Management of Priapism with Chronic Myeloid Leukaemia- A Rare Presentation and Our Experiences

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Abstract

Background: Ischaemic priapism is one of rare presentation of Chronic myeloid leukaemia due hyper viscosity and it needs urgent urological intervention. Objective: To evaluate the need of urological intervention in Priapism with Chronic Myeloid Leukaemia (CML). Methodology: This is a retrospective study from January 2014 to July 2019 in the department of Urology, DMCH. Among 126 patients 6 were diagnosed as Priapism with CML. Records of these patients were evaluated and analyzed. Results: Among 126 patients 6 patients were diagnosed as priapism with CML Mean age of the patient was 46.7 years, 2 of them were farmer and 2 had history of taking herbal drugs immediate before sexual intercourse and developed Priapism. Five patients were newly diagnosed as CML after admission in Urology ward. Of these six cases five were managed with minimal invasive procedure such as intermittent pseudo-ephedrine injection and wide bore needle corporal aspiration with pseudo-ephedrine irrigation as emergency management, only one needed surgery that was proximal shunt. After emergency management all patients were sent to hematologist for definite management of CML. Conclusion: Priapism with CML is a medical emergency which need immediate minimal intervention with concomitant haematological management.

Key wards: Chronic Myeloid Leukemia, Priapism, Hyperleukocytosis, Proximal Shunt.

Introduction

Priapism in adult male patients with leukaemia is 1% to 5%. Among different causes, chronic myeloid leukaemia (CML) is one of rare causes of priapism and is a medical emergency. Most of the times priapism requires prompt evaluation and urgent urological intervention. According to American Urological Association guideline-2003, Priapism is defined as full or partial erection that continues more than 4 hours beyond sexual stimulation and orgasm or is unrelated to sexual stimulation. Ischaemia due to reduction in venous outflow by aggregation if leukaemic cells in the corpora cavernosa and the dorsal veins causes marked rigidity and pain are the key mechanism of priapism in CML. Time dependent changes occur in the corporal metabolic environment due to progressive hypoxia, hypercarbia, and acidosis in ischaemic priapism. Typical complains of penile pain after 6 to 8 hours and the examination reveals a rigid erection in this type of priapism. When priapism presents in the urology setting, evaluation and management of the predisposing condition must accompany interventions directed at the penis. We report our experiences who presented with priapism as the manifestation of chronic myeloid leukemia.

Materials and Methods

This is a retrospective study from January 2014 to July 2019 in the department of Urology, DMCH. Total 126 patients of Priapism were admitted during this period. Among them 6 patients were diagnosed as Priapism with CML and was included in this study. Records of
these six admitted cases of Priapism with CML were evaluated. Detailed history regarding age, occupational status, predisposing factors such as taking of any herbal drugs, history of having any previous diagnosed disease, duration of presentation at hospital after the penile erection were evaluated.

Investigations that were done during hospital admission such as complete blood count, peripheral blood count, colour Doppler study of penile vessels, cavernous blood gas analysis were analyzed. Six cases of priapism were found to have CML and they were referred to haematology department of Dhaka Medical College Hospital, Dhaka for further evaluation and management plan. Haematology department confirmed their diagnosis and advised treatment. Treatments taken immediately after admission and during the admission period were evaluated and analyzed.

Results

Among 126 patients 6 patients were diagnosed as priapism with CML and were included in this study. Mean age of the patient was 46.7 years, 2 of them were farmer and others were involved in different works. Among the 6 patients 2 had history of taking herbal drugs immediate before sexual intercourse and developed Priapism. Five patients were diagnosed as CML after admission in Urology ward, 1 was previously diagnosed. Three patients were presented within 24 hours and managed with assurance, painkiller and intermittent pseudo-ephedrine injection to corpora cavernosa. Other 3 patients were presented after 24 hours among them 1 needed proximal shunt surgery and 2 of them needed wide bore needle corporal aspiration with pseudo-ephedrine irrigation as emergency management. After emergency management of Priapism all patients were sent to Haematology department of DMCH for definite management of CML.

Table-1: Analysis of history of patients diagnoses as priapism with CML.

| Case Number | Age in years | Occupation       | Past Diagnosed Disease | Duration of presentation in hours |
|-------------|--------------|------------------|------------------------|----------------------------------|
| 1           | 45           | Rickshaw puller  | Herbal Drug            | 18                               |
| 2           | 58           | Farmer           | -                      | 48                               |
| 3           | 40           | Businessman      | -                      | 12                               |
| 4           | 38           | Worker           | Herbal Drug            | 29                               |
| 5           | 60           | Farmer           | CML                    | 6                                |
| 6           | 39           | Small Traders    | -                      | 72                               |
| Mean±SD     | 47±10        |                  |                        | 31±25                            |

Table-2: Immediate urological management.

| Case Number | Pain Killer | Proximal Shunt | Cavernosal Pseudo-Ephedrine | Wide bore Cavernosal aspiration and Pseudo-Ephedrine |
|-------------|-------------|----------------|-----------------------------|------------------------------------------------------|
| 1           | Needed      | --             | --                          | --                                                  |
| 2           | Needed      | --             | Needed                      | --                                                  |
| 3           | Needed      | --             | --                          | --                                                  |
| 4           | Needed      | --             | Needed                      | --                                                  |
| 5           | Needed      | --             | --                          | --                                                  |
| 6           | Needed      | --             | --                          | Needed                                              |

Discussion

Out of total incidences of priapism, etiologically only 1%-5% priapism is due to Leukaemia and in 20% cases etiology is malignant and non-malignant haematological conditions. CML accounts 50% of haematological condition causes ischaemic priapism. In our series, among 126 patients, we found 6 case of priapism is due to CML and it accounts only around 4% of total; which is similar with world-wide above findings. As, Ischemic Priapism due to CML is a urological emergency, it needs urgent multi-disciplinary intervention required within 4-6 hours to prevent erectile dysfunction. According to American Urological Association, European Urological Association of Urology and British guidelines, systemic treatment of underlying disorder like CML with urgent sequential urological intervention is necessary to prevent permanent erectile dysfunction. In CML induced priapism, after initial diagnosis and systemic evaluation, concurrent tyrosine kinase inhibitors (TKIs) or cytoreductive therapy with or without leukapheresis and intra-cavernous treatment is required. As ischemic priapism is a compartment syndrome it requires treatment directed at the penis primarily. Sequential urological intervention includes- initial conservative measures like intra-cavernosal aspiration under local anesthesia, cavernosal irrigation, intra-cavernosal therapy of adrenoceptor agonist like Phynylephrine and finally, surgical therapy like shunting or primary penile implantation after 36 hours of priapism. In our setting, 3 patients were presented within 24 hours and managed with painkiller and intermittent pseudo-ephedrine injection to corpora cavernosa. Rest 3 patients were presented after 24 hours and 1 needed proximal shunt surgery and 2 of them needed wide bore needle corporal aspiration with pseudo-ephedrine irrigation as emergency management.

Conclusion

Priapism is a urological emergency. Hyper leucocytosis causes low volume ischaemic Priapism requiring multidisciplinary approach with urologist and
haematologist working at tandem to clinch the diagnosis. The unusual presentation of CML needs to be borne in mind while dealing with cases of priapism.

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