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Acute Priapism: A Revealing Inaugural Mode of Chronic Myeloid Leukemia. A Case Report and Review of the Literature

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Abstract

Priapism is a painful and prolonged erection occurring without any sexual stimulation and not resulting in ejaculation. It most often occurs in patients with sickle cell disease. We report here the case of acute recurrent priapism revealing inaugural mode of CML in a 22-year-old patient with no particular pathological history. The study of the onco-hematological karyotype revealed a karyotype with 46 chromosomes with a clonal chromosomal anomaly: The t (9; 22) "Philadelphia chromosome". The evolution was favorable under imatinib.

Keywords

Priapism, Chronic Myeloid Leukaemia, Philadelphia Chromosome

1. Introduction

Priapism is a prolonged, often painful, pathological erection occurring without sexual arousal and not leading to ejaculation. It is a urological emergency because the functional prognosis of the penis may be at stake. It is a condition that is encountered in our African context often in sickle cell patients. When this etiology is not found, other causes, particularly hematological ones, are sought. Simple symptomatic management of so-called "idiopathic" priapism, without thorough etiological research, will lead to recurrence with possible severe functional sequelae. Less than a hundred cases of priapism occurring in leukemic

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subjects or revealing leukemia have been described in the literature [1]-[8]. In this case, we report an acute recurrent priapism in a 22-year-old man with no previous history of leukemia, which ultimately revealed chronic myeloid leukemia. The evolution after the realization of a cavernous-spongy shunt, and the beginning of the treatment of the chronic myeloid leukaemia was favourable.

2. Observation

This was a 22-year-old patient admitted for a painful, involuntary and prolonged erection that had been evolving for 13 hours. This episode had been preceded by a first one 48 hours before, with a favorable evolution after cavernous evacuation puncture and injection of alpha-stimulant drugs.

The history did not reveal any personal or family medical or surgical history, no notion of medication taken before the onset of the symptomatology, nor of perineal trauma.

The clinical examination revealed an apyretic patient, in good general condition, with an erect penis, with hard cavernous bodies, contrasting with a supple glans. Local infiltration was noted with an ecchymotic aspect of the penis extending to its root and to the bursa related to the previous iterative punctures. The rectal examination was normal.

The haemogram showed white blood cells at 403,500, haemoglobin (Hb) at 6 g/dl and platelets at 460,000/mm^3; uraemia at 0.19 g/l, creatinemia at 15 g/dl, C-reactive protein at 88 mg/l. A blood smear showed anisopoikilocytosis of red blood cells with rare erythroblasts; white blood cells: myelmia; platelets: thrombocytosis

The myelogram showed a significant hyperplasia of the granular lineage at 96% without signs of dysplasia with myeloblasts at 10%, eosinophils at 4% and basophils at 6% (**Figure 1**).

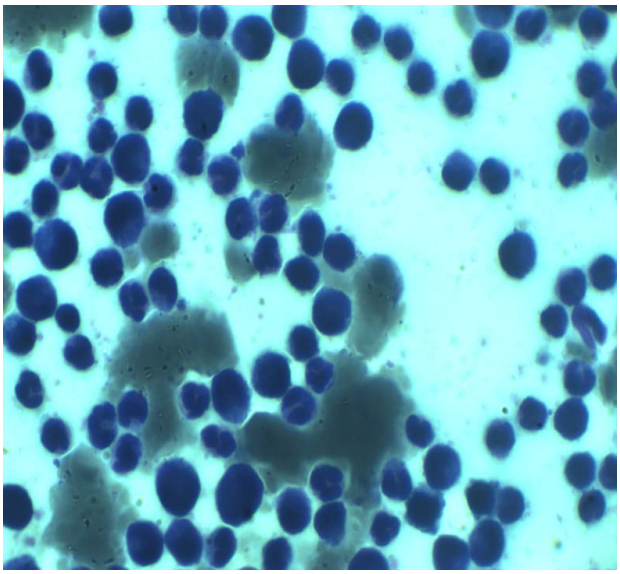

Figure 1. Myélogramme.

The karyotype study confirmed the diagnosis of CML, showing a 46-chromosome karyotype with a clonal chromosomal abnormality: the t (9; 22) "Philadelphia chromosome".

The patient benefited from a cavernous-spongiosa shunt because of the persistence of priapism despite several evacuation punctures. Hyperhydration with physiological serum at a rate of 5 liters per day was also instituted. In view of the repeated punctures, which caused infiltration and an ecchymotic aspect of the penis, exposing the patient to gangrene, antibiotic therapy with ceftriaxone 2 grams per day was instituted.

In relation to chronic myeloid leukemia, the patient was put on a Tyrosine kynase inhibitor (ImatinibR: 400 mg/d), associated with Allopurinol 300 mg/d.

The evolution was marked by a regression of the pain after 24 hours, a detumescence of the penis, a resorption of the ecchymosis of the external genitalia. At the end of the episode, there was a loss of erectile function.

On the haematological level, the control blood count done 1 month after the treatment showed a normalization of the rate of the different blood lines. Cytogenetic tests for the Philadelphia chromosome were negative. On the molecular level, PCR tests for BCR-ABL abnormalities were also negative.

3. Discussion

Priapism is a urological emergency; and the publications reported in the literature on this subject concern, for the most part, cases secondary to sickle cell disease (the main etiology in Africa), or to the use of psychotropic drugs (the main etiology in Europe) [9] [10]. Kamal *et al.* [10] in a series of 28 patients noted 1 case of priapism revealing CML while Fall *et al.* [11] in Senegal found 2 cases out of 63 patients collected over a 10-year period. This could be explained by the rarity of these two conditions, and the even lower probability of presenting with this complication during leukemia. Indeed, the most frequent circumstances for the diagnosis of CML are an anemic syndrome, an alteration of the general state of health, a prolonged fever, and clinically hepatomegaly and splenomegaly [12]. Furthermore, a proportion of priapism seen in emergency departments and labelled as "idiopathic", representing 30% of the causes of priapism [8], could be related to an undiagnosed disease, after a rapid interrogation to rule out sickle cell disease, drug intake or the use of intra-cavernous injections, and in the absence of a minimal biological work-up including a blood count. It should be noted in our case that the patient went home following remission of priapism after cavernous puncture during the first episode. He was admitted and further investigated on the occasion of the recurrence 48 hours later. Nevertheless, a few publications have been reported in the literature on priapism of leukemic origin (**Table 1**).

The time of consultation is the essential element determining the therapeutic choice and the functional prognosis of the penis. In the Senegalese series [11], 50% of patients were seen between the 24th hour and the 5th day, justifying radical surgical treatment (creation of a cavernous-spongy fistula) after failure of

Table 1. Literature review of cases of priapism in chronic myeloid leukemia.

Authors	Number	Time to consultation	Treatment	Erectile dysfunction
Shaeer *et al.*	1	4 days	Failed puncture, imatinib.	Yes
Yoshida *et al.*	1	2 days	Failed puncture, shunt caverno-spongieux, imatinib	No
Tazi I	1	22 hours	Puncture, imatinib	No
Chang M.W *et al.*	1	19 hours	Puncture, interferon alfa, allopurinol	No
Ergenc H *et al.*	1	18 hours	Imatinib, leukapheresis	No
Rojas B *et al.*	1	36 hours	Leukapheresis, Cavernous-spongy shunt	Yes
Nerli R.B *et al.*	1	24 hours	hydratation	No
Fall *et al.*	2	24 hours	Cavernous-spongy shunt	No
Sarr A	1	6 hours	Puncture	No
Dhar J	1	4 hours	Puncture	No

cavernous puncture. Portillo [13] in Spain found a consultation time varying between the 4th and 72nd hour. In our case, the patient had consulted 13 hours after the onset of symptoms. The patient benefited from cavernous puncture. Tazi *et al.* [1] in Morocco and Chang *et al.* [5] in China observed detumescence after evacuation puncture in patients admitted for priapism due to haemopathy and who consulted within 24 hours. Rojas, Nerli, Yoshida [2] [3] [4] used cavernous-spongiosa shunt in patients with priapism due to haematological disease and who consulted beyond 24 hours.

The initial management of priapism in CML, as in other etiologies, involves aspiration of the corpora cavernosa and injection of a vasoconstrictor, with the aim of obtaining detumescence. In daily practice, it is common to receive patients after the 6th hour, making it difficult to obtain detumescence due to the onset of ischemia and fibrosis of the cavernous tissue [3] [4]. In our case, detumescence was obtained after puncture. This management included a second, etiological aspect related to chronic myeloid leukemia and included hyperhydration, allopurinol, imatinib and prevention of possible infection by antibiotic therapy. Stabilization of the basic pathology will prevent a recurrence of priapism. The evolution was marked by a loss of erectile function. Of the reported cases of priapism in leukemia (**Table 1**), only Shaeer and Rojas [4] [6] also described erectile dysfunction after treatment.

4. Conclusion

Priapism can be the telltale sign of CML, although it is rare. We must suspect it in a subject without any particular medical or surgical history and presenting an abnormality of the blood lines on the haemogram.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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