Case Report

Priapism in a Pediatric Chronic Myeloid Leukaemia Patient: Unusual Presentation of a Rare Disease in Children

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Abstract

A rare case of chronic myeloid leukemia (CML) presenting with priapism is reported in a pediatric patient. CML accounts for around 15% of hematological malignancies. It is still rarer in the pediatric age group. Investigations revealed hyperleukocytosis due to CML. When conservative management failed, priapism was relieved by shunt procedures and the patient was treated with oral chemotherapy. This case demonstrates the importance of early identification of the underlying cause of priapism, and that the possibility of CML may be kept in mind in pediatric patients too, as it directly influences the management and early diagnosis may preserve sexual function in later life.

Keywords: *Chronic myeloid leukemia, pediatric patient, priapism, sexual function*

Introduction

An abnormal, prolonged, painful erection of the penis persisting for longer than 4 h which occurs without accompanied sexual arousal may be termed as priapism. These cases are a medical challenge where the underlying cause is to be diagnosed and treated.[1] Leukemia accounts for 0.7% cases of malignant priapism.[2] Of this, 50% of cases are attributed to chronic myeloid leukemia (CML). Priapism in adult leukemic patients is about 1%–5%.[3] Priapism in children due to CML has been rarely seen. Treatment includes conservative management and treatment of the underlying disease.[1] We report here an interesting case of CML in a child who presented with priapism.

Case Report

A 15-year-old boy presented with the progressive painful erection of the penis for 2 days. It was not associated with any sexual stimulation, intake of medicines, or trauma. No similar history of such painful erection in the past was reported. He reported intermittent mild episodes of epistaxis and weight loss of approximately 10 kg in 4 months. On examination, he had mild pallor, a massive spleen which was palpable up to the umbilicus. The penis was erect, firm, swollen, and tender with prominent superficial veins. Hemoglobin was 9 g/dL, hematocrit was 28%, white blood count was 135,000/mm^3, and platelets were 197,000/mm^3. A peripheral blood smear [Figures 1 and 2] demonstrated immature leukocytes in various stages of differentiation (myeloblast 3%, myelocyte 18%, metamyelocyte 19%, neutrophils 50%, lymphocytes, 6%, eosinophils 3%, and monocytes 1%), mild normocytic, normochromic anemia. About 75% of the BCR/ABL fusion genes were detected in 111 informative interphase cells analyzed [Figure 3]. Bone marrow analysis was suggestive of CML. Color Doppler ultrasonography suggested a low-flow type of priapism. Renal and liver function tests were within the normal limits. HIV status was nonreactive using ELISA method.

An attempt was made to manage the patient by cavernosal aspiration, irrigation, and phenylepinephrine injection, but the penis was persistently erect. Hence, the distal shunt procedure – glanocorposal shunt – was done. This relieved the priapism minimally, but the pain did not subside. The shunt procedure was revised after few hours when proximal corporospongiosal shunt was done. Partial detumescence was achieved with some relief of pain. At the same time, the patient was supported with intravenous fluid hydration, analgesics, and moist oxygen inhalation. After complete workup and establishing the diagnosis

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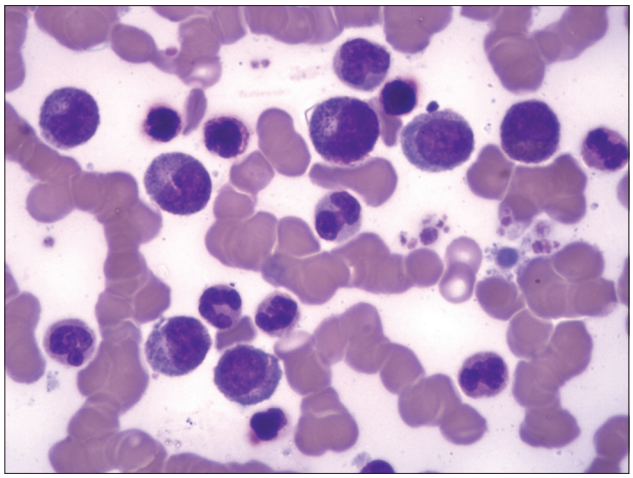

Figure 1: Peripheral blood smear showing increased granulocytes including immature forms (×100)

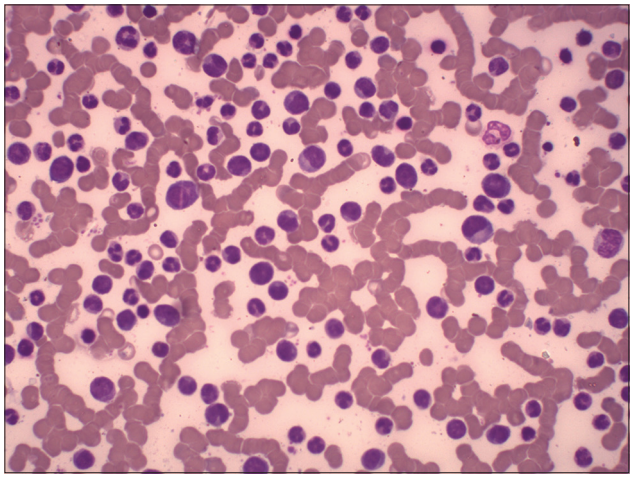

Figure 2: Peripheral blood smear showing marked increased granulocytes including immature forms (×40)

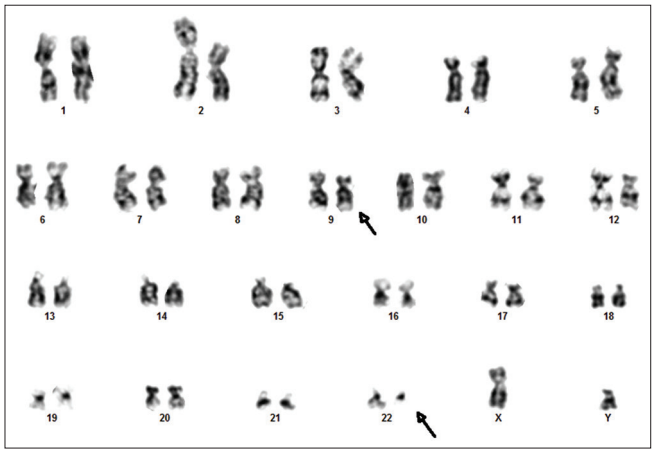

Figure 3: Genetic testing, abnormal male karyotype of 46 chromosomes including translocation (9;22)

of CML, the patient was started on tablet imatinib (400 mg/day) and tablet allopurinol (300 mg/day) along with symptomatic treatment. After 2 days, priapism reduced to normal flaccid state.

Blood counts were repeated after 1 month. Splenomegaly regressed gradually and came to normal size after 3 months of imatinib therapy. At present, the patient is on imatinib tablet (400 mg/day), with regular blood investigations every month. By combining urologic and oncologic treatment, the patient recovered completely and there was no future episode in 2-year follow-up. Furthermore, the remarkable hematologic response was noted with the latest total leukocyte count of 7000/mm^3 with no immature granulocytes and normal platelet count.

Discussion

Priapism, a rare entity, is a medical emergency. It is extremely important that prompt diagnosis of the underlying systemic disease should be made and early effective treatment should be administered to avoid long-term complications of erectile dysfunction and impotence. It is frequently seen in adults and rarely in children. In our literature review analysis, only four cases of the pediatric age group have been reported from 2006 to 2016.[3-6] However, pediatric priapism may be under-reported.

The American Urology Society divides priapism into two subtypes, namely, ischemic (low-flow) priapism or nonischemic (high flow) priapism.[7] Ischemic priapism is a nonsexual, persistent erection characterized by little or no cavernous blood flow and abnormal cavernous blood gases (hypoxic, hypercarbic, and acidotic). It requires prompt management as irreversible cellular damage and fibrosis occurs if not relieved within 48 h.[8] It may lead to erectile dysfunction in later life and future episodes of persistent and prolonged priapism (stuttering priapism).

The other less common type is nonischemic priapism. This is neither painful nor fully rigid, persistent erection. This results in increased arterial flow that overwhelms venous outflow. Irreversible damage or fibrosis is rare. Trauma to the penis or perineum is the main cause of nonischemic priapism.

The common causes of priapism in children are sickle cell disease (65%), leukemia (10%), trauma (10%), idiopathic (10%), and pharmacologically induced (5%). CML is responsible for up to 50% of leukemic cases of priapism. Increased number of circulating leukocytes in mature and immature forms may cause hyperviscosity leading to venous obstruction from thrombi, causing priapism in hematologic malignancy. Many studies have shown that a white blood cell count greater than 100.0 × 10^9/L is likely to cause blood hyperviscosity.

A detailed history and thorough physical examination, gas analysis of the blood within the corpora cavernosa, and penile Doppler ultrasound study can classify the type and indicate the etiology of priapism.

Multidisciplinary treatment with a medical oncologist, surgeon, urologist, radiologist, nursing, and psychology provide the best chances of cure. The initial goal of

therapy is immediate relief of priapism. At presentation, the patient is treated conservatively. Penile anesthesia or systemic analgesia is the first-line treatment to control pain and decrease patient's distress. Sometimes, local nerve block or sedatives are necessary to control pain. Surgery is reserved only if conservative treatment fails. Surgical procedures include aspiration-irrigation, intracorporeal sympathomimetics, or shunt procedures.

Management of the underlying cause is the mainstay of treatment. Imatinib, a tyrosine-kinase inhibitor (TKI), is the initial drug of choice for the majority of patients with CML. It inhibits cellular proliferation and produces a 92%–98% decrease in CML colony growth *in vitro*.[9] Imatinib was the first TKI licensed for use in CML patients in chronic phase , and its introduction was associated with significant improvements in response and survival. Imatinib at a dose of 400 mg/day is the gold standard first-line treatment for CML.[10]

Newer drugs, such as dasatinib and nilotinib, are also being used nowadays. Drugs under clinical trials include bosutinib, ponatinib, and saracatinib.

Earlier series of case reports show successful detumescence with local radiation therapy, open surgical shunting, or combination of the two treatments.[9] Recent literature suggests the use of cytoreductive modalities such as chemotherapy or combination chemotherapy and leukapheresis.[10]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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