Primary Plasma Cell Leukemia Presented as Progressive Paraplegia: A Case Report

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Abstract: Plasma cell leukemia (PCL) is a rare plasma cell disorder. It is the leukemic variant of multiple myeloma. A 52-year-old man with an atypical presentation of primary plasma cell leukemia is reported. The patient presented with paraparesis which progressively worsened to paraplegia. MRI of the spine showed an extradural mass causing cord compression and multiple bony erosions from soft tissue masses. Peripheral blood film examination and bone marrow aspiration showed numerous plasmablasts. Atypical cells expressed surface and cytoplasmic lambda light chain on immunochemical studies, surface CD45 and CD38. To our knowledge, primary PCL presenting with progressive paraplegia has not been reported in the literature.

Key Words: cord compression, multiple myeloma, paraplegia, plasma cell leukemia

P lasma cell leukemia (PCL) is a rare variant of multiple myeloma (MM) occurring in 2–4% of all cases of MM.\(^1\,^2\) It is a malignant proliferation of plasma cells in the peripheral blood and bone marrow. The diagnosis is made when there are more than 20% of plasma cells in the peripheral blood with an absolute plasma cell count of more than \(2 \times 10^9/L\).\(^1\,^2\) PCL can be classified as primary when it presents in the leukemic phase or as secondary when there is leukemic transformation of a previously recognized multiple myeloma.\(^3\) In this report, we highlight an unusual case of primary PCL in a 52-year-old man who presented with progressive paraplegia secondary to an extradural mass and multiple soft tissue masses.

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Accepted May 15, 2008.

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Southern Medical Journal • Volume 102, Number 1, January 2009

Case Report

A 52-year-old man first presented at a district hospital with a three-week history of progressive bilateral lower limb weakness. It was associated with peripheral numbness and bowel and urinary incontinence. He had no known medical illnesses prior to presentation. He also complained of loss of appetite and weight. He denied any fever, cough, night sweats or contact with any pulmonary tuberculosis patient and he had no history of trauma. He was a heavy smoker for 30 years and smoked about 30 cigarettes per day. There was no family history of malignancy or leukemia. He was treated for two weeks before being transferred to Hospital Universiti Kebangsaan Malaysia, Kuala Lumpur.

On examination, he was dehydrated and ill-looking. There was a left orbital swelling as well as soft tissue swelling at the left rib and scapular area measuring 3 cm and 12 cm, respectively. There were bilateral crepitations over the chest. There was no hepatomegaly or splenomegaly. Examination of the upper limbs showed normal bilateral power and reflexes while the lower limbs showed total paraplegia and absent reflexes. There was a loss of sensation at the level of T4 and below. The anal tone was lax with loss of perianal sensation.

Blood investigations showed hemoglobin of 8.9 g/dL, white cell count \(30.2 \times 10^9/L\) (neutrophils \(21.0 \times 10^9/L\), lymphocytes \(4.6 \times 10^9/L\), monocytes \(4.2 \times 10^9/L\)), platelets \(180 \times 10^9/L\), sodium \(138 \text{ mmol}/L\), potassium \(4.8 \text{ mmol}/L\), urea \(23.1 \text{ mmol}/L\), creatinine \(351 \text{ mmol}/L\), total protein \(71 \text{ g}/L\), albumin \(30 \text{ g}/L\), calcium \(2.71 \text{ mmol}/L\), phosphate 1.23

Key Points

- Primary plasma cell leukemia presenting with cord compression is an unusual presentation of the disease.
- Despite its poor prognosis, combination chemotherapy has been shown to improve patients’ condition.
mmol/L, erythrocyte sedimentation rate 66 mm/h, C-reactive protein 3.38 mg/dL and lactate dehydrogenase of 1598 IU/L. Sputum culture grew *Pseudomonas aeruginosa* but the blood culture was negative. Chest x-ray showed bilateral haziness over the mid- and lower zone bilaterally. There were multiple erosions over the 6th to 8th ribs. Thoracolumbar x-ray showed an old fracture of L2. Full skeletal survey showed no lytic lesions.

An urgent magnetic resonance imaging of the spine (Fig. 1) showed an extradural mass involving the left side of T4 and T5 vertebrae and the right side of the T7 vertebra, causing narrowing of the spinal canal and the right exiting nerve root. There was also a prevertebral mass noted which extended from T6 to T7. Computed tomography (CT) scan of the thorax and abdomen (Fig. 2) showed a huge mass at the right and left side of the 6th and 7th ribs respectively. CT scan of the brain and orbit showed infiltration of the orbital region with no brain parenchymal involvement.

An initial diagnosis was made of paraplegia secondary to spinal cord compression secondary to an extradural mass, most probably metastatic disease, with septicemia secondary to bronchopneumonia. However, tumor markers were negative. Peripheral blood film showed the presence of numerous plasmablasts (69%). Bone marrow smear showed the presence of plasma cells (20%) and its variant as well as plasmablasts (21%). Immunohistochemical studies showed the cells strongly expressed CD38 and lambda chain which was consistent with plasma cell leukemia. B-cell markers were negative. Cytogenetic studies showed a complex chromosomal abnormality with a few hyperdiploidy. A CT-guided biopsy (Fig. 3) was performed of the mass over the left 7th rib which showed infiltration by plasmacytoid cells of various levels of maturity. Immunohistochemical studies showed restriction for lambda light chain and were positive for CD79a, but negative for CD20, CK and HMB45. These features were consistent with a plasma cell neoplasm. A final diagnosis of primary plasma cell leukemia with multiple bony lesions and soft tissue infiltrations, complicated by paraplegia secondary to cord compression, was made.

The patient was given one course of combination chemotherapy (vincristine, doxorubicin and dexamethasone) once his septicemia had resolved. After the first course of chemotherapy, the multiple soft tissue swellings reduced in size with some improvement of his paraplegia. He was due for a monthly course of chemotherapy. Unfortunately, he presented with septicemic shock three weeks after the chemotherapy and died two months after the diagnosis.

Discussion

Most clinical data of PCL were collected from case reports and a few review articles. The clinical presentation of primary PCL is more aggressive than that of multiple myeloma. A higher presenting tumor burden, and higher frequencies of extramedullary involvement, anemia, thrombocytopenia, hypercalcemia, and renal impairment have been reported. Interestingly, in primary PCL,
the presence of lytic bone lesions seems to be lower than those usually observed in MM. Primary PCL commonly presented with aesthenia, renal insufficiency, bone pain, hepatosplenomegaly, and lymphadenopathy, while secondary PCL patients were older and had a greater incidence of lytic bone lesions and lower platelet counts. However, the involvement of the central nervous system causing paraplegia in primary PCL has not, to our knowledge, been reported.

The prognosis for both primary and secondary PCL is poor, although there are better response rates to treatment in patients with primary PCL. The prognosis for primary PCL with spinal cord compression remains unknown. Spinal cord compression in multiple myeloma showed an excellent response to combination treatment of laminectomy and radiotherapy. A third of patients with complete paraplegia and about 40% of patients with a history of weakness for 3 weeks to 3 months responded to treatment.

At present, the optimal treatment for PCL is not well defined. Nevertheless, higher response rates were seen with combined chemotherapy compared to a single agent or the standard myeloma regimen. Autologous stem cells have been reported to produce excellent responses, with up to a 2- to 3-year disease-free survival rate. However, death related to sepsis remains the main cause of death.

References

Fig. 3 Biopsy of the mass over the left 7th rib showing infiltration by plasmacytoid cells of various maturity.