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Acute lymphoblastic leukaemia presenting as paediatric priapism

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ABSTRACT

A case of spontaneous priapism in a ten year old child with acuta lymphoblastic leukaemia is presented. Emergent management options like drainage, sympathomimetic agents, and medical treatment to reduce circulating white cell counts and shunt surgery are discussed.

Key Words: Paediatric priapism, acute lymphoblastic leukaemia, priapism drainage

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Introduction

Priapism is a penile erection which lasts for more than four hours. It occurs in the absence of sexual stimulation. It is a urological emergency. Prolonged priapism has the risk of penile fibrosis leading to erectile insufficiency. Priapism is rare in children. Sickle cell disease is the common cause of priapism in children. Leukaemia is an uncommon cause of priapism in paediatric age group [1].

Case report

Ten year old male child presented with persistent painful erection for last 24 hours (Fig 1). He had one episode of vomiting, dribbling of urine and difficulty in urination, fever. There was no history of trauma to penis, penile manipulation, haematuria or pain in abdomen. He had no known medical disorder. On Examination he had hepatosplenomegaly. There was no free fluid in abdomen. Penile examination revealed both corpora to be turgid tender. The glans was not turgid. Both testes were normal.

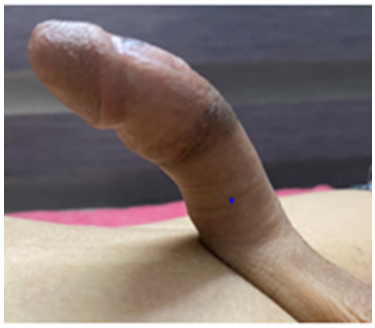

Fig. 1. Clinical photograph showing erect penis.

Laboratory investigations revealed haemoglobin: 10.2; total Leukocyte count: 289000; platelet count: 62000. On peripheral smear the blasts were 90%. His coagulation parameters were deranged; INR: 2.07; APTT Control: 26; patient: 42 LDH-2107. With these findings a diagnosis of ischaemic priapism due to haematological malignancy was kept. Paediatrician and medical oncology opinion was sought. They advised bone marrow aspiration. His bone marrow aspirate was sent.

Priapism was managed on emergent basis. Patient was taken for corporal aspiration with 22G scalp vein. Initially dark red colour blood about 50 cc was drained which gradually turned to bright red colour with detumescence of penis. But soon after removal of scalp vein priapism reappeared. Patient was shifted to intensive care unit for monitoring. He was treated with intravenous fluids, antibiotics and oral ibuprofen.

Bone marrow aspiration was done which showed Acute Lymphoid Leukaemia (ALL). Later patient got drowsy and became irritable. Chest X ray did not show mediastinal mass or enlargement. CT brain showed bilateral intracranial bleed (Fig 2). Repeat aspiration of the corpora was attempted but aspirate was minimal and dark red colour blood that could not be aspirated. Considering his poor general condition and life threatening cerebral complications, priapism treatment took a backseat.

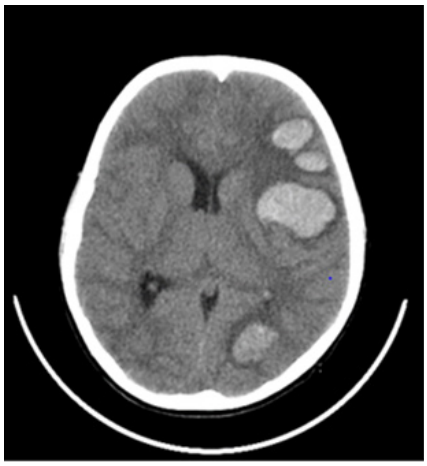

Fig. 2. CT brain showed bilateral intracranial bleed.

In view of high total leucocyte count and high risk for Tumour Lysis Syndrome (TLS) patient was given TLS prophylaxis with oral allopurinol (50 mg TDS), soda-bi-carb tablets and intravenous fluids. Inj Dexamethasone 4 mg was started, TLC and TLS profile (creatinine, potassium, calcium, phosphate) was monitored regularly as ALL is found to be very sensitive to steroids and can result in TLS. His flow cytometry immunophenotyping revealed 65% blasts on peripheral smear. On flow cytometric analysis these blasts expressed CD1a, CD3 (Dim), cCD3, CD4 (variable), CD5, CD7, CD8, CD 38. The findings were consistent with the diagnosis of ALL T cell lineage.

He was referred to an oncology centre where priapism resolved gradually over a period of five days. At the oncology centre he was started on International Berlin-Frankfurt-Münster Study Group (ICBFM 2002) protocol. Induction phase A (28 days cycle) included administration of daunorubicin, vincristine and intrathecal methotrexate (IT MTX) weekly along with oral prednisolone for 28 days. Inj l Asparaginase (leunase) was administered for 10 doses. Post Phase a bone marrow was in remission. Phase B included administration of cyclophosphamide, low dose Cytarabine (Ara-c), oral 6 Mercaptopurine (6 MP) and IT MTX. Post Induction Phase 2 consolidation was planned, which included administration of high dose of methotrexate (5 gram/m^2) every 15 days for 4 cycles.

Post consolidation reinduction for 6 weeks was administered which included similar drugs as in induction,with doxorubicin replacing daunorubicin. Post Reinduction cranial irradiation was administered. Post irradiation patient started on maintenance phase with oral 6 MP and methotrexate. At last follow up he is doing well. He is getting normal erections now. He has been followed up for nine months.

Discussion

Prolonged penile erection lasting for more than four hours unrelated to sexual stimulus is defined as priapism. Widely accepted types of priapism are: ischaemic, stuttering and non-ischaemic. Ischaemic priapism is the commonest one in paediatric age. Non-ischaemic is rare. Subsequent discussion pertains to ischaemic type. Prolonged priapism is a risk factor for penile fibrosis and erectile dysfunction. Priapism is a urological emergency. Priapism is most common in the fifth decade of life. Leukaemia presenting as priapism is uncommon especially in paediatric age group. It is possible that paediatric priapism may be underreported [1]. Sickle cell disease is the commonest cause of priapism in children. Leukaemia constitute around 10% of cases of paediatric priapism [1]. It is postulated that abnormally large blast cells have a tendency to adhere together. These cause sludging in the cavernous bodies [2]. Prolonged erection cases structural damage to the penis and may lead to erectile dysfunction [3].

Management of adult priapism is clearly defined [4]. But there are no clear cut guidelines for the initial management of paediatric priapism. Different options suggested are medical management, drainage of priapism, instillation of sympathomimetic agents, anticancer treatment and shunt surgery.

Castagnetti et al., [5] have reported medical management alone in four cases of leukaemia in children aged 9 to 13 years. Three of these children had chronic myeloid and one had acute T cell lymphoblastic leukaemia. The patients underwent chemotherapy and leukopharesis without drainage of priapism. Initial improvement of priapism was observed by two to thirteen days. It took from five days to three months for complete resolution of priapism. No erectile dysfunction was reported in these children over the follow up period of four to eight years.

Current guidelines [4] warrant against systemic management of ischaemic priapism as the only treatment. In patients with an underlying disorder, such as sickle cell disease or hematologic malignancy, systemic treatment of the underlying disorder should not be undertaken as the only treatment for ischemic priapism. Concurrent intracavernous treatment is recommended. The therapeutic success claimed with systemic management alone are associated after prolonged ischaemic periods. All priapism will resolve over time even without treatment but at the cost of erectile function.

Drainage of the priapism is the mainstay of intracavernous treatment. On its own, aspiration has a success rate of approximately 30%. The use of sympathomimetic injections with or without irrigation has a success rate of 43%–81% [6]. Phenylephrine is the favoured sympathomimetic for intracavernous use as it has lower risk of cardiovascular side effects. Phenylephrine is usually used in concentration of 100 ug/ml-200 ug/ml injected 1 ml every 5 minutes. The patient should be monitored for acute hypertension, headaches, reflex bradycardia, tachycardia, arrhythmias and palpitations. Lower concentrations are recommended in children [6].

There are some concerns about the intracavernous use of sympathomimetic in paediatric patients with ischaemic priapism because of risk of arterial vasospasm and worsening the ischemia. Ischaemic priapism is managed in a step-wise fashion. If aspiration/irrigation, intracavernous injection of sympathomimetic drugs fail surgical intervention in form of shunt surgery is recommended.

Conclusion

We report a rare case of ALL presenting as priapism in paediatric age group managed with drainage and anticancer treatment.

Declarations

Written consent from the parent is obtained.

Consent for publication

Written consent to publish this material has been obtained from the parent. Proof of consent to publish from study participants is available and can be furnished on demand time.

Availability of data and material

All the data pertaining to the case is presented herewith. Additional data about follow up is available if needed

Competing interests

Nil, Not applicable

Funding

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References

[1] Donaldson JF, Rees RW, Steinbrecher HA. Priapism in children: A comprehensive review and clinical guideline. J Pediatr Urol. 2014; 10: 11-24.

[2] Steinhardt GF, Steinhardt E. Priapism in children with leukemia. Urology 1981; 18: 604-6.

[3] Burnett AL, Bivalacqua TJ. Priapism: Current principles and practice. Urol Clin North Am. 2007; 34: 631-42.

[4] Montague DK, Jarow J, Broderick GA, et al. American urological association guideline on the management of priapism. J of Uro. 2003; 1318-24.

[5] Castagnetti M, Sainati L, Giona F, et al. Conservative management of priapism secondary to leukemia. Pediatr Blood Cancer. 2008; 51: 420-3.

[6] Rodgers R, Latif Z, Copland M. How I manage priapism in chronic myeloid leukaemia patients. Br J Haematol. 2012; 158: 155-64.