Priapism in chronic myeloid leukemia: A rare case report

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SUMMARY

Chronic Myeloid Leukemia (CML) is a clonal myeloproliferative neoplasm characterized by the BCR::ABL1 fusion gene and accounts for about 15% of adult leukemia cases. Its global prevalence has increased due to improved survival with tyrosine kinase inhibitor (TKI) therapy. Priapism is a rare urological emergency in CML, usually associated with severe hyperleukocytosis. This report describes a 32-year-old man presenting with generalized body pain, spontaneous bruising, and a painless penile erection lasting more than four days without sexual stimulation. Examination revealed splenomegaly (Schuffner 4), while laboratory tests showed extreme leukocytosis (420×10^9/L), anemia, and thrombocytopenia. The BCR::ABL1 (b3a2) transcript was detected by RT-PCR, confirming CML. Treatment included leukapheresis, hydroxyurea, imatinib, and a Winter procedure due to persistent priapism. Bone marrow biopsy confirmed CML in the chronic phase. Although leukocytosis and spleen size decreased, the patient developed erectile dysfunction, likely due to irreversible ischemic injury. This case highlights priapism as a rare but serious initial manifestation of CML caused by leukostasis, emphasizing the need for prompt cytoreduction and early urologic intervention.

INTRODUCTION

Chronic myeloid leukemia (CML) is a myeloproliferative neoplasm with an annual incidence of two cases/100 000. It accounts for approximately 15% of newly diagnosed adult leukemia cases.[1] In 2024, an estimated 9280 new CML cases will be diagnosed in the United States (US), and about 1280 patients will die of CML (due to its high prevalence today). Since the introduction of imatinib in 2000, the annual mortality in CML has decreased from 10%–20% to 1%–2%.[2] The CML-specific mortality is 0.5%–1%. Consequently, the prevalence of CML in the US, estimated at about 30 000 cases in 2000, has increased by approximately 9000/year to an estimated 150 000+ cases in 2024. Early estimates indicated the CML preva- lence to reach a plateau of about 180 000 cases by 2030–2040.[3]

Priapism is a urological emergency. Ischemic priapism is the most common type, with one of the causes being a hematologic disorder in the form of leukemia.[4] The incidence of priapism is 1.5 per 100,000 people. About 20% of cases of priapism are caused by hematological disorders. In patients with leukemia, 50% of cases of priapism are due to CML.[5]

However, based on the current incidence of near 9000 cases/year in the US (population expansion) and an estimated annual overall mortal- ity of 1%–2%, the prevalence plateau (annual incidence equal to annual mortality of 9000 cases) is now estimated to be 9000 100/2 = 400–450 000 cases in the US, which may not be reached until 2040–2050 with full TKI optimal treatment. Considering a world population of 8 billion and optimal CML management worldwide with the availability of affordable generic BCR::ABL1 tyrosine kinase inhibi- tors (TKIs), the world prevalence of CML (25 times that of the US) might reach above 10 million cases. These projections depend on difficult-to-estimate variables like the population growth in the US and worldwide, and the TKIs treatment penetration, optimization, and affordability.[6]

CASE PRESENTATION

A 32-year-old man was referred from the regional hospital with chief complaints of body aches, spontaneous bruising, and lumps. These complaints were accompanied by significant weight loss, from over 100 kg to 77 kg in 4 months. The patient also complained of frequent, unexplained bruising. There was no history of previous illnesses or drug allergies. The patient's father had a family history of chronic kidney failure. 3 days beforee, the patient experienced prolonged erections without sexual stimulation—a condition that suggested priapism. Laboratory tests revealed a white blood cell (WBC) count of 454,970 /mm^3, hemoglobin of 7.3 g/dL, hematocrit of 22.5%, and a platelet count of 178,000 /mm^3. An abdominal ultrasound revealed splenomegaly with a spleen size of Schuffner 4. Based on the clinical and laboratory findings, the patient was referred to the internal medicine department for further evaluation.

Initial treatment began with hydroxyurea 2×1000 mg per day, imatinib 2×400 mg per day, and calcium carbonate. The treatment plan included a Winter procedure to treat priapism and leukapheresis to reduce the leukocyte count. As part of the initial chemotherapy regimen, the patient was also given cytarabine 2×20 mg subcutaneous leukapheresis and a 600 ml packed red blood cell transfusion. A central double lumen (CDL) was inserted for leukapheresis.

After leukapharesis, the leukocyte count decreased from 420 to 385 x10^9/L, and the spleen size also gradually decreased to Schuffner 2, then reached Schuffner 0 in 2 weeks. Furthermore, after Hb 10, a Winter procedure was finally

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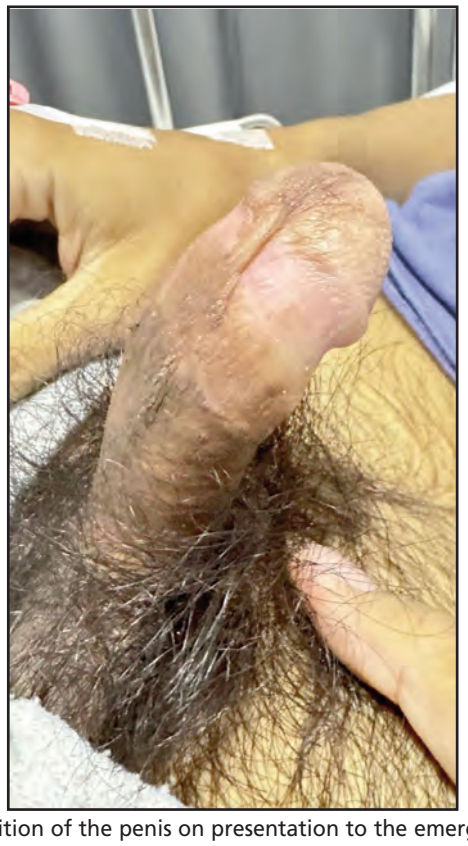

Fig. 1: Initial condition of the penis on presentation to the emergency department

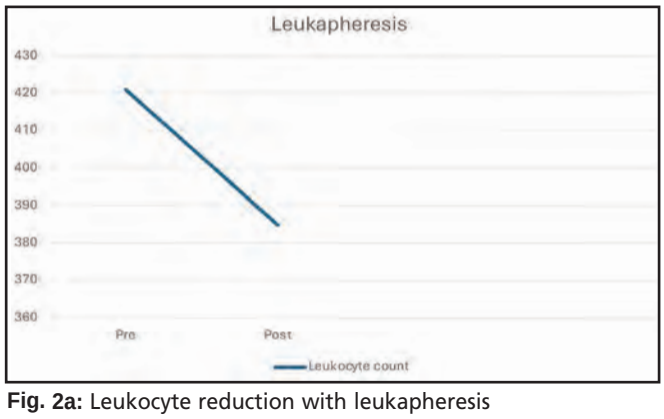

Fig. 2a: Leukocyte reduction with leukapheresis

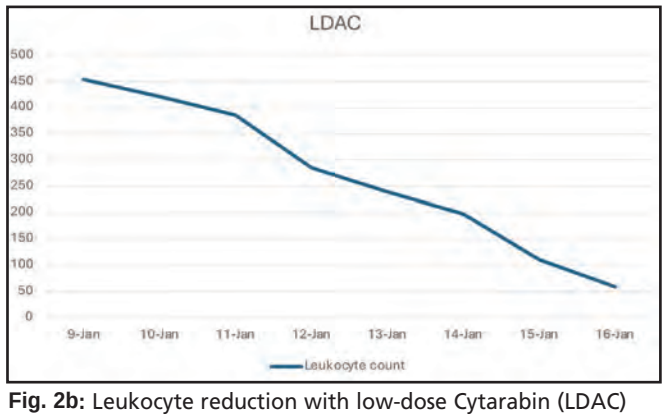

Fig. 2b: Leukocyte reduction with low-dose Cytarabin (LDAC)

performed and the hydroxyurea dose was increased to 3 x 1000 mg per day. In the following week, the patient entered the aplasia phase, with leukocytes decreasing to 1,650/µL, so imatinib and hydroxyurea therapy were temporarily discontinued.

A month later, imatinib therapy was resumed at a dose of 400 mg per day, and hydroxyurea was administered twice daily at 500 mg. Molecular testing revealed a positive BCR-ABL result with a b3a2 fusion, confirming the diagnosis of CML. Priapism persisted until the patient reported erectile dysfunction in the second month. At the end of the second month, a bone marrow puncture (BMP) was performed, the results of which supported the definitive diagnosis of CML.
During follow-up, the patient's overall condition steadily improved as his underlying disease was brought under control. The episodes of priapism resolved completely without recurrence, and no further urologic complications were observed. Despite the previous prolonged erections, erectile function was fully preserved, and the patient maintained normal sexual function. His clinical course remained stable, with continued improvement in hematologic parameters, and he experienced a significant enhancement in quality of life. Overall, the priapism was successfully managed, the condition resolved, and the patient's erectile function remained intact, reflecting a positive long-term outcome.

Bone marrow aspiration/biopsy (H&E stain) showing marked hypercellularity with predominant myeloid lineage proliferation, reduced fat spaces, relative suppression of erythroid precursors, and increased megakaryocytes — features consistent with Chronic Myeloid Leukemia (CML).

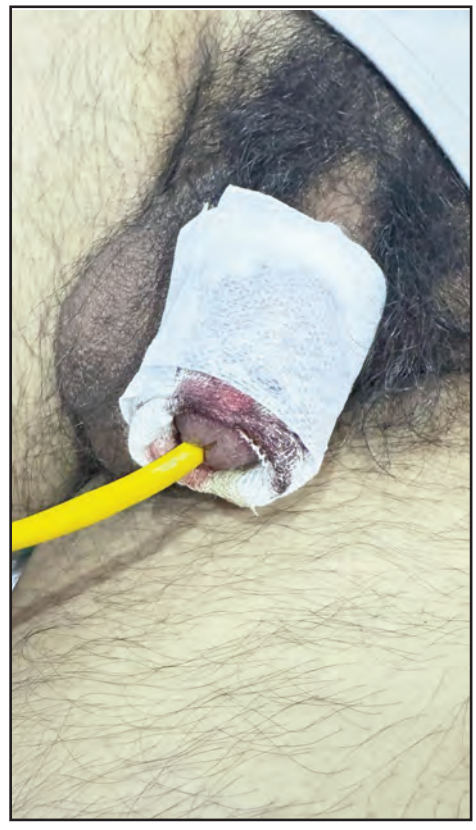

Fig. 3: Postoperative penile condition following the Winter procedure

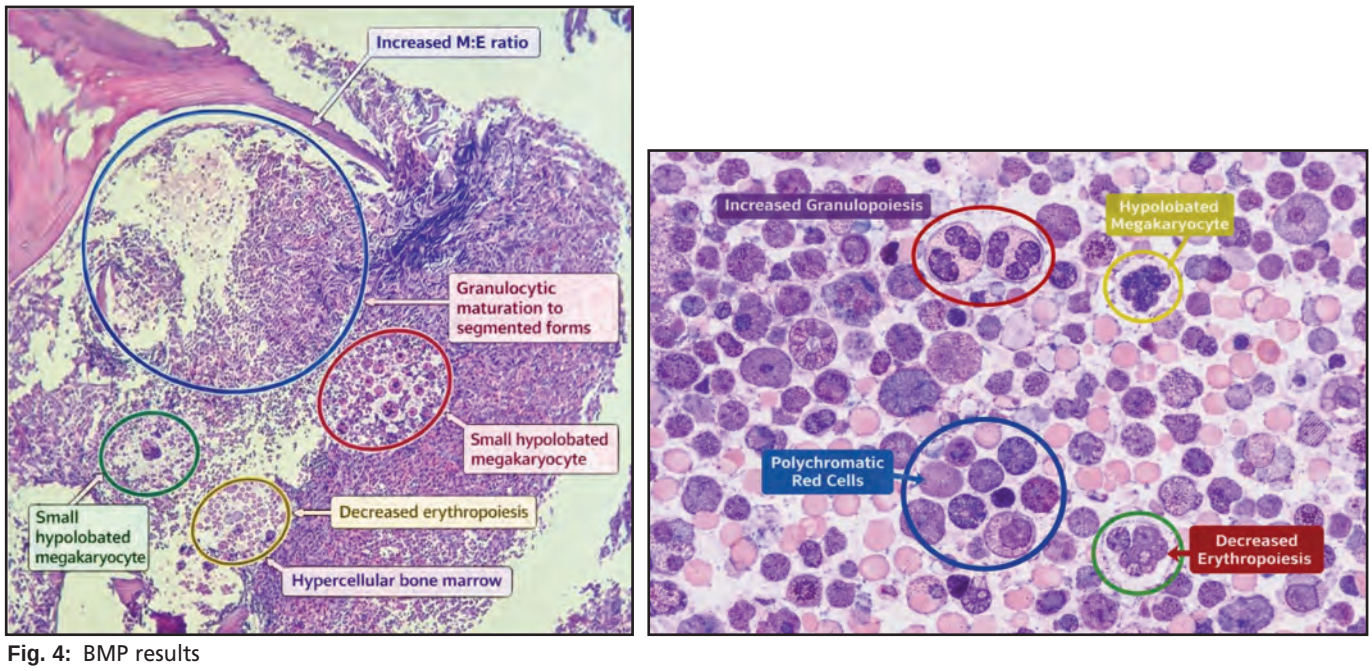

Fig. 4: BMP results

The bone marrow section demonstrates marked hypercellularity due to excessive proliferation of myeloid precursors, with cells present at all stages of granulocytic maturation (myelocytes, metamyelocytes, band forms). There is a myeloid predominance with a left shift, while erythroid elements are relatively decreased, resulting in an increased myeloid-to-erythroid (M:E) ratio. Megakaryocytes may be increased and often appear small or hypolobated, a characteristic finding in CML. These marrow features correlate with CML, a myeloproliferative neoplasm driven by the BCR-ABL fusion gene (Philadelphia chromosome), leading to uncontrolled proliferation of the myeloid lineage.

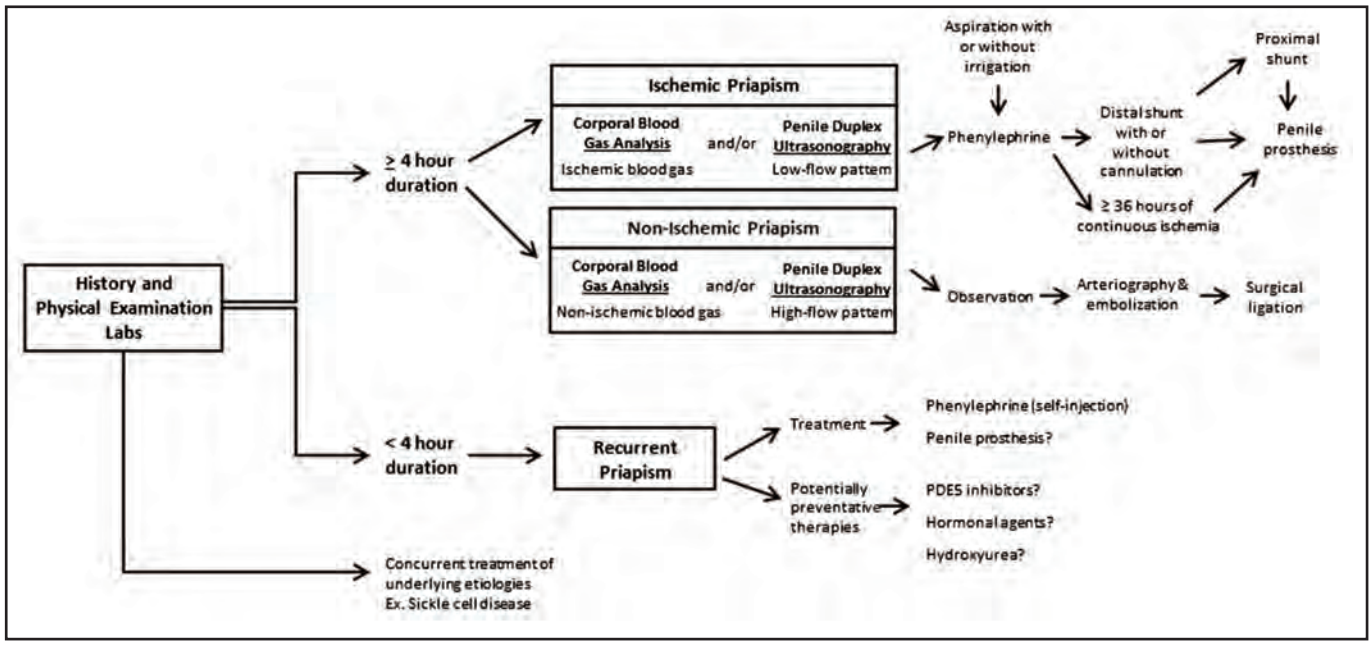

Fig. 5: Algorithm of priapism management[11]

DISCUSSION

CML characterized by the proliferation of abnormal and immature myeloid cells in the bone marrow, disrupting the production of normal blood cells. CML is a myeloproliferative neoplasm characterized by the presence of the Philadelphia chromosome (Ph+), resulting from a reciprocal translocation between chromosomes 9 and 22, resulting in the BCR::ABL1 fusion gene encoding a constitutively active tyrosine kinase, leading to uncontrolled cell proliferation and evading apoptosis.[7] Initial examination of the patient revealed hyperleukocytosis, anemia, and thrombocytopenia, as well as splenomegaly up to Schuffner 4. Extreme hyperleukocytosis in CML can lead to complications such as leukostasis, an emergency condition caused by the aggregation of leukemic cells in the circulation that disrupts blood flow to vital organs, including the penis, as occurred in this patient who experienced ischemic priapism without sexual stimulation. Priapism is a rare but characteristic complication of CML, occurring due to obstruction of the microcirculation by leukemic cells that causes ischemia of the penile cavernous tissue.[5] Severe splenomegaly, in this case reaching Schuffner 4, reflects the overwork of the spleen in destroying abnormal blood cells, as well as the extramedullary site of hematopoiesis due to bone marrow failure.[3]

Molecular testing identified the **BCR::ABL1** fusion gene with the **b3a2** transcript, producing the p210 protein and confirming the diagnosis of **CML**. CML is diagnosed based on morphological, cytogenetic, and molecular findings, with detection of the Philadelphia chromosome or **BCR::ABL1** by RT-PCR being essential and serving as a therapeutic target. Bone marrow examination showed myeloid hyperplasia, consistent with **chronic-phase CML**, characterized by <10% blasts and generally good treatment responsiveness.[7] Patient therapy begins with hydroxyurea to rapidly control the leukocyte count. Hydroxyurea is a cytotoxic agent that inhibits ribonucleotide reductase, inhibiting DNA synthesis, and reducing the leukocyte count in emergencies such as leukostasis.[3] Leukapharesis is performed to mechanically reduce the extreme leukocyte count (420 x 10^9/L), which is essential to reduce the risk of complications such as stroke, visual impairment, or subsequent priapism. Definitive therapy begins with imatinib mesylate, a first-generation tyrosine kinase inhibitor, which specifically inhibits BCR::ABL1 activity by occupying the ATP-binding site on the protein. Imatinib's effectiveness in altering the natural history of CML is significant, increasing 10-year survival from approximately 20% to 85–90%.[6,8]

After several weeks of therapy, the spleen size decreased significantly and priapism symptoms began to resolve, although by the end of February the patient was reported to have lost erections, indicating permanent cavernous tissue damage due to prolonged ischemia. Treatment of priapism involves urologic interventions such as the Winter procedure, which involves aspiration of blood from the corpus cavernosum to relieve pressure and restore circulation. Rapid intervention is crucial, as ischemic priapism lasting more than 4–6 hours can lead to tissue fibrosis and permanent erectile dysfunction.[4]

On January 23, the patient entered the aplasia phase, marked by a drastic decrease in leukocyte count to 1,650/µL, so imatinib and hydroxyurea therapy were temporarily discontinued. Aplasia can occur in response to cytotoxic therapy that severely suppresses the bone marrow, so close monitoring and transfusion support are necessary during this phase. After improvement, imatinib therapy was resumed at a dose of 400 mg/day, the standard first-line regimen for patients with chronic CML. Imatinib has a good safety profile and a low resistance rate of approximately 10% over 10 years, and its effectiveness in inducing major molecular remissions makes it a global mainstay therapy for chronic CML.[9]

CML is one of the hematological malignancies with the best prognosis since the advent of targeted therapy using tyrosine kinase inhibitors (TKIs). With adherence to therapy and regular molecular monitoring to ensure hematological, cytogenetic, and molecular response, most patients can live near-normal lives. However, long-term monitoring remains crucial, as some patients can progress to the accelerated or blastic phase, although the incidence is only around 5–6% within 10 years.[8,10] The key to successful therapy lies in early diagnosis, appropriate therapy selection, patient adherence to long-term treatment and regular monitoring. Until the end of the last century, drug therapy for CML was limited to nonspecific agents such as busulfan, hydroxyurea, and interferon- alpha (IFN-a). IFN-a therapy resulted in suppression of the Ph-positive cells and improved survival but had modest efficacy and sig- nificant toxicities. Allogeneic hematopoietic stem cell transplantation (allo-HSCT) is curative but carries the risks of morbidities and mortality. Allo-HSCT is an option for younger patients with good performance status and organ functions, and who have an appropriate donor.

The CML therapeutic landscape changed dramatically with the development of the small molecule BCR::ABL1 TKIs that potently interfered with the interaction between the BCR::ABL1 oncoprotein and adenosine triphosphate (ATP), blocking cellular proliferation of the malignant clone.[9] This "targeted" approach altered the natural history of CML, improving the 10-year survival rate from approxi- mately 20% to 80%–90% over 20 years of follow-up.[10] While the 10-year survival rate is about 85%, many patients now die from causes unrelated to CML (old age, second cancers, and others). With imatinib as frontline therapy, the 10-year CML-specific survival rate is 90%, the 10-year rate of CML resistance is only 10%, the CML-specific mortality rate is 10%, and the 10-year incidence of blastic transformation only is 5%–6%.[8]

Imatinib mesylate was the first TKI to receive approval by the United States Food and Drug Administration (FDA) for the treatment of patients with CML-CP. It acts via competitive inhibition at the ATP-binding site of the BCR::ABL1 oncoprotein, which results in the inhibition of phosphorylation of proteins involved in cell signal transduction. It efficiently inhibits the BCR::ABL1 kinase and also blocks the platelet-derived growth factor receptor and the C-KIT tyrosine kinase.[7]

CONCLUSION

This case underscores that ischemic priapism, while uncommon, represents a potentially serious and urgent complication of chronic myeloid leukemia, typically arising from extreme hyperleukocytosis and leukostasis. The accumulation of leukemic cells can obstruct microcirculation, including within the penile cavernous tissue, leading to prolonged, painful erections without sexual stimulation. If not addressed promptly, this can result in tissue ischemia and permanent erectile dysfunction. Early recognition and rapid intervention with cytoreductive measures, such as leukapheresis and hydroxyurea, are therefore critical to relieve obstruction, restore normal blood flow, and prevent irreversible damage. Furthermore, definitive urologic management, when indicated, can safely resolve the priapism once hematologic parameters are stabilized. Beyond the acute management, effective long-term control of CML through targeted therapy with tyrosine kinase inhibitors like imatinib is essential, as these agents specifically inhibit the BCR::ABL1 oncoprotein, induce hematologic and molecular remission, and reduce the risk of disease progression and mortality. Patient adherence, close monitoring of therapeutic response, and achievement of major molecular remission are key determinants of durable outcomes. Importantly, this case highlights the value of a multidisciplinary approach, involving hematology, urology, and emergency care teams, to optimize both functional and disease-related outcomes. Overall, it demonstrates that with timely recognition, coordinated care, and targeted therapy, even rare and potentially devastating complications like ischemic priapism can be successfully managed, with complete resolution and preservation of erectile function.

CONFLICT OF INTEREST

The authors declared that they have no conflict of interest.

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