CORRESPONDENCE

Priapism Associated with Homozygous Hb E State: A Causal Association or an Incidental Finding?

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Dear editor,

Priapism, that is, persistent penile erection that continues hours beyond, or is unrelated to, sexual stimulation, is a relatively uncommon disorder and is a medical emergency. Typically, only the corpora cavernosa are affected [1]. The term priapism was derived from the Greek god Priapus, son of Aphrodite who was born with oversized genitals [2]. The haematological causes for priapism include Sick cell anaemia, Leukaemia, Multiple myeloma, Paroxysmal nocturnal haemoglobinuria, Thalassemia, Thrombocythemia and Henoch–Schönlein purpura [3]. As far as hemoglobinopathies are concerned, priapism is most often encountered in sickle cell disease in 38–42 % of cases followed by sickle/beta⁺ thalassemia [4, 5]. Priapism is also noted in patients with sickle cell trait even though the incidence is low as compared with sickle cell anaemia [6–9]. However, so far priapism in Hb E homozygosity is not reported in literature. In this correspondence we intend

to bring to notice an unusual association of priapism with Hb E homozygosity.

A 27 years old male, resident of North Eastern part of India was referred to our haematology OPD to rule out any underlying haematological disorder as the individual had an episode of priapism 3 months ago. The patient had an episode of unprovoked painful persistent erection for more than 8 h for which he sought medical advice. The surgeon at that medical center had managed with therapeutic needle aspiration from corpora cavernosa combined with flushing of cavernosa with normal saline to clear the sludged blood. The patient's symptom subsided subsequently with the intervention and was referred to our center for further evaluation. On detailed clinical history, the individual was not on any medication for chronic illnesses; neither was he abusing any psychoactive drugs, alcohol, no history of prior trauma to the perineal region. His physical examination revealed mild pallor. There was no hepatosplenomegaly or lymphadenopathy. Systemic examination did not reveal any abnormality. On USG abdomen, spleen was not reported as enlarged (Span 13 cms).

On investigations, haemoglobin, total leucocyte count and platelet count were 9.1 gm/dL, $7.1 \times 10^3/\mu\text{L}$ and $118 \times 10^3/\mu\text{L}$ respectively. Peripheral smear examination revealed microcytic hypochromic red cells and target cells with normal differential leucocyte count and reticulocyte count of one percent. There were no sickle cells or atypical cells in peripheral smear and sickling test was also negative. Subsequently Hb HPLC was performed which revealed with Hb A₂ + E of 92.6 % (Retention time—3.68 min), Hb A of 6 % (Retention time 2.29 min) and Hb F (Retention time 1.06 min) of 1.6 % (Fig. 1; Table 1). This was followed by parental study which revealed Hb A₂ + E in mother and father of 29 and 27 % respectively suggestive of both parents being heterozygous for Hb E,

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SAMPLE ID# 000000000000000000876			
ANALYTE ID	%	TIME	AREA
F	1.6	1.06	27429
P3	5.0	1.84	88956
Unknown 1	0.9	2.05	16105
Ao	6.0	2.29	105574
A2	92.6	3.68	1640975
TOTAL AREA			1879039
F	1.6%	A2	92.6%

Fig. 1 Hemoglobin chromatogram of the patient suggestive of Hb E homozygosity

Table 1 Hematological parameters of patient and parents

Parameters	Patient	Father	Mother
Hemoglobin (g/dL)	9.1	14.0	10.8
Total leucocyte count ($\times 10^3/\mu\text{L}$)	7.1	10.81	11.16
Platelet count ($\times 10^3/\mu\text{L}$)	118	133	173
Hematocrit (%)	34.2	40.2	34.2
MCV (fL)	59.7	76.3	79.5
MCH (pg)	16.0	26.6	25.1
MCHC (g/dL)	26.8	34.8	31.6
RDW-CV (%)	18.9	15.4	15.5
RBC count ($\times 10^6/\mu\text{L}$)	5.93	5.27	4.3
Hb F (%)	1.6	0.7	1.0
Hb A0 (%)	6	67.6	64.6
Hb A2 + Variant (%)	92.6	27	29

hence a diagnosis of Hb E homozygosity was made in the patient.

Colour Doppler USG did not reveal any vascular abnormalities, such as a cavernous artery fistula or pseudoaneurysm. In the absence of any other cause and the

available evidence of Hb E homozygosity, this case would be the first in literature in which Hb E homozygosity is associated with priapism. The patient is presently asymptomatic and is on follow up.

Priapism has an incidence of 1.5 per 100,000 and can occur in all age groups from newborn to elderly [10]. Typically, there is a bimodal peak of incidence, between 5 and 10 years in children and 20–50 years in adults. Sick cell disease is the commonest aetiology in childhood while pharmacological agents and myeloproliferative disorders are associated with many cases of priapism in adults. Types of priapism include ischemic (low flow), non ischemic (high flow) and stuttering type of which the ischemic type is a medical emergency which can result in fibrosis of corpora and erectile dysfunction. There are some uncommon aetiologies for priapism described in literature which include glucose phosphate isomerase deficiency, Fabry's disease, a high concentration (i.e., 20 vs 10 %) of fat emulsion in total parenteral nutrition, or with paradoxical thromboembolic events associated with heparin or warfarin use [11]. The possibility of our patient simply having a chance co-occurrence of two pathologies cannot be entirely excluded. HbE trait and homozygosity are extremely common in north-east India with Baruah et al. reporting EE-state in 21 % of 9,000 patients screened in upper Assam and Bhattacharyya et al. reporting 19.2 % of tribal Totos in West Bengal to be E homozygotes [12, 13]. Ideally, a pro-thrombotic state work-up would also be indicated in our patient.

To conclude, the association of Hb E homozygosity with priapism is not described so far. Even though we could not hypothesize the underlying pathogenetic mechanism causing priapism in Hb E homozygosity, it is an interesting and unique finding which is worth reporting.

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