

PRIAPISM

a rare and unusual presentation in chronic myeloid leukemia

[a case report]

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Introduction :

The term priapism has its origin in reference to the Greek god Priapus, who had a disproportionate permanent erection. Priapism is caused by imbalance of penile blood inflow and outflow. It is a condition in which a penis remains erect for hours in the absence of stimulation or after stimulation has ended. There are three types: ischemic (low-flow), non-ischemic (high-flow), and recurrent ischemic (intermittent). Most cases are ischemic. Ischemic priapism is generally painful while nonischemic priapism is not. The etiology is usually known, management guidelines are generally directed at rapid treatment versus rapid diagnosis, and it is uncommon and rare for clinicians to workup the underlying cause. Although Sickle Cell Disease is a predominant etiology of veno-occlusive Priapism cases in the literature, there are a wide variety of other causes ranging from urinary retention to insect bites. Hematologic dyscrasias are also a major risk factor for ischemic priapism. Priapism may occur in patients with excessive white blood cell counts. We report a case of a young 25-year-old adult who presented to the Emergency Department with priapism as the first and only manifestation of chronic myeloid leukemia.

Case report :

A previously healthy 25-year-old man presented to the Emergency Department with history of an erect penis of over 16 hours duration. His penis was erect, painful and engorged when he arrived at emergency department. There was no history of trauma, sexual stimulation in the past 24 hours, malaise, night sweats, joint pain, and cough. His medical history gave no clue and no specific orientation except a smoking history of 4 packs-year, no other reported medical conditions, medication list, or symptoms. The patient also stated that this was the first episode of Priapism he suffered from and it was not related to any sexual stimulation.

The physical examination revealed an erected, painful and swollen penis. The rest of the physical examination was clueless: no signs of swelling in the liver, lymph nodes, and spleen. Furthermore the patient was a bit anxious but in a good overall condition, no pallor was noticed.

In front of the clueless history, a complete blood count (CBC) has been requested.

The laboratory data showed: hemoglobin (Hb) 10.3 g/dl, hematocrit 29.3%, white blood count (WBC) 501 570/mm³ and platelet 269,000/mm³. The WBC showed a 20% of Myelocytes and a 14% of Metamyelocytes. (Fig.2)

Urologists were contacted for a rapid and efficient treatment of the Priapism. The treatment was initiated by corporal aspiration at the Emergency Department. The patient also has been given treatment for the pain. The erection gradually reduced.

The laboratory data and the bone marrow examination later requested helped identifying the underlying etiology, the patient was diagnosed as a case of Chronic Myeloid Leukemia (CML). He has been transferred to the Hematology department for a specific care of his CML.

Discussion :

The condition priapism was named after the Greek god Priapus, thought to be the son of Zeus. It is believed that a jealous Hera or Aphrodite cast a spell over his mother while pregnant (either Aphrodite or Chloe) causing Priapus to be born with the affliction bearing his name and resulting in him being disowned by his mother. Priapism is an urological emergency, which must be treated early to prevent erectile dysfunction. Priapism is a rare condition on its own with an incidence of 1.5 cases per 100,000 person-years.^{1 5} Priapism is a pathological condition characterized by penile erection that persists for longer than six hours and is unrelated to sexual stimulation. It is further categorized by Hauri D, et al. and Winter CC, et al. as high flow (non-ischemic) and low-flow (ischemic) where low-flow is caused by hematologic disease; mainly sickle cell disease and rarely leukemia, if untreated, results in necrosis of the cavernous muscle and subsequent fibrosis and erectile dysfunction.^{2 3}

Kamal K et al. in their retrospective study of a series of 28 cases of low-flow priapism seen lately after 12 hours in a period of 21 years from 1991 to 2011 : Priapism is considered as a major andrological emergency that needs to be treated without delay before any investigation. The prognosis depends mainly on the age of the patients and the precocity of their treatment. They also noticed in their study the failure of the medical treatment in all cases and the success of the aspiration in only 4 cases. The 24 other cases needed surgery. Twenty patients underwent distal caverno-cancellous anastomosis and 4 had proximal anastomosis.

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The average follow-up was 46 months and the erectile capacity was preserved in 44% of our patients.⁹ In our case report the treatment chosen was aspiration because of the 16 hours duration Priapism. With this treatment the erection gradually reduced. The age of our patient (25 years) and the precocity of treatment may have avoided the need of surgical treatment.

The proposed algorithm by their study is interesting to be used in an emergency department and applied in order to permit a right procedure in front of any Priapism seen early or lately.⁹ (Fig. 3)

Although low-flow Priapism is essentially and frequently a manifestation and presentation of SCD it is rare and uncommon to be a presentation of Chronic Myeloid Leukemia (CML). Our case is unique as priapism was the only presentation of the underlying leukemia. The importance of prompt diagnosis and treatment of priapism cannot be overemphasized. Few others case reports have shown a same uncommon presentation of CML.^{1 4 6 7 8} In the case reported by Rajendra B. Nerli and al. the patient also presented a swollen spleen and liver.

Chronic myeloid leukemia (CML) is an indolent form of leukemia of the myeloid lineage and may exist in 3 distinct phases, (1) chronic phase, (2) accelerated phase, and (3) blast crisis. The genetic translocation responsible for this disease is a reciprocal translocation between chromosomes 9 and 22, also known as the Philadelphia chromosome, creating

The fusion gene BCR-ABL1 that leads to a constitutively active kinase that drives oncogenesis. Depending on the CML phase and biology, patients may have a profoundly elevated WBC at diagnosis, which can present as leukostasis, or hyperviscosity syndrome^{1 5}.

Hyperviscosity syndrome is associated with serious clinical manifestations including visual discomfort (retinopathy, retinal hemorrhages), shortness of breath (pulmonary insufficiency), priapism, and other symptoms related to end organ damage¹. In our case the Priapism is a result of the hyperviscosity syndrome induced by the highly elevated WBC (over 500 000/mm³).

Conclusion :

Rare but with a high complication rate, Priapism is an uncommon presentation of Chronic Myeloid Leukemia.

Priapism management needs rapid urologic treatment to prevent erectile dysfunction and other severe complications that may arise from priapism.. Priapism due to hyperviscosity can be the first presentation and only in patients with Chronic Myeloid Leukemia as seen in our case report, and CML should therefore always be considered in a patient with priapism. High Index of Suspicion, as well as early full blood count and peripheral blood/Bone marrow examination among patients presenting with priapism will facilitate prompt and correct diagnosis.

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Prélèvement transmis au Laboratoire

RESULTAT PROVISOIRE EN ATTENTE DE VALIDATION BIOLOGIQUE

HEMATOLOGIE

NUMERATION FORMULE SANGUINE
(Technique Cytométrie de Flux sur SYSMEX XT 4000 i)

	MORMES	ANTECEDENT
HEMATIES	3,48 M/mm ³ (4.2 à 9.7)	
Hémoglobine.....	10,3 g/l (14.0 à 17.0)	
Hématocrite.....	29,3 % (40 à 52)	
V.G.M.....	84,2 fl (60 à 95)	
T.G.M.H.....	29,6 pg (28 à 32)	
C.G.M.H.....	35,2 % (30 à 35)	
LEUCOCYTES	501 570 /mm ³ (4000 à 10000)	
Polynucléaires neutrophiles..	53,0 % (40 à 75)	
soit...	!!!!!! /mm ³ (2000 à 7500)	
Lymphocytes.....	2,0 % (20 à 45)	
soit...	10 031 /mm ³ (1500 à 4000)	
Monocytes.....	3,0 % (2 à 8)	
soit...	15 047 /mm ³ (200 à 800)	
Polynucléaires éosinophiles..	3,0 % (1 à 4)	
soit...	15 047 /mm ³ (100 à 600)	
Polynucléaires basophiles....	2,0 % (0 à 1)	
soit...	10 031 /mm ³ (0 à 150)	
Blastes	0 %	
Myéloblastes	0 %	
Promyélocytes	3 %	
Myélocytes	20 %	
Métamyélocytes	14 %	
PLAQUETTES	269 Mille/mm ³ (150 à 450)	
Volume Plaquettaire Moyen ...	13,0 fL (N : 7.2 à 11.1)	

Figure 2 : Complete Blood Count of our patient

PRIAPISME

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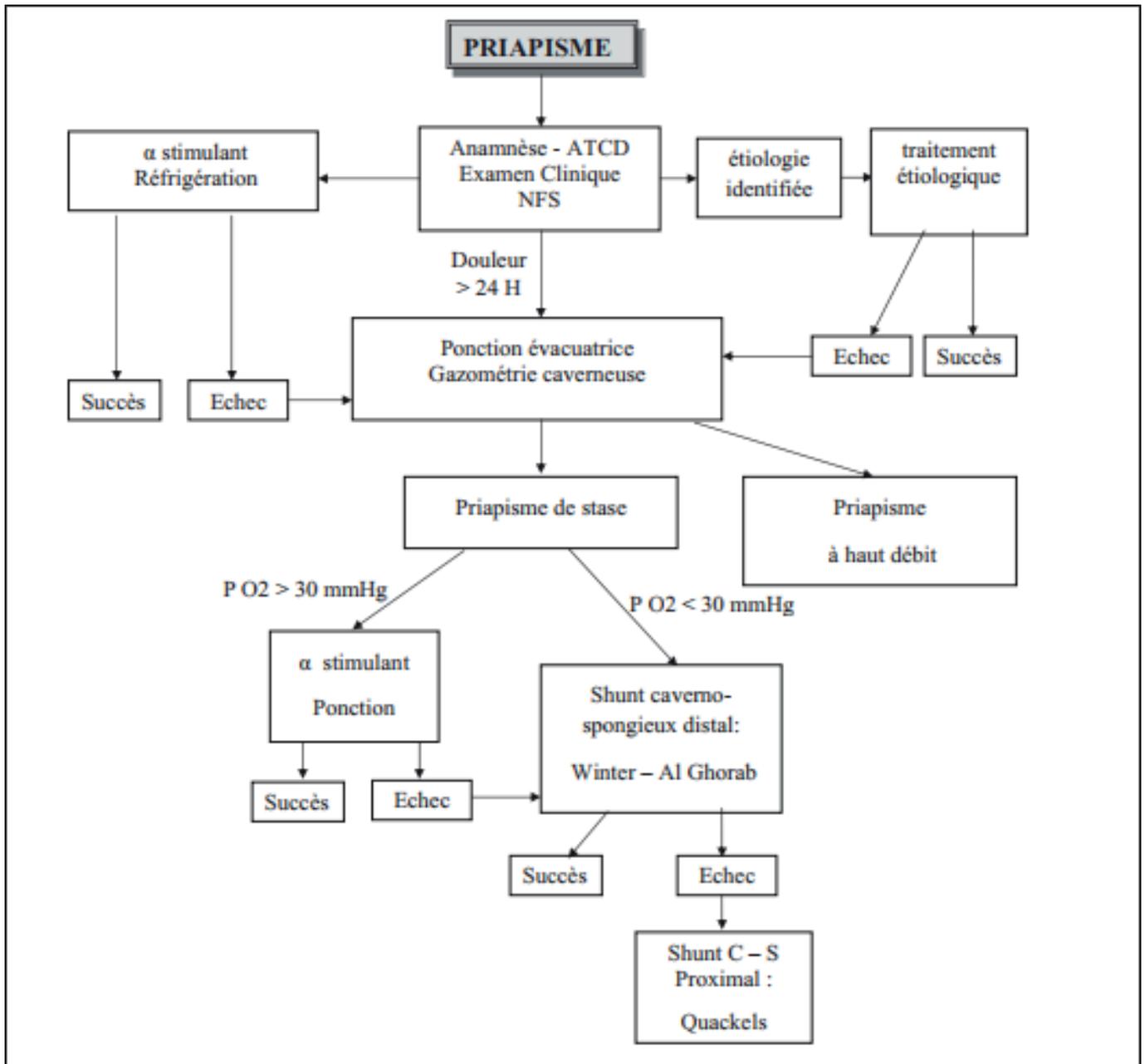


Figure 3 : Priapism management algorithm

{K. Kamel and al. Prise en charge du priapisme à bas débit vue tardivement : à propos de 28 cas. African Journal of Urology Volume 22, Issue 4, December 2016, Pages 297-304}