Chronic myelogenous leukemia presenting with Morel Lavallee lesion – case report

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Abstract

Chronic myelogenous leukemia (CML) is a myeloproliferative neoplasm defined by dysregulated and uncontrolled proliferation of mature and maturing granulocytes with uniform distribution. It is associated with the fusion of BCR (on chromosome 22) and ABL1 (on chromosome 9) resulting in a BCR: ABL1 fusion gene. This fusion gene produces BCR: ABL1 protein which causes the constitutive activation of Tyrosine kinase, implicated in the pathogenesis of chronic myelogenous leukemia. The Morel Lavallee lesion is a closed soft tissue degloving injury resulting from the separation of the hypodermis from the underlying fascia, which in turn leads to hematoma collection between the layers. It is a rare manifestation of chronic myelogenous leukemia (CML). In this report, we describe a 50-year-old female who presented with right abdominal swelling, associated with easy fatigability, and significant weight loss of 1 month. Later confirmed to be chronic myelogenous leukemia with bone marrow aspiration and BCR/ABL1 fusion mRNA by RT-PCR.

Keywords: chronic myelogenous leukemia, myeloproliferative neoplasm, Morel Lavallee, BCR/ABL1

Introduction

Chronic myelogenous leukemia is a myeloproliferative disorder characterized by uncontrolled production of granulocytes, mainly neutrophils, and myeloid expansion of the bone marrow. There are three phases of chronic myelogenous leukemia, the chronic phase, accelerated phase, and blast crises phase. Although it can

directly evolve from the chronic phase to the blast phase, Usually, the disease progresses from the chronic phase to the blast crises phase.

The clinical manifestation of CML is variable depending on the stage of the disease at diagnosis.20-50 % of patients are asymptotic at the time of diagnosis and 85 % of patients are diagnosed in the chronic phase(1, 2).

Systemic symptoms such as fatigue (34 %), malaise (3 %), weight loss (20 %), drenched sweating (15 %), abdominal fullness (15 %), and bleeding episodes due to platelet dysfunction (21%) are the commonest manifestations (2). CML can rarely present with soft tissue hematoma which can be treated with both tyrosine kinase inhibitors and conservative management(3, 4). The Morel Lavallee lesion is a close spontaneous soft tissue injury resulting in hematoma collection between fascial layers. The diagnosis of CML is suspected by the typical findings on the peripheral blood and bone marrow, then confirmed by the presence of the Philadelphia chromosome, BCR/ABL1 fusion gene, or the BCR/ABL1 fusion mRNA by conventional cytogenetics, fluorescent in situ hybridization (FISH) or reverse transcription polymerase chain reaction (RT-PCR) respectively.

Hydroxyurea can be used for cytoreduction while awaiting confirmation of the diagnosis. Tyrosine kinase inhibitors are the backbone of management. We report a case of Morel Lavallee lesion in the anterior abdominal wall as an initial presentation of chronic myelogenous leukemia.

Case presentation

A 50-year-old female presented with progressively increasing upper abdominal swelling of one-month duration. She also has easy fatigability, unquantified but significant weight loss, and loss of appetite

The patient has no history of trauma or heavy lifting. She also denied any previous episodes of bleeding and her family history was also negative for bleeding disorders.

Physical examination was significant for splenomegaly of 8 cm along the splenic growth line, a 20x10 cm hard, non-tender mass on the right side of the umbilicus (figure 1), and ascites. Her laboratory evaluation reveals a complete blood count with a WBC of 97,900, hemoglobin of 9.6, and platelet count of 919,000. Abdominal ultrasound found a right anterior thoracoabdominal mass and splenomegaly measuring 20cm.

Abdominopelvic contrast CT was significant for a right abdominal wall (intramuscular) huge hematoma collection (figure 2) and massive splenomegaly. Lactate dehydrogenase was elevated to 949 IU/L.

Renal function tests, liver enzymes, coagulation profile, and serum electrolytes were in the normal range.

Bone marrow examination showed myeloid hyperplasia (with myeloid to erythroid ratio of 15:1). Neutrophils and bands 68%, metamyelocytes 6%, promyelocytes 8%, blasts 3% suggestive of CML-chronic phase. The diagnosis of CML was confirmed after RT-PCR was done on the patient's blood and was positive for BCR/ABL1.

Ultrasound-guided fine needle aspiration from the mass revealed inflammatory cells with a hemorrhagic background.

Treatment and outcome

The patient was started on hydroxyurea, allopurinol, and hydration. Imatinib 400mg p.o/day was begun after cytoreduction.

She received conservative care for the hematoma accumulation and was discharged with appointments to the surgery and hematology clinics. In two weeks, she has significantly improved.

Conclusion

It is important to keep in mind that unexplained soft tissue hematoma can be an initial manifestation of chronic myelogenous leukemia. However, the exact cause of the Morel Lavallee lesion is not yet known, platelet dysfunction, acquired Von Willebrand disease, and acquired Glanzmann's thrombasthenia are the most likely causes. supportive care and tyrosine kinase inhibitors are the main mode of management. Surgical interventions can be considered based on indications.

Discussion

CML is a malignant neoplasm, which is characterized by the expansion of the erythroid, myeloid, and platelets in the peripheral blood and myeloid hyperplasia of the bone marrow. It is associated with the BCR/ABL1 fusion gene. This gene encodes an abnormal protein which leads to the constitutive activation of tyrosine kinase, which eventually results in neoplastic proliferation(5). CML accounts for 15-20% of leukemias in adults(6) and Ionization radiation exposure is the only known risk factor(1, 7).

The average diagnostic age ranges from 60 to 65 years(8). About 50% of cases are asymptomatic and are identified incidentally by routine complete blood count tests(2). Anemia, splenomegaly, fatigue, weight loss, and night sweats are the commonest initial manifestations(5). Extramedullary involvement such as the lymph nodes, skin, and soft tissues can be seen in patients with blast crises(9). very rarely, soft tissue hematoma can be the first manifestation of CML. The soft tissue hematoma may be explained by platelet dysfunction, acquired Von Willebrand disease, or acquired Glanzmann's thrombasthenia(3).

The clonal expansion of dysfunctional megakaryocytes can explain platelet dysfunction in CML, which explains the anticipated improvement in their function following the initiation of tyrosine kinase inhibitors(3, 10-12).

The Morel-Lavallée lesion is a closed traumatic soft-tissue degloving injury, first described by the French physician, Dr. Victor-Auguste-Francois Morel-Lavallee in 1863. This damage is the result of the separation of the hypodermis from the fascial tissue due to a shearing force. Hematoma collection forms due to disruption of the soft tissue's perforating blood and lymphatic vessels(3, 13). The subcutaneous tissues beside the greater trochanter are the most frequently affected areas by the Morel-Lavallee lesion(13). We were able to find three case reports in which the Morel Lavallee lesion was the initial manifestation of CML. The first patient was a 16-year-old male athlete who presented with swelling on his knee, confirmed to be CML after investigation, and was managed both surgically and with a tyrosine kinase inhibitor(14).

The second patient was a 55-year-old male who came with left thigh swelling and was also diagnosed with CML after workup and treated with imatinib and conservatively(4). The third case was a Morel Lavallee lesion affecting the posterior chest wall, he was treated conservatively and started on dasatinib(3). Our case is the fourth CML patient presenting with Morel Lavallee lesion, and the first case that involved the anterior abdominal wall. She was managed conservatively.

platelet dysfunction, acquired Glanzmann's thrombasthenia, acquired Von Willebrand deficiency, were attributed to the spontaneous hematoma in the previous case reports(10-12).

Even though it's not clear why this patient developed hematoma in unusual site without any prior trauma, platelet dysfunction is the likely etiology.

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Lists of abbreviation

BCR/ABL1-break point cluster region/ Abelson tyrosine-protein kinase 1

CML-chronic myelogenous leukemia

CT-computed tomography

FISH-fluorescent insitu hybridization

mRNA-messenger ribonucleic acid

P.O-per os.

RT-PCR-reverse transcription polymerase chain reaction

WBC-white blood cell

Declarations

Ethics approval and consent to participate

There are no ethical concerns about this case report.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Availability of data and materials

All of data and materials for this case report are available from corresponding author.

Competing interests

We declare that there are no competing interests.

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Authors' contributions

Dr. Gebeyehu Tessema Azibte -writing, reviewing and editing

Dr. Getnet Yigzaw Mossie-source, writing original draft

Dr. Zekarias Seifu Ayalew-reviewing and editing

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Not applicable

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