Unusual lower back pain with monocytosis: A case report

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Received April 14, 2015; Accepted July 12, 2016

DOI: 10.3892/ol.2016.5190

Abstract. There are numerous causes of lower back pain. In the oncological setting, spine metastasis from a solid tumor is the most common. However, hematological disorders should also be taken into consideration. The current study presents a case of chronic myelomonocytic leukemia with the initial presentation of chronic lower back pain, followed by symptoms that included urinary retention, stool incontinence and left gum swelling, in a patient who was eventually diagnosed with granulocytic sarcoma (GS) over the sacral region. GS is a rare presentation of a tumor consisting of extramedullary leukemic infiltrations, which develop at different sites and cause different symptoms. Prompt and correct diagnosis of this type of disease may be crucial to improve the survival outcome by the early initiation of adequate treatment.

Introduction

Granulocytic sarcoma (GS) is a rare form of tumor consisting of extramedullary leukemic infiltrations. GS can develop at various sites, including the skin, orbital region, central nervous system (CNS), lymph nodes and spleen (1). GS may present at an initial leukemia diagnosis, during disease progression or relapse, or after hematopoietic stem cell transplantation (SCT) (2). The symptoms of GS depend on its size and location, and include tenderness or abnormal bleeding. The incidence of GS in acute myeloid leukemia (AML) is 2.5–9.1% (3), but it is less frequent in chronic myelogenous leukemia and other myeloproliferative diseases.

Chronic myelomonocytic leukemia (CMMoL) is a subtype of myelodysplastic/myeloproliferative neoplasm (4). The clinical presentation of CMMoL is often non-specific, including fatigue, dyspnea, petechiae, hemorrhage, skin lesions or splenomegaly. Diagnostic criteria include monocytosis of >1,000 cells/mm³ for >3 months, exclusion of all possible infectious etiologies and no specific gene rearrangements or mutations (5).

Cauda equina syndrome is associated with compression of the lumbosacral nerve roots below the level of the conus medullaris, which presents as tenderness and sphincter dysfunction (6). The current study presents a rare case of lower back pain and monocytosis as the initial presentation. Radiographic and immunohistochemical studies confirmed a diagnosis of CMMoL with granulocytic sarcoma located at the sacral region.

Case report

A 62 year-old woman presented with a 1-year history of mild lower back pain with soreness radiating to the left leg. The patient had previously taken over-the-counter analgesics and had intermittently undergone physical therapy. However, the tenderness persisted and then worsened, with the development of weakness of the left lower leg on February 28, 2015. Other pertinent symptoms included urinary retention, stool incontinence and left gum swelling, prompting referral to the Emergency Department of the Tri-Service General Hospital (Taipei, Taiwan). Upon physical examination, the patient exhibited swelling and tenderness of the left posterior mandibular region. A neurological examination showed hypoesthesia over the L3-5 and S1 dermatomes, along with a loose anal sphincter and unsteady gait. Bilateral muscle power was grade 4 (7), and the Babinski sign was absent. A peripheral blood smear revealed monocytosis, anemia and thrombocytopenia: White blood cell count, 80,030/µl; hemoglobin, 10.7 g/dl; platelets, 32,000/µl; monocytes, 38.2%; and metamyelocytes, 2.9%. An enlarged spleen was palpable 5 cm below the left costal margin, but no pulmonary or breast masses were found. Spine magnetic resonance imaging showed several heterogeneously contrast-enhancing masses up to 5.6 cm in diameter, located at the S1 to S3 vertebrae. Multiple enhancing nodules were identified in the spinal canals from L4 to the caudal end (Fig. 1). The patient was immediately admitted with a diagnosis of cauda equina syndrome. A bone marrow biopsy and spine biopsy were performed. The bone marrow was hypercellular and demonstrated dysplastic megakaryocytes with hyposegmentation, left-shifted myeloid hyperplasia and an increased myeloid:erythroid ratio (>10/1). Cluster of differentiation (CD)34 (mouse anti-human CD34 class II monoclonal antibody; 1:100 dilution; clone, QBEnd 10; Dako, Glostrup, Denmark), was positive in ~15% of cells, and there was also increased numbers of CD68-positive cells (mouse

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Key words: back pain, granulocytic sarcoma, chronic myelomonocytic leukemia
anti-human CD68 monoclonal antibody; 1:100 dilution; clone, PG-M1; Dako). The two pathology reports indicated CMMoL with spine involvement (Fig. 2A-D). Cytogenetic studies were normal (46XX). Flow cytometry revealed 18% of monoblasts lacking the expression of cluster of differentiation (CD)4, CD34 and CD117, and with increased expression of CD56. The patient was therefore diagnosed with CMMoL, intermediate-2, according to the CMMoL-specific prognostic scoring system (8), and treated with low-dose cytosine arabinoside (10 mg/m² every 12 h) for 10 days. Palliative radiotherapy was also arranged, but was not performed as the condition of the patient deteriorated. The patient succumbed to severe sepsis half a month later.

Discussion

In patients who complain of lower back pain, a thorough survey of all possible underlying systemic diseases, with an evaluation of neurological signs, should be undertaken, particularly in elderly patients. A total of 5-14% of cancer patients develop spinal metastasis with cord compression (9). Lung, breast, prostate, renal and gastrointestinal tumors are all prone to progress to or present with spinal metastasis (10). A detailed workup should be performed to search for the primary etiology. In addition to solid tumors, metastatic spinal lesions of hematological origin should be taken into consideration, particularly in patients with an abnormal hemogram.

The incidence and prevalence of CMMoL are not high, and the exact frequencies are unknown (11). Granulocytic sarcoma is more common in acute leukemia and less common in chronic leukemia (1). Leukemia cutis is the most common type of GS that presents in CMMoL, accounting for 10.2% of cases during disease progression (12). Certain cases transform to AML rapidly. The remaining sites of involvement are the pericardium, jejunum and testis, respectively (13-16). Extramedullary involvement of the CNS is rare, and only one pediatric patient has been reported following SCT (17). This juvenile CMMoL patient experienced bone marrow relapse 8 months after GS formation in the CNS. According to the aforementioned findings, it is known that the presence of GS in CMMoL patients indicates a dismal outcome and a possible sign of blastic crisis. The treatment for CMMoL varies depending on the severity of the disease status and the specifics of the presentation. Patients with a lower blast count (<5%) may initially require only supportive care if their symptoms are mild. However, conventional chemotherapy, hypomethylating agents or allogeneic
SCT may be required for those with bone marrow blast counts of >10% or for those who exhibit other signs of advanced or aggressive disease. The treatment response of hypomethylating agents is 25-70% (5). Patients may also require >4 cycles of therapy to reach a clinical response, which is similar to myelodysplastic syndrome (18).

The present study reported a case with an unusual presentation of back pain that was finally diagnosed as a hematological malignancy. In conclusion, GS should be taken into consideration for patients with a spinal mass and abnormal hemogram. The presence of GS in CMMoL predicts a poor outcome. A prompt biopsy is important to ensure the correct diagnosis and to guide the appropriate treatment, thus preventing rapid progression of the disease.

Acknowledgements

The authors would like to thank Mr. Chian-Ming Chen, Miss Hsin-Yi Liu and Dr Wen-Chuan Tsai from the Tri-Service General Hospital for their expertise in analyzing the patient's bone marrow smear, spinal biopsy specimen and flow cytometry results. The authors would also like to thank Dr Anthony Janckila (Department of Microbiology and Immunology, University of Louisville, Louisville, KY, USA) for providing a critical review of the manuscript.

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