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PRIAPISM : RARE PRESENTING MANIFESTATION OF CHRONIC MYELOID LEUKEMIA AND ITS MANAGEMENT – CASE SERIES OF 5 PATIENTS

Clinical Hematology

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ABSTRACT

Priapism is defined as a persistent penile erection that continues hours (4 h) beyond, or is unrelated to, sexual stimulation. Priapism has incidence of 1.5 cases per 100 000 person-years. Haematological conditions are the cause of 20% of cases of priapism in men . Chronic myeloid leukemia (CML) accounts for half of this cases. Based on this case series we can conclude that as CML and various other hematological disorders can present as priapism , hence all the patients with priapism need to undergo basic hematological work up at diagnosis of priapism. Early diagnosis and prompt management of CML associated priapism can preserve male potency.

KEYWORDS

Introduction

Priapism is defined as a persistent penile erection that continues hours (4 h) beyond, or is unrelated to, sexual stimulation. Priapism has incidence of 1.5 cases per 100 000 person-years. Haematological conditions are the cause of 20% of cases of priapism in men. Chronic myeloid leukemia (CML) accounts for half of this cases. The primary mechanism is the aggregation of leukaemic cells in the corpora cavernosa and the dorsal veins of the penis. Various treatment options in form of penile aspiration, shunt surgeries are available with variable success rate. Patients presented as priapism and later dignosed to have CML at our centre in last 5 yrs are discussed in this article, along with their management and review of literature. One case with classic presentation has been described in detail.

Case

30 yrs old male,admitted in our hospital for persistent, painful erection of penis associated with tightness, uncomfortable sensation since last 20 hrs. (Image 1) There was no history of sexual stimulation, trauma ,drug use, previous similar episodes, bone pains, jaundice, swelling over body, blood transfusion.

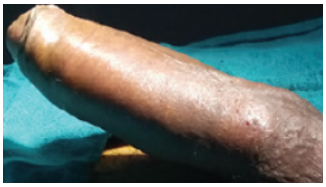

Image 1: Persistent & painful erection of penis (Priapism)

On examination patient was in agony, 2cms splenomegaly noted, penis was rigid with soft glans and engorged superficial veins. Patient's complete hemogram was suggestive of hemoglobin-11.2 g/dl,Total WBC count -2,85,000/cumm, platelet count of 4.62 lakhs/cumm with to shift to the left (Myelocytes 22%,Metamyelocytes 10%,Band Forms 61%). (Image 2)

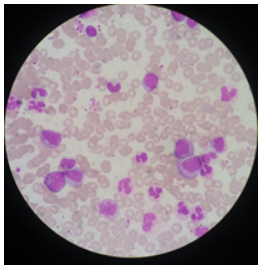

Image 2: Peripheral smear in CML CP showing shift to left (Leishman stain , 100X)

Patient diagnosed with ischemic type priapism on basis of history and clinical examination. With suspicion of chronic myeloproliferative disorder patient started on C. Hydroxyurea 500 mg , 2 capsules 3 times a day with hydration 3 litre/m2 and allopurinol 100 mg three times a day. Tumor lysis syndrome profile was normal. Soon after admission multiple intracavernous aspirations with irrigation and injections of Phenylephrine also tried,but in view of persistence of priapism, patient undergone procedure of 'AI-Ghorab shunt', shunting bilateral corpora cavernosa on same day. Patient's bone marrow aspiration and bone marrow biopsy done suggestive of CML- chronic phase. Patient's FISH revealed double fusion of BCR-ABL in 98% of interphase cells. Patient's hydroxyurea stopped and started on C.Imatinib 400 mg daily, allopurinol continued for next 7 days. Patient regularly followed on further visits with RQ PCR for BCR ABL . Patient s RQ PCR BCR

ABL IS Ratio at 6 months and at 1 year was 1.7 % and 0.2 % respectively and he also maintained ability of erection and ejaculation.

Clinical Presentation of cases

Priapism was seen in 5 (1.52%) out of 317 CML patients at presentation in the last 5 yrs

Case No	Age (yrs)	Duration	Prior history of priapism episodes	Type of priapism	Other symptoms
1	30	20 hrs	No	Painful	-
2	26	3 Days	Yes, 2 episodes (Not Treated)	Painful	Weight loss, Abdominal discomfort, Dyspnoea on exertion
3	25	2 Days	No	Painful	Fever ,Abdominal discomfort
4	28	7 hrs	No	Painful	Abdominal discomfort, Dyspnoea on exertion
5	29	3 Months	Yes,3 episodes (Not Treated)	Painless	Fever, weight loss

Clinical Examination & Investigations of cases

Case no	Splenomegaly (below left costal margin)	Hepatomegaly (below right costal margin)	HB (gms/dl)	WBC Count	Platelet count	% FISH for BCR-ABL Positivity	BMA Finding
1	+, 2 cms	+, 3 cms	11.2	285 x 10^9/L	462 x 10^9/L	98%	CML-CP
2	+,15 cms	-	8.9	292 x x 10^9/L	490 x 10^9/L	84%	CML-CP
3	+,1 cms	-	11.3	607 x 10^9/L	320 x 10^9/L	100%	CML-CP
4	+,15 cms	-	7.0	441 x 10^9/L	422 x 10^9/L	98%	CML-CP
5	+,3 cms	-	10.5	284 x 10^9/L	370 x 10^9/L	98%	CML-CP

Treatment & Follow Up of cases

Case No	Medical management	Surgical treatment	CML course (RQPCR for BCR ABL monitoring- IS Ratio)	Outcome of priapism assessed at 6 months in form of EHS2
1	Hydroxyurea , Allopurinol ,Hydration Imatinib	Aspiration Shunt	0.2 at 12 months	Grade 4
2	Hydroxyurea , Allopurinol ,Hydration Imatinib	Aspiration Shunt	0.86 at 12 months F/B hematological relapse at 46 months	Grade 2
3	Hydroxyurea , Allopurinol ,Hydration Imatinib	Aspiration Shunt	Blast crisis with leukemia cutis at 3 months	Grade 1
4	Hydroxyurea , Allopurinol ,Hydration Imatinib	Aspiration Shunt	0.8 at 6 months	Grade 4
5	Imatinib	-	0.09 at 12 months	Grade 1

Aspiration - Intracavernous aspirations & phenylephrine injections, Shunt - Al-Ghorab shunt

CCyR -Complete cytogenetic response ,HU-Hydroxycarbamide, ALP- Allopurinol

EHS-Erection Hardness Score, grade1: Penis is larger but not hard, grade 2: Hard but not hard enough for penetration, grade 3: Hard enough for penetration but not completely hard, grade 4: Completely hard and fully rigid

Results

1. Priapism was seen in 5 (1.52%) out of 317 newly diagnosed CML patients at our centre in the last 5 yrs
2. 103 (32.65%) out of 317 newly diagnosed CML patients were of age 30 yrs or less at presentation & priapism seen in 5 i.e. 5.1% patients of this age group
3. Age ranged from 25 -30 yrs with mean WBC count of 3,82,000/ cumm
4. Two of our patients presented within 24 hrs of symptoms onset
5. One patient presented after 3 months so didn't undergo any urological intervention
6. Four patients undergone intracavernous aspiration & phenylephrine injections , followed by surgical shunt procedure
7. Erectile dysfunction developed in three out of five patients when assessed at 6 months post diagnosis.
8. Imatinib was started in all the patients after confirmation of diagnosis
9. While one patient progressed to blast crisis with leukemia cutis at 3 months of presentation and one had hematological relapse at 46 months, others achieved satisfactory molecular remission at different time points.

Discussion

Priapism is defined as either low-flow (ischemic) or high-flow (non-ischemic). Low-flow or ischemic priapism results from pathologically decreased penile venous outflow that results in stasis. Intracavernosal blood sampling reveals acidosis and a decrease in oxygen tension. Clinically low-flow priapism manifests as a rigid, painful penile shaft with a soft glans. This type is more common and represents an actual emergency because irreversible cellular dam age and fibrosis occur if treatment is not administered within 24 to 48 hours. It will result long term sequel of erectile dysfunction or predisposition to frequent, prolonged episodes of priapism. The cause of low-flow priapism including idiopathic,hematologic disorders, tumor infiltrate, or drugs induced.

High-flow or arterial priapism differs in that it is secondary to trauma to perineal area or penis which results in increased arterial inflow into the cavernosal sinusoids, which overwhelms venous outflow and clinically, the entire penis is partially rigid and painless . In contrast to low-flow priapism, intracavernosal blood sampling from patients with high-flow priapism reveals bright red oxygenated blood, and thus irreversible cellular damage and fibrosis are rare. The type of priapism is usually due to penis or perineum trauma that results in injury to the internal pudendal artery.This establishes a fistula between the cavernosal artery and the corpus cavernosum that unregulated inflow occurs. It is not an actual emergency in patients with high-flow priapism, and treatment can be on an elective basis.

Priapism can occur at any age and two peaks in age distribution is described. A pediatric peak, 5-10 years old, is noted owing to sickle cell disease. The secondary peak occurs in patients with active sexual activity age of 20-50 years.

The primary mechanism is the aggregation of leukaemic cells in the corpora cavernosa and the dorsal veins of the penis. Three other mechanism are described:(1) venous congestion of the corpora cavernosa resulting from mechanical pressure on the abdominal veins by the splenomegaly (2) infiltration of the sacral nerves with leukemic cells (3) infiltration of the central nerve system, but there is no evidence to support this in leukaemia

About the management of priapism, initial penile intervention may utilize therapeutic aspiration with or without irrigation. If priapism persists even after aspiration/irrigation, intracavernous injection of sympathomimetic drugs should be performed. If the erection persists for 24-48 hours , patient should have a surgical shunt performed to establish new venous outflow and restore normal arterial flow to the corpora cavernosa.

The importance of prompt diagnosis and treatment of priapism cannot be overemphasized, as there is definite incidence of impotence following this condition. Reports suggest that priapism lasting 5–10 days leads to impotence in 35–90% of men.[xv] So decompression of the penis should be done as frequently as possible during the first 24 hours. There are few case reports of CML presenting as priapism, which has

presentation similar to our cases and were managed similarly with the combined urological and hematological intervention. These reports also reinforces the fact that early urological and hematological intervention can preserve potency in the CML induced priapism patients.

CONCLUSION

As CML and various other hematological disorders can present as priapism , hence all the patients with priapism need to undergo basic hematological work up at diagnosis of priapism. Early diagnosis and prompt management of CML associated priapism can preserve male potency.

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