Sickle Cell Disease with Recurrent Priapism

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Abstract

Sickle cell disease is a very common entity in central India, especially in Maharashtra. Patients usually present with recurrent haemolytic and/or vaso-occlusive crises. Although unusual, Priapism i.e. spontaneous painful erection of penis can be a debilitating manifestation of crisis. Especially when it occurs in prepubertal males it can lead to impotence in later life. It is also one of the least discussed complications of sickle cell disease. Here we present a case of sickle cell disease with recurrent priapism in a young boy of 16 years of age. He failed to respond to standard line of treatment. When all modalities failed he was given Tablet Gabapentin after which his priapism was totally relieved. We would like to recommend the use of Gabapentin in such cases of recurrent priapism.

Introduction

Priapism is an uncommon but debilitating complication of sickle cell disease (SCD). The prevalence has been reported as 8.2%. Majority of the patients have a single episode but recurrent episodes occur in about 30% patients. If such episodes occur in prepubertal stage, they may lead to impotence in the patient. Hence it is important to be aware of this complication in sickle cell disease patients. We report a case to emphasize this and review the therapeutic options for priapism.

Case Report

A sixteen year old male patient was admitted with complaints of painful erection of penis 2-3 hours prior to admission. Patient was a diagnosed case of Sickle cell disease (SS pattern). There was no history of fever, joint pains or bony pains. There was no history of chest pain or breathlessness. He did not have burning in micturition. He gave history of similar episodes of painful erections in past. The episodes lasted for 3-4 hours and sometimes resolved spontaneously. Twice he underwent blood drainage from corpora cavernosa. The frequency of episodes had increased in past 2 months. On examination the patient was in severe agony and pain. He was febrile, pulse rate – 110/min and respiratory rate of 16/min. He did not have pallor, icterus, joint swelling or sternal or bony tenderness. Systemic examination was normal. Local examination confirmed priapism. His investigations revealed haemoglobin of 9 gms%, peripheral smear showed sickled erythrocytes and ultrasound of abdomen revealed splenic infarcts. Penile Doppler showed venous obstruction with low flow. Rest of the investigations were normal. So priapism was probably a manifestation of vaso-occlusive crisis of sickle cell disease.

Initially he was treated with IV fluids, Tab. Diclofenac sodium 50 mg twice daily, Tab. Folic acid 10 mg daily and Cap. Hydroxyurea 500mg daily. He responded to this treatment and was asymptomatic for 20 days. Then again he started developing episodes of priapism. He was given a trial of Tab. Terbutaline 5 mg HS for 7 days and later Tab. Pseudoephedrine for 7 days but there was no relief. He was very afraid of corpora cavernosa blood drainage every time and was not willing for the same. In consultation with paediatric surgeon he was then started on Tab. Gabapentin 300 mg at bedtime. He responded to Gabapentin and has been symptom free for last 3 months.

Discussion

Sickle cell disease is a common disorder in this part of India and is associated with repeated episodes of vaso-occlusive crisis resulting in acute chest syndrome, splenic infarcts, avascular necrosis of head of femur etc. Amongst the least discussed complications is priapism which is painful erection of penis in the absence of sexual desire. Priapism in sickle cell disease occurs due to infarction of the venous outflow tract which results in increased blood volume in the paired lateral corpora cavernosa and single corpus spongiosum causing erection. The basis for this is stasis, hypoxia and acidosis of venous blood during normal erection resulting in sickling of erythrocytes within the venous sinusoids of corpora cavernosa thus causing obstruction of venous outflow. Priapism can be prolonged or stuttering and ischaemic or non-ischaemic. If it persists for longer duration it may lead to fibrosis and impotence. Penile Doppler with venous blood gas analysis can help to classify priapism.

Sharpsteen Jr et al. reported priapism in 8.2% sickle cell cohort. Death occurred in nine adult patients (25%) within 5 years of the first episode of priapism. They concluded that priapism in adult males identifies those at high risk for other sickle cell-related organ failure syndromes and, as such, is another complication indicative of severe disease.

Decisions for treatment of priapism are based extensively on diagnostic findings as the treatment for ischaemic and non ischaemic and stuttering priapism is different. In 60% of cases however embolization or surgery is advocated. Embolization is preferred, but if patient does not respond then as a last resort penile exploration with ligation of sinusoidal fistulas or pseudoaneurysm may be performed.

Definite first line treatment in ischaemic priapism consists of evacuation of blood from corpora cavernosa along with irrigation and injection of an alpha adrenergic sympathomimetic agent. If intracavernous treatment fails then surgical shunting should be tried, especially if priapism persists beyond 12 hours.
Drugs used in treatment of ischaemic priapism include terbutaline, pseudoephedrine, baclofen, bicalutamide and flutamide. Because of lack of consistent literature for use of oral agents, these medications cannot be recommended.2

Perimenis P et al tried Gabapentin in 3 patients with recurrent idiopathic priapism and patients responded in 48 hours.7 Our patient also responded well to gabapentin. As few workers think of priapism to have a neuromuscular pathogenetic mechanism. Gabapentin can be postulated to target this mechanism. Gabapentin can be given to patients with recurrent priapism.

Education, combined with more research of treatment options, may help patients avoid the damaging social, psychological, and medical implications of this bothersome and often embarrassing complication of SCD.

References


