CASE REPORTS

Priapism as a Result of Chronic Myeloid Leukemia: Case Report, Pathology, and Review of the Literature

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ABSTRACT

Introduction. Priapism is rare-presenting feature in male patients with chronic myeloid leukemia (CML). Several hypotheses for pathogenesis have been described. Management has been controversial; some authors described resolution following priapism-specific interventions, and others recommended addition of CML-specific therapy or even CML-specific therapy alone.

Aim. In this report, we describe presentation and management of a man with refractory priapism that was the first presenting manifestation of CML. We also report, for the first time, the pathology sections of the sinusoidal tissue in such cases. Literature is reviewed for similar cases and their outcome.

Methods. A 21-year-old male patient presented with painful priapism that started 6 days earlier and failed aspiration–irrigation. CBC revealed marked leucocytosis. Oncology care diagnosed CML, and treatment with Imatinib was commenced with prior semen cryopreservation. Following remission, a penile prosthesis was implanted, assisted by optical corporotomy. Sinusoidal tissue biopsy was stained by hematoxylin/eosin (H&E) and CD34.

Main Outcome Measures. Pathology sections of cavernous tissue following CML-induced priapism.

Results. The penile implant survived without complications. H&E examination of the sinusoidal tissue biopsy revealed leukemic infiltration associated with vascular endothelial damage. CD34 staining showed the mixed picture of leukemic infiltrates, intact vascular endothelium with lumena showing leukemic cells, alternating with destroyed vessels, and no vascular lumena and ruminants of endothelial cells.

Conclusion. Priapism can be the first manifestation of previously undetected CML. The pathological picture of sinusoidal tissue in such cases is presented. In the case at hand, a complete blood picture was helpful in early diagnosis of CML and early initiation of targeted chemotherapy along with the corporal irrigation/aspiration or shunt surgery. It is therefore recommended to have a CBC examined at presentation of any case of ischemic priapism of unknown etiology, early initiation of CML therapy along with aspiration/irrigation, preferably cryopreserving a semen sample before CML therapy. **Shaeer OKZM, Shaeer KZM, AbdelRahman IFS, El-Haddad MS, and Selim OM. Priapism as a result of chronic myeloid leukemia: Case report, pathology, and review of the literature. J Sex Med 2015;12:827–834.**

Key Words. Priapism; CML; Chronic Myeloid Leukemia

Introduction

Priapism is an emergency condition characterized by full or partial penile erection that continues more than 4 hours beyond sexual stimulation and orgasm or is unrelated to sexual stimulation. There are two types of priapism: ischemic (veno-occlusive or low flow) priapism and nonischemic (arterial or high flow) priapism [1].

The most common cause of ischemic priapism is idiopathic, while the cause of nonischemic pria-

pism is mostly perineal or penile trauma. Other causes of ischemic priapism include intracorporal injection of vaso-active substances such as papaverine, phentolamine, and prostaglandin E1, as well as hematological diseases such as sickle cell disease, hyperviscosity syndromes such as chronic myeloid leukemia (CML) polycythemia vera, and multiple myeloma and hypercoagulable state. Further causes of nonischemic priapism include penile revascularization, neurogenic condition, and intracorporal injection of vaso-active substance [2].

Malignant priapism, or priapism secondary to locally invasive or metastatic cancer, is a rare phenomenon. Ahmed et al. in 2012 reported a case of priapism secondary to malignant urothelial cancer who underwent a radical cystectomy with negative margins but positive nodal disease 3 months prior to presentation [3]. A similar case report was published by Casoli et al. in 2002 [4]. A case report for patient with lung cancer presented with Yokoi et al. on a patient with squamous cell carcinoma of the lung treated with pneumonectomy [5]. Lin et al. described a case of priapism in a patient with adenocarcinoma of the prostate after radical radiotherapy [6]. Priapism also reported secondary to local infiltration of the penis from bladder and prostate cancer [3,7]. The best approach for such case is not well defined; however, surgical resection seems to be the most radical approach however it may be a very aggressive treatment in a palliative setting.

Priapism is rare-presenting feature in male patients with CML, which is a clonal myeloproliferative neoplasm characterized by the presence of the Philadelphia (Ph) chromosome [8]. CML is characterized by reciprocal translocation of chromosomes 9 and 22 (t[9;22][q34;q11]), resulting in the BCR-ABL1 gene rearrangement, which gives rise to uncontrolled growth of myeloid cells in the bone marrow. The exact reason for the translocation is not clear but is probably because of simultaneous chromosomal breaks and repair during mitosis, facilitated by close proximity of chromosomes 9 and 22 in the interphase nucleus [9]. Only radiation seems to be a risk factor as it has been associated with an increased incidence of CML [10]. The peak incidence is in the fifth and sixth decades of life, with an annual incidence of one to two cases per 100,000 people. More than 10% of newly diagnosed adult leukemia diagnosed in the United States are CML cases per year [11].

In CML, priapism may be secondary to hyperleucocytosis with leucostasis/hyperviscosity which usually occurs with extreme elevation of the white cell count to $>100 \times 109$/L that finally lead to venous obstruction due thrombi and microthrombi [12]. Another contributing factor may the overproduction of cytokines and adhesion molecules by leukemia cells that lead to increased sequestration of cells in the microvasculature.

The first case of CML, priapism was described early in 1974, by Schreibman et al. [13]. 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 [14]. Only few case reports were published describing the occurrence of priapism as complication of CML [13,15–27].

In this report, we describe presentation and management of a young man with refractory priapism that was the first presenting manifestation of CML. We also report, for the first time, the pathology sections of the sinusoidal tissue in such cases and postulation of pathogenesis based on the pathology findings.

Methods

A 21-year-old male patient presented with painful priapism that started 6 days prior to presentation. The patient denied any preceding intake of erection-enhancing medications whether oral, parenteral or intracavernosal, recreational drug use, or exposure to trauma, and was unaware of any systemic illness he might have had, with the exception of mild dyspepsia or sense of gastric fullness with meals. General examination was uneventful with the exception of palpable splenomegaly, three fingers below costal margin.

Before presenting to our practice, the patient was exposed to several trials at cavernosal aspiration/irrigation and injection of epinephrine, which all failed. The penis was fully rigid, with bluish discoloration, and was relatively warm, a sign of inflammation.

Penile prosthesis surgery was decided. Preoperative investigations were performed, including sickling test which turned out negative. Complete blood count (CBC) revealed marked leukocytosis (410,000 cells/cm^3) and infiltration with immature cells. Hematologic malignancy was suspected, and the patient was referred to an oncologist.

Leukapheresis using a cell separator lowered white blood cell (WBC) counts rapidly. Patient was prescribed Imatinib 400 mg daily with meals, which was well tolerated. Chromosomal translocation analysis using the polymerase chain reaction (PCR) revealed the Philadelphia (Ph) chromo-