CASE REPORT

Chronic myeloid leukaemia presenting as priapism: A case report from Khyber Pakhtunkhwa

Adnan Khan,[1] Ihtesham Shafiq,[2] Mohammad Hassan Shah,[3] Sarbiland Khan,[4] Gulrukh Shahid,[5] Maria Arabdin[6]

Abstract

Priapism is known as a very rare complication of leukaemia. We report a 16-years boy who presented with the persistent painful erection of the penis for eleven days. He had aspiration without achieving sustained detumescence. The patient underwent an emergency irrigation and decompression of priapism by a consultant urologist. During the hospital stay blood morphology, bone marrow aspiration and BCR-ABL were done to confirm the diagnosis of chronic myeloid leukaemia. Our report thus explains the relevance of all physicians in the diagnosis and management of patients with priapism.

Keywords: Priapism, Leukaemia, CML.

Introduction

Priapism is characterized by painful, prolonged and irreducible erection not leading to ejaculation. It is a medical emergency with significantly grim prognosis because of the risk of impotence which is almost 50% despite appropriate management. Sickle cell anaemia, chronic myelogenous leukaemia, chronic lymphocytic leukaemia, and acute lymphoblastic leukaemia are some of the haematologic disorders that can result in priapism.[1] In adult leukaemic patients, the incidence of priapism is known to be approximately 5%[2] and in paediatric it is even rarer. Priapism mainly occurs due to blood disorders, in which veins are blocked as a result of small emboli and thickening of blood because of increased number of circulating leukocytes in mature and immature forms.[3] The most accepted mechanism behind the development of painful and persistent erection is sludging of blood in the corpora cavernosa. Permanent fibrosis of the cavernosal tissue may not occur in leukaemic priapism as it does in other types. Here we report a 16-year-old patient whose first presentation is priapism, which is a rare clinical presenting feature in Acute Myeloid Leukaemia.

Case Presentation

A 16-year young boy presented in January 2017 with 11 days' history of persistent, painful, penile erection, and bleeding during micturition. He had aspiration without achieving sustained detumescence. On examination, pulse was 100 beats per minutes, blood pressure was 110/60 mmHg. Abdominal examination revealed the liver to be 2cm below right costal margins and the spleen was 4cm below left costal margin, while lymphadenopathy was not present. The patient had an

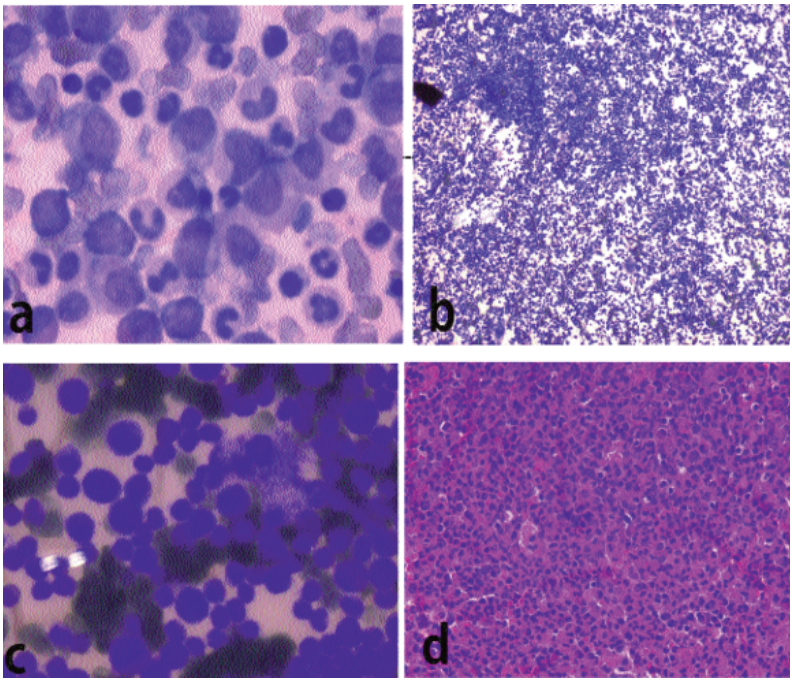

Figure: a) Peripheral smear showing myeloid hyperplasia with neutrophilia, myelocytes, metamyelocytes. b) Bone marrow aspiration slide 10 power image showing hypercellular marrow. c) Bone marrow 100 power image showing myeloid hyperplasia with neutrophils and metamyelocytes peak. d) Bone marrow trephine showing increased megakaryocytes, myeloid hyperplasia (H&E staining).

[1,2,6]Rehman Medical Institute, [3-5]Rehman Medical College, Peshawar.
Correspondence: Adnan Khan. Email: adnan-khan@outlook.com

erect penis, which was tender with venous engorgement. He had no trauma to the penis and no history of using sexual stimulation. There was no fever and weakness. Laboratory investigations showed haemoglobin 5.7gm./dl; Total leukocyte count 614.8x10^9/l, Myeloblast 65%, Promyelocytes 10%, Myelocytes 26%, Metamyelocytes 18%, Band cells 4%, Neutrophils 30% and Lymphocytes 7%: ESR 2mm, platelets 709x10^{12}/L. Urea, Uric acid, and Creatinine were normal. Peripheral smear showed myeloid hyperplasia with neutrophilia, myelocytes, metamyelocytes (Figure-a). Bone marrow aspiration showed myeloid hyperplasia with neutrophils and metamyelocyte predominating (Figure-b, c). Bone marrow Trephine showed increased megakaryocytes, and myeloid hyperplasia (Figure-d).

Detection of BCR-ABL confirms the diagnosis of Chronic phase CML with priapism was made. Hydroxyurea 1gram daily were commenced immediately. Aspirin 100 mg (Disprin CV), Allopurinol 300 mg daily with adequate hydration was also started.

Four pints of blood were transfused in 2 days and the patient was then shifted to operation theater for Corporal irrigation and Winter shunting. The erection gradually reduced following aspirations and washes.

The patient responded to the treatment and were able to achieve an erection with manual stimulation. At present, the leukaemia is in remission.

Discussion

The condition priapism was named after the Greek God Priapus, son of Zeus. It is believed that a jealous Hera cast a spell over his mother while pregnant causing Priapus to be born with the condition bearing his name.

Priapism is a urological emergency, which must be treated early to prevent erectile dysfunction. Priapism is a rare condition with an incidence of 1.5 cases per 100,000 person-years. 20% of cases of priapism are caused by Haematological conditions. Priapism secondary to penile metastasis is rare and one of the etiological factors.[4]

The first case of CML, priapism was described early in 1974, by Schreibman et al.[1] It occurs in 1-2% of CML male patients, with a bimodal age distribution of 5-10 and 20-50 years old, yet described in all age groups.[5] Only few case reports were published describing the occurrence of priapism as a complication of CML.

Hyper-leukocytosis is regarded to be the cause of priapism in patients with leukaemia. Three different mechanisms have been described which are as follows: congestion of the corpora cavernosa occurring due to mechanical compression of the abdominal veins by the enlarge spleen; Sludging of leukaemic cells in the corpora cavernosa and the penile dorsal vein; infiltration of the sacral nerves and central nervous system with leukaemic cells.

Management of priapism depends on whether it is high or low-flow. Leukaemic priapism is usually ischaemic (low-flow) and therefore a urological emergency requiring urgent intervention. Our patient had hyperleukocytosis and features consistent with low-flow. Because leukaemic priapism is a relatively rare occurrence, there is no standard treatment recommended for it. However American Urological Association recommends that systemic treatment of an underlying disorder should not be undertaken as the only treatment for ischemic priapism.[6] It is required to administer Intra-cavernous treatment concurrently. As ischaemic priapism is a compartment syndrome it requires treatment directed at the penis primarily. The patient had the aspiration of the corpora cavernosa and irrigation with epinephrine and achieved detumescence which was not sustained. Allopurinol 300mg daily and hydroxyurea 2g daily were commenced immediately to ensure cytoreduction.

He had Winter shunt and achieved detumescence which was sustained. This strongly supported the recommendation made above concerning combining the treatment for the primary cause of priapism with intracavernosal intervention. Winter shunt is a form of cavernoglanular shunt. Other cavernoglanular shunts include the Ebbehoj, T-shunt and Al-Gorab shunts. These are distal shunts. Other forms of treatment for priapism include proximal shunts such as the Quackles and caverno-venous shunts.

Our case is exceptional as priapism known to be a very uncommon complication of leukaemia. The importance of urgent diagnosis and management of priapism cannot be overemphasized, as there is a definite incidence of the risk for impotence following this complication. Apart from the initial management of priapism, further investigations and proper treatment of the underlying cause is more important. Urgent management with combined urological therapy along with oncological treatment for priapism, the patient was rapidly relieved of this clinical problem.

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