Thoracic Cord Compression by epidural Multiple Myeloma: A Rare Presentation of Multiple Myeloma

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Abstract
Multiple myeloma is a hematopoietic disorder and multicentric disease, with the most common localization being the spine. A 53-year-old male presented with progressive paraplegia, superficial and deep sensory disturbance below the level of T2. Spinal magnetic resonance image showed an epidural mass compressing the spinal cord at the level of T1 with intact bone structure. The patient underwent surgical posterior spinal decompression. Microscopic examination and immuno-histochemical studies confirmed the diagnosis of multiple myeloma of kappa subtype. The patient was subsequently started on steroids and chemotherapy for myeloma. Extra osseous epidural tumors causing compression myelopathy without evidence of destruction or collapse of vertebral bodies are relatively rare; to our knowledge very few cases exist in the literature.

INTRODUCTION
Multiple myeloma is a B-cell malignancy characterized by accumulation of monoclonal plasma cells. It accounts for approximately 1% of all malignant diseases and represents about 10% of hematologic malignancies. Typically, the disease involves the bone marrow and breaks through the cortex, invading the surrounding tissue. Usually multiple myeloma is a widespread and multicentric disease, with the most common localization being the spine. Therefore, collapse of vertebral bodies with narrowing of the spinal canal and circumscribed tumor extension into the adjacent epidural space is a common complication of the disease. Spinal cord compression occurs in approximately 5% of patients with multiple myeloma. We report an unusual case of multiple myeloma that showed spinal compression syndrome due to space-occupying epidural mass without focal vertebral involvement.

CASE REPORT
A 53 old African American male with no significant past medical history came with the complaint of weakness in bilateral lower extremities form 2 days. Patient started feeling mild weakness and numbness in both the legs. Initially the weakness was not hindering his daily activities. Weakness gradually progressed to an extent that he was not able to bare weight on his legs. He also started having bladder and bowel incontinence on the day of presentation, which made him to seek medical assistance. Before this episode of weakness, he was having generalized body pains for about one month. He denied any trauma to the back, fever, headache, nausea, vomiting or diarrhea or any difficulty in breathing or loss of appetite or weight. On initial examination Patient had a power of 4/5 in both the lower extremities and decreased sensations in both legs extending up to below the umbilicus. Reflexes were normal. Rectal examination showed decreased anal sphincter tone. Patient was immediately started on corticosteroids for suspected card compression. In mean while, examination of the patient revealed further deceased power in the bilateral lower extremities to 3/5. Neurosurgery was called immediately. Initial labs were normal including the liver and renal parameters. The MRI showed cord compression at the level of T1. There was 3 cm infiltrative mass at the level of T1 extending into the spinal canal causing the compression of the cord.
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Patient was immediately taken to the surgery and debulking of the mass was done and sent for pathology study. Post-operative MRI showed decompression of the cord. The biopsy of the mass showed plasma cell malignancy. Immuno-histochemical analysis of the bone marrow showed neoplastic cells of plasma cell origin. These plasma cells expressed monoclonal lambda light chain with absent kappa staining. Post-operative course was uneventful and patient regained the power of 4/5 in both lower legs. Radiotherapy was started. Slowly patient regained the strength and was able to ambulate with walker.

Figure 1
Fig 1. Infiltrative mass at level of T1 vertebrae.

Figure 2
Fig 2. Spinal cord compression by the infiltrative mass.

Figure 3
Fig 3. Post operative MRI showing patent spinal canal.

Figure 4
Fig 4. Low power view showing multiple plasma cells.
DISCUSSION

Multiple myeloma is a malignant proliferation of plasma cells usually showing diffuse bony involvement with predilection of the spine. A frequent complication of multiple myeloma is pathologic fractures. Bony involvement typically is lytic nature. Extramedullary multiple myeloma is very rare, comprising less than 5% of all plasma cell neoplasms. Spinal cord compression is usually caused by primary involvement of the vertebral body with tumor extension into the adjacent spinal canal. In these cases, plain radiographs or CT generally show a large, lytic bone lesion or collapse of the vertebral body in the corresponding segment. MRI has been established as a sensitive method in detecting localized or diffuse bone lesions in multiple myeloma. In our case, no reliable evidence of bone involvement was seen on conventional radiographs, and the vertebral bodies were of normal height and shape. On MRI, the marrow signal in the vertebral bodies was normal; however, MRI demonstrated spinal cord compression over T4 level due to a separate extensive epidural mass in the dorsal spinal canal, which was clearly distinguishable from surrounding bone structures and obviously of extra osseous origin.

The case reported by Matsui et al. occurred in a 52-year-old man who presented with paraparesis due to immunoglobulin D (IgD) myeloma mass that occupied the lumbar epidural space separated from the vertebral body. The patient died from an epidural tumor in the high thoracic region 19 months after the onset of symptoms.

Palmbach et al. reported a case of multiple myeloma in a 75-year-old woman with progressive paraplegia due to a large epidural and extraosseous manifestation without evidence of lytic lesion or collapse of vertebral bodies. Only MRI showed multiple patchy bone lesions. The patient died 3 months after admission as a result of an extensive pneumonia and respiratory insufficiency.

Watanabe et al. reported a case of isolated epidural mass IgD multiple myeloma with intact bone structure at the level of C7 to T2 in an 85-year-old man who presented with compression myelopathy.

To the best of our knowledge, the present report is the sixth to describe an isolated epidural mass of multiple myeloma without evidence of destruction or collapse of vertebral bodies. In these cases, we believe that spinal involvement may be caused by extension of Paraspinal lymph node into the inter-vertebral foramen. Others suggested that the tumors could originate from lymphoid tissue present in the epidural space.

Treatment options for extra osseous epidural multiple myeloma include surgical resection and local radiation therapy (4000cGy over 4 weeks). The response to conventional chemotherapy of extramedullary multiple myeloma is very poor.

The median survival of patients with myeloma is
approximately 2.5 years, although this rate appears improved to approximately 4.5 years in the younger patients. With spinal involvement, 75% of patients die within 1 year of diagnosis while almost all die within 4 years.\cite{7,8}

Although quite rare, extra osseous multiple myeloma without evidence of bone destruction or collapse of vertebral body should be included in the differential diagnosis of epidural mass lesions, causing spinal cord compression.

References
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