

A CASE OF HB-SC DISEASE PRESENTING AS PRIAPISM

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Priapism is a common symptom in Haemoglobin SC and SS disease, which results from thrombosis in the vascular spaces in the cavernous tissue of penis. Thrombosis is due to altered physico-chemical properties of HbS which has glutamic acid substituted for valine in the sixth position from N-terminal in Beta Chain. When deoxygenated, HbS is less soluble and tactoids form in red cells which cause distortion and damage to the red cell membrane. Sickled red cells become rigid and mechanically fragile resulting in haemolysis, repeated vascular occlusions and infarction of various interbak organs like kidneys, mesentery, intestine, lungs, brain and bones.

CASE REPORT

A 30 years old Balochi farmer from Loralai was admitted with priapism of 7 days duration. There was no history of dysuria, haematuria or trauma. On clinical examination he was slightly pale; spleen, liver, lymph nodes and kidneys were not palpable and there was no other abnormality. On investigation, Hb-10.2 G/dl; total and differential leucocyte count, reticulocyte count, platelet count and blood urea were normal. The peripheral blood did not show any malarial parasite. When a drop of blood was sealed under a cover slip on a slide, red cells acquired sickle shape after 24 hours. Sodium metabisulphate test was not done. Paper electrophoresis revealed a distinct band of HbS and C.

DISCUSSION

In this rare case, HbSC was responsible for priapism. Other cases of priapism described are leukemias (both myeloid and lymphatic), new growth of urethra, primary and secondary carcinoma of bladder or testes, sickle cell anaemia, trauma to penis, or injury to upper dorsal region of spinal cord¹. Frequent priapism, sexual impotence and reduced fertility has been reported as a complication in sicklers from African subcontinent².

Neuropsychiatric symptoms like delirium and temporal lobe epilepsy or grand mal epilepsy may also be seen³. Sudden, unexpected deaths are not uncommon in SS or SC disease and are due to superimposed bacterial, viral or malarial infections⁴. The mothers with SC disease behave normally during pregnancy⁵.

Treatment is mainly conservative in the form of use of sedatives or analgesics and correction of fluid and electrolyte balance.

The case reported above was treated on conservative lines and his priapism was relieved within a week.

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