CASE PRESENTATION

Priapism in Two Children with Sickle Cell Disease at Qateef Central Hospital

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ABSTRACT

Two Saudi children with sickle cell disease were admitted at Qateef Central Hospital with sickle cell crises. The first patient from south-west of Saudi Arabia, was admitted with severe abdominal crisis. The second patient was from Eastern Province, was admitted with major sequestration crisis. Priapism was diagnosed in both patients after admission. Both of them were successfully treated conservatively. The possibility of this complication should be borne in mind when a child with sickle cell disease is presented in a crisis.

Priapism is a painful persistent penile erection without sexual excitation and is due to engorgement of corpora cavernosa.[1] Although the prevalence of priapism is reported to be as high as 40-50 percent in patients with sickle cell disease,[2,3,4] early report by Perrine RP, et al from Eastern Province of Saudi Arabia revealed only one case of priapism among 130 male patients with sickle cell disease, the age of which was not mentioned.[5] Al-Awamy, et al, recently reported no evidence of priapism among 42 patients with sickle cell disease aged 3-42 years.[6] However, 2 adult cases have been reported more recently.[7] Despite the fact that the disease in south-west of Saudi Arabia is considered more severe, no cases of priapism have been reported from that area.[8,9]

We report these two cases of priapism to indicate the presence of this complication in Saudi children with sickle cell disease, to highlight the possibility of under diagnosis and successful conservative management. To the best of our knowledge, these are the first two reported cases of priapism in Saudi children with sickle cell disease.

THE CASES

Patient 1

A 4 year old male from south-west of Saudi Arabia, admitted through emergency room with a complaint of severe abdominal pain for 24 hours. He was a known case of sickle cell disease, diagnosed and followed up in our hospital. No history of fever, vomiting or urinary symptoms and no history of trauma. There was a past history of three previous admissions to our hospital with vasoocclusive crises and one admission with *H.Influenza Septicaemia*. On physical examination, the patient was alert, but in pain. Heart rate 90 per minute, respiratory rate 25 per minute, temperature 36.5^{0}C and blood pressure 110/70 mmHg.

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Table 1
The results of relevant laboratory investigations for both patients

Haematological parameters	*Patient 1*	*Patient 2*
WBC, x 10^9/1	22	15
RBC, x 10^{12}/1	2.58	00.67
Hb, gm/dl	5.8	1.3
Hct, %	20.1	5.1
MCV, fl	77.9	75.9
MCH, pg	22.5	19.4
MCH, gm/dl	28.9	25.6
Platelets, x 10^9/1	245	55
Reticulocytes, %	19	—
HbS, %	83	69.1
HbF, %	17	29.5

Abdomen was distended with moderate generalised tenderness. No organomegally was noted and bowel sounds were normal. No other physical abnormalities was recorded. Genitalia were not examined by the admitting paediatric resident. The results of relevant laboratory investigations are summarised in Table 1.

Patient was admitted as a case of sickle cell disease with abdominal crisis. When the consultant reviewed the case, penile erection was noted, and a review of the history revealed that this had been present before admission. Patient was treated initially with hydration and analgesia, but priapism failed to respond to this treatment. Partial exchange transfusion was done after which detumescence began within 24 hours. Post transfusion haemoglobin "S" level decreased from 83 percent to 26 percent.

Patient 2

A 7 year old male from Eastern Province of Saudi Arabia, admitted for the first time through the emergency room with a complaint of fever for three days, sudden pallor, vomiting and abdominal pain for one day. No history of urinary symptoms and no history of trauma. He was not known to have sickle cell disease but he had two siblings with the disease. On physical examination, the patient was restless with severe pallor. Temperature was 36^0C, respiratory rate 52 per minute, heart rate 160 per minute and blood pressure 96/40 mmHg. Abdomen was distended with moderate tenderness. Spleen was palpable 6cm below left costal margin. No other physical abnormalities was recorded. Genitalia were not examined by the admitting paediatric resident. The results of relevant laboratory investigations are summarised in Table 1. Patient was admitted as a case of sequestration crisis and urgent blood was requested. When the consultant reviewed the case, penile erection was noted. After blood transfusion, haemoglobin level was raised gradually from 1.3g/dl to 10.3g/dl and haemoglobin "S" level decreased from 69.1% to 16.3%. Detumescence began within 24 hours after blood transfusion.

DISCUSSION

Priapism has been reported in the paediatric age group as early as neonatal period.[10] Paediatric patients most often had priapism owing to sickle cell disease.[11] Although sickle cell disease is common in Eastern Province of Saudi Arabia,[12] priapism was rarely reported in adults and not reported in children.[5,6,7] This rarity was ascribed to high level of HbF and co-existing alpha-thalassaemia.[6,7]

Due to social reasons, many patients in our area are reluctant to discuss the genitalia, and may hide symptoms from their doctors. Some doctors are also reluctant to ask about the genitalia in symptomatic inquiry and even ignore their examination. These reasons may contribute to the rarity of reported cases of priapism in our area.

In these two cases, priapism was not diagnosed at admission, because failure to examine the genitalia. Therefore, we should emphasise the importance of genital inspection in patients presenting with sickle cell crises. On the other hand, Saudi patients with sickle cell disease and parents of affected children should be educated about this condition and the need for early intervention to prevent complications like impotence.

In both patients, haemoglobin "S" was decreased to less than 30 percent, after which detumescence began within 24 hours. This drop in haemoglobin "S" was attained by an exchange transfusion in patient 1, however in patient 2, it was attained by infusion relatively large volume of packed red blood cells, given to correct the severe, life-threatening anaemia. This approach of conservative management should be considered in cases of priapism before surgical intervention.

CONCLUSION

Priapism may be under-diagnosed in Saudi patients with sickle cell disease. It should be suspected in patients with sickle cell disease, presenting with abdominal pain. Conservative management should be attempted before surgical intervention.

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