Priapism in CML

Sir

CML is a common leukemia among adults in India. Uncommon presenting symptoms of CML include tinnitus, diplopia, papilloedema and priapism and may be due to leukostasis associated with greatly exaggerated blood leukocyte count. About 20% of all cases of priapism are due to various hematological disorders such as Sickle cell anemia, CML, AML or ALL.¹

CASE: A, 55 year old male presented with complain of continuous, painful erection of penis for 2 days. On further questioning patient complained of generalized weakness and fullness of abdomen for last 4 months. He denied history of any drug (esp. antihypertensive, antipsychotics and any herbal medicine) / alcohol intake. On examination. Patient had sternal tenderness. spleen was enlarged up to umbilicus and liver enlarged, 2 cm below right subcostal margin. Penis was erect, stiff and painful. Investigations: Hb 9.5 g, WBC 4,20,000/mm³ differential-polymorphs 58% lymphocyte 8%, basophils 6%, eosinophils 1%, immature cells 27% (promyelocytes 6%, metamyelocytes 16%, myelocytes 5%), Platelet 2,80,000/mm³. Urine examination, Renal and Liver function test were normal. S. uric acid 7 mg%. Patient under went shunt surgery. following which he unproved, presently, he is on hydroyurea with close monitoring of blood counts.

COMMENTS

Approximately 20% of priapism cases are related to the hematologic disorders. Leukemia accounts for 1 to 5% of these.
Priapism can be low flow (ischemic) or high flow (non ischemic) type. Low-flow priapism results from decreased penile venous outflow causing stasis and presents as a painful, rigid erection. More common than high-flow priapism, low-flow priapism is a medical emergency because irreversible cell damage and fibrosis can occur if treatment is not initiated within 24-48 hours. Low-flow priapism can be drug induced or caused by hematologic disorders (i.e. Sickle cell anaemia, thalassemia and Leukemias) and tumour infiltration.2

High-flow priapism results from increased arterial inflow into the cavernosal sinusoids, which overwheels venous outflow. Clinical presentation is painless erection; irreversible cellular damage and fibrosis are rare. High-flow priapism often is the result of penile or perineum trauma and is not an emergency, treatment is elective.3

First-line treatment of priapism is aspiration of blood from the base of the corpora cavernosa. The success rate with aspiration alone is approximately 30%. If the treatment is unsuccessful, instillation of the sympathomimetic agent phenylephrine hydrochloride (250-500 µg) every five minutes is used until the swelling of the penis has reduced, or subsided, (the Phenylephrine solution is made by mixing 10 ml/ml of phenylephrine with 19 ml of normal saline).4 In cases of priapism related to hematologic malignancies, leukapheresis or cytotoxic therapy such as Hydroxyurea, Imatinib may be used to reduce the number of circulating white blood cells. If medical management fails, surgery can be considered. This includes placing a shunt between the corpora cavernosa and glans, which allows for blood to flow in and out of the corpora cavernosa.5

REFERENCES:


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