CASE REPORT

Priapism due to essential thrombocythaemia: a rare causation

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SUMMARY

Priapism is rarely caused by essential thrombocytosis, a disorder characterised by increased number of megakaryocytes. We report a case of a 21-year-old man who presented with priapism and on investigation was found to have essential thrombocytosis as the cause.

BACKGROUND

Essential thrombocythaemia is an acquired myeloproliferative disorder that is characterised by a persistent elevation of platelet count with a high tendency to thrombosis and haemorrhage. It is related with expansion of megakaryocytic lineage and is considered to be a clonal disease arising from multipotent stem cell.¹ Haematological dyscrasias are a major risk factor for ischaemic priapism, of which primary thrombocythaemia is a rare cause. We report a case of a 21-year-old man who presented with a history of stuttering priapism for 2 years and presented with ischaemic priapism later, which was managed with repeated aspirations and instillation with phenylephrine, and was diagnosed with primary thrombocythaemia as underlying aetiology.

CASE PRESENTATION

A 21-year-old man presented with complaint of painful erection of penis for the last 6 days with history of similar episodes for the last 2 years, which resolved spontaneously. Pain was dull aching and increasing in intensity. There was no history of trauma or drug intake. Local examination revealed rigid, erect penis.

INVESTIGATIONS

Laboratory investigations revealed haemoglobin of 12.2 g/dL, white blood cell count of $17 \times 10^9/L$ cells/mm³ and platelet count of $830 \times 10^9/L$. Immediate corporal blood gas analysis revealed PO₂ of 28 mm Hg, PCO₂ of 65 mm Hg and pH of 7.23 suggestive of ischaemic priapism. Penile Doppler imaging was suggestive of arterial insufficiency with velocity of blood flow in cavernosal artery of <25 cm/s. Later, blood and bone marrow examination showed normocytic, normochromic type of global blood picture with increased megakaryocytes with hyperlobulated nuclei. Myeloid:erythroid ratio was 3.5:1 with normal granulocyte and erythroid precursor maturation. Haemoglobin electrophoresis revealed normal Haemoglobin A(HbA) type. Findings were suggestive of primary

thrombocythaemia. Further blood examination was positive for Janus kinase 2 (JAK2) V617F gene mutation and negative for BCR-ABL gene mutation. Ultrasonography abdomen showed mild splenomegaly.

DIFFERENTIAL DIAGNOSIS

The diagnoses were priapism due to essential thrombocytosis and stuttering priapism due to sickle cell disease.

TREATMENT

The patient was managed with repeat aspiration and instillation with phenylephrine (figure 1). After two sessions, priapism resolved. In consultation with the haematology department and as per guidelines, the patient was started on aspirin 75 mg tablet once a day and hydroxyurea 500 mg capsule twice a day.²

OUTCOME AND FOLLOW-UP

The patient was discharged in satisfactory general condition. At 6 months of follow-up, the patient was asymptomatic and had no history of repeat stuttering priapism. His haematological parameters were normal, with platelet count of 4.0 lac/mm^3 with normal haemoglobin and total leucocyte counts.

DISCUSSION

Priapism is a condition of full or partial erection of penis for more than 4 hours and is unrelated to sexual stimulation. It is a genuine urological emergency and should be managed immediately, as it can result in irreversible necrosis and fibrosis of corporal bodies, ultimately leading to permanent erectile dysfunction.³ Treatment options include therapeutic aspirations and intra-cavernous sympathomimetic drugs, followed by distal and proximal shunt procedures if necessary. Haematological dyscrasias are a major risk factor for ischaemic priapism, with various causes including sickle cell disease, thalassaemia, hereditary spherocytosis, paroxysmal nocturnal haemoglobinuria and so on.⁴ Our patient was diagnosed as a case of primary thrombocythaemia as the platelet count was $>4 \text{ lac/mm}^3$, haemoglobin was <16 g/dL, was JAK2 V617F gene mutation-positive and BCR-ABL gene mutation-negative, and bone marrow features suggestive of increased megakaryocytes, as per revised WHO diagnostic criteria.⁵ Since the discovery of JAK2 mutations in patients with essential thrombocytosis, these

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Figure 1 Intraoperative photograph showing penile aspiration and injection of phenylephrine.

markers have significantly improved the diagnostic criteria. Around 50%–60% of patients with essential thrombocythosis harbour JAK2 V617F mutation.⁶ Priapism is a rare complication of primary thrombocythaemia, and we could find only six such cases reported in the literature. We report a case of priapism in a young adult patient in which haematological work-up was suggestive of essential thrombocythosis; he was managed with aspiration and phenylephrine instillation in corporal bodies and treatment for essential thrombocythosis as advised by the haematological department.

Learning points

- ▶ Stuttering priapism should be considered as an alarm system before the dreaded occurrence of ischaemic priapism.
- ▶ Patients with stuttering/ischaemic priapism should always undergo a detailed haematological work-up to find its cause as haematological dyscrasias are a major risk factor.
- ▶ Early diagnosis and management of the underlying haematological condition is a paramount prophylactic measure to prevent ischaemic priapism.
- ▶ Early and effective management is the key to avoid irreversible loss in ischaemic priapism.

Contributors All of the authors declare that they have participated in the design, execution and analysis of the paper, and that they have approved the final version. SG: Concept, design, supervision, processing, writing the manuscript and critical analysis. AKS: Supervision, processing, writing the manuscript and critical analysis. BPS: Concept, design, supervision, processing, writing the manuscript and critical analysis. AA: Concept, supervision, writing the manuscript and critical analysis.

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