), and decadron (15 mg IV every 6 hours for 4 days), with a 30-day interval, but no clinical response was observed. Her general condition and renal function deteriorated progressively. She died 5 months later, when she had pulmonary infection with diffuse alveolar infiltration and urinary tract infection by Morganella morganii in spite of antibiotic treatment.

**Discussion**

Extradural plasmacytoma is a rare clinical entity. It may be primary, or part of systemic involvement of multiple myeloma [7, 8].

Spinal cord or nerve root compression occurs in about 10% to 20% of myeloma patients [9]. The pathogenesis is vertebral collapse or infiltration of myeloma cells into the extradural space [5, 6]. Cord compression occurs most often at the thoracic level, while root compression usually involves the lumbosacral nerve roots [10]. Typically, plain radiographs and CT scans demonstrate a significant osteolytic lesion or collapse of the vertebral body in the corresponding segment [5]. Plain radiography is the most suitable tool to screen for bony lesions in multiple myeloma, but MR imaging has been suggested for screening of spinal lesions in patients with normal radiographs [3]. There have been few reports of solitary, extramedullary, and extradural plasmacytoma without spreading from the surrounding spine bony lesions, and all reported cases presented with spinal cord compression [11–14]. In the present case, MR imaging scan revealed a large extradural epidural tumor mass from the T11 to the L2 level and no evidence of vertebral bone involvement was noted over the length of the spine. However,

![Fig. 1. Extramedullary plasmacytoma manifesting as an epidural mass from the bony level of T11 to L2 (arrow). The mass (A) is isointense to muscle on this sagittal T1-weighted image.](image-url)
the patient presented with root compression at the L5, S1 level rather than cord compression, as in most previously reported cases.

According to the histologic classification of multiple myeloma by Bartl et al, the high-grade plasmablastic form has a uniformly poor prognosis with a median survival of 10 months [15]. It has been reported that this subtype is usually associated with lower hemoglobin and serum albumin concentrations, higher calcium and β₂-microglobulin concentrations, and a higher percentage of bone marrow plasma cells. This subtype also has more frequent renal insufficiency and a higher plasma cell labeling index. Extramedullary lesions develop more frequently in this form, which may contribute to the shorter survival [16, 17]. The prognosis is more favorable in primary extramedullary plasmacytoma, which occurs mainly in the upper aerodigestive tract [8]. However, when extramedullary plasmacytoma manifests as part of systemic involvement of multiple myeloma, it is usually associated with an anaplastic pathology and a more aggressive clinical course [18, 19]. These observations could explain why our patient responded poorly to treatment and deteriorated progressively.

We conclude that multiple myeloma is one of the most common causes of spinal cord and nerve root compression. 

Fig. 2. The bone marrow is diffusely infiltrated by plasmablastic cells with round vesicular nuclei and apparent nucleoli. The normal hematopoietic component is hardly seen. (H & E) A) x 100, B) x 400.

Fig. 3. Plain x-ray of the lumbar spine shows no osteolytic or osteoblastic abnormality.
compression [20]. When a patient presents with low back pain, the possibility of multiple myeloma must be evaluated. If there is no evidence of bony destruction or collapse of vertebral bodies on the routine bone survey, extraosseous epidural plasmacytoma should be added to the differential list, although it is rare. In most cases, MR imaging study is necessary to confirm these extramedullary lesions. Decompressive surgery or radiotherapy in conjunction with systemic chemotherapy for the underlying multiple myeloma is currently considered the standard treatment [21].

References