ORIGINAL

Pattern of Admissions of Children with Sickle Cell Disease in Qateef Central Hospital, Saudi Arabia

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ABSTRACT

Sickle Cell Disease (SCD) in the Eastern Province of Saudi Arabia is thought to be a benign form of disease. This is not true for all patients and it contradicts our clinical observation. Therefore, we conducted this study to look at the patterns of different presentations of SCD patients who required admission to Qateef Central Hospital (QCH). The study was conducted prospectively between March 1990 and July 1991. All SCD patients less than 13 years were included. The total number of patients admitted to the paediatric medical ward during the study period was 2886, out of which 561 were SCD patients (19.4%). Out of these, painful crises were responsible for 298 admissions (53%), anaemic episodes for 204 admissions (36.4%), infection for 83 admissions (15%), acute chest syndrome for 28 admissions (5%) and others for 20 admissions (3.6%). The SCD patients form a large proportion of paediatric admissions. They present with various complications of the disease.

Sickle Cell Disease is a chronic haemolytic disorder, manifested by a wide variety of clinical and haematological features. Some patients are entirely asymptomatic and are detected only during population screening, whereas others constantly develop painful episodes and other complications. SCD is a common problem in the Eastern Province of Saudi Arabia with a carrier rate of 20%[1]. The disease is thought to be a benign form in this area[2]. This is not true for all patients and it contradicts our clinical observations. Therefore we conducted this study in QCH to look at the pattern of different presentations of SCD paediatric patients at the time of admission to the hospital.

METHODS

A prospective study was conducted at QCH from March 1990 through July 1991 (18 months). All

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paediatric patients (< 13 years) with SCD (Homozygous and SB-thalassaemia) admitted during this period were included. The diagnosis of SCD was made on the basis of clinical as well as positive sickling test and Hb-electrophoresis using Helena Laboratories Super Z Electrophoresis Kit. Painful crisis is defined as an episode of pain in the extremities, back, chest and abdomen. Anaemia is defined as a reduction of haemoglobin concentration below the steady state values[3]. Acute sequestration crisis is defined as acute fall in haemoglobin associated with an increase in spleen size sufficient to cause clinical presentation[4]. Infections are divided into major and minor. Major infections are osteomyelitis, septicaemia, septic arthritis and splenic abscess. Acute chest syndrome is defined as a combination of fever, chest pain, leukocytosis, clinical and or radiological evidence of lung abnormalities[5].

RESULTS

The total number of paediatric admissions to paediatric medical ward during the study period was 2886, out of which 561 were patients from the Eastern Province with SCD (316 patients). This accounted for 19.4% of total paediatric admissions. SB°-thalassaemia was diagnosed in 33 admissions, whereas the rest of SCD admissions were patients with SS pattern. There were 325 males and 236 females with a ratio of 1.4:1. Most patients were admitted once during the study period (220 patients). Fewer patients required more frequent admissions (Fig 1).

Painful crises (Table 1) were the cause for 298 admissions (53%). These admissions occurred more frequently in older age group. The commonest site for painful crises were the limbs, which accounted for 112 admissions (37%). The other sites are shown in Table 2. Upper respiratory tract infection was associated with 26 admissions (8.7%). Other precipitating factors of painful crises were trauma in 9 admissions (3%), physical exertion in 3 admissions (1%) and emotional disturbances in 3 admissions (1%).

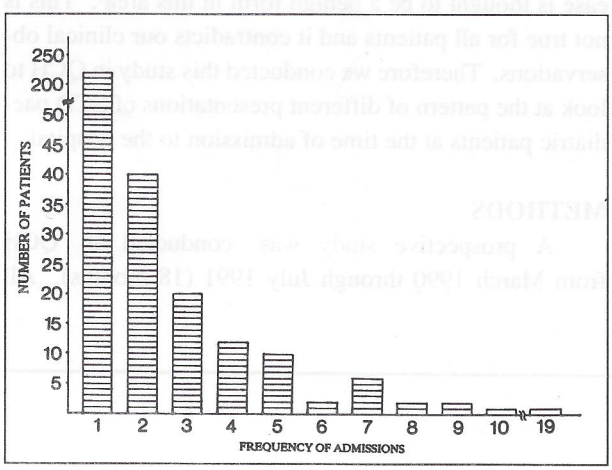

Figure 1

Table 1
Causes of Admissions

Causes	No. of Admissions	%
Painful crises	298	53
Anaemias	204	36.4
Infections	83	15
Acute Chest Syndrome	28	5
Others	20	3.6

Table 2
The site of painful crises

Site	No	%
Limbs	112	37
Back	080	27
Abdomen	060	20
Joints	036	12
Generalised bodyache	027	9
Chest	008	3
Mandible	001	1

Out of 204 admissions for anaemic episodes, 97 (47.6%) were for acute haemolytic crises, out of which 39 with confirmed G6PD deficiency, while in others the cause of haemolysis was not known. In 42 admissions (20.6%), patients were admitted for hypertransfusion regimen (36 SB°-thalassaemia and 6 hypersplenism). Anaemia was associated with acute splenic sequestration crises (ASSC) in 28 admissions, infections in 19, aplastic crises in 8, acute chest syndrome in 5 and severe epistaxis in 5.

Major infections were found in 30 admissions; 19 osteomyelitis, 8 septicaemia, 2 septic arthritis and one splenic abscess. In cases of osteomyelitis, 9 patients had salmonella. In one patient, gram stain showed gram negative bacilli while pus culture was negative. The rest of patients with osteomyelitis (9 patients) had clinical and

radiological evidence of osteomyelitis while pus culture was negative. In septicaemic patients, 5 had salmonella, 2 had brucella and one who had splenectomy grew staphylococcus epidermidis. In patients with septic arthritis, gram stain showed gram positive cocci in one patient, and pus culture was negative in both patients.

Other infections were upper respiratory tract infection in 54 admissions (46 viral, 5 B-haemolytic streptococci and 3 candida albicans), viral gastroenteritis in 4, scarlet fever in 2, quinzy in 2, dental abscess in 1 and chicken pox in 1. In 28 admissions (5%), the diagnosis of acute chest syndrome was made.

Central nervous system complications were found in 6 admissions; 3 convulsions, 2 confusional state and 1 coma. Convulsions were in the form of generalised tonic clonic state and was associated with fever in one patient. CT scan was done in the three and revealed no abnormalities. Confusion was associated with severe lower limbs painful crisis in one patient and salmonella septicaemia in the other. Coma was found in patient with major sequestration crisis and severe anaemia (Hb 1.9 gm/dl). This patient continued to be unconscious for 8 days, after which he recovered with residual central nervous system sequalae.

Cardiovascular complications were found in 4 admissions; 2 congestive heart failure, 1 premature ventricular contractions and 1 deep vein thrombosis. Congestive heart failure was associated with severe anaemia in the two admissions. Premature ventricular contraction (pulsus trigeminus) were associated with a painful crisis and disappeared after recovery of pain. Deep vein thrombosis occurred in a patient with nephrotic syndrome.

Renal complications were found in 4 admissions. Two patients had polyuria with dehydration. Nephrotic syndrome (first relapse) was found in one admission and haematuria in another.

Major type of priapism was found in 3 admissions. It was associated with a major attack of acute splenic sequestration crisis in one admission and severe abdominal pain in the other two.

DISCUSSION

SCD in the Eastern Province of Saudi Arabia is thought to be a benign form of disease and rarely required admissions[2,6]. This is not supported by this study, which shows that SCD patients presented with various complications and formed almost 20% of total paediatric admissions in QCH. Some of these patients required frequent admissions during the study period (18 months) up to 19 times. This high frequency of admissions further indicates the magnitude of the problem of some of our SCD patients and the effect on daily living activity of these patients and their families.

The type of complications leading to clinical presentation are similar to those reported in Western Saudi Arabia and other populations[7-11]. They presented with almost all known acute complications of the disease.

The most common reasons for admission were painful crises (53% of all admissions) as was found in other studies[6,10,12]. The frequency of these admissions increased with increasing age group. This may indicate that the older the patient the more prone he/she to the precipitating factors of these complications.

The next common reasons for admission were anaemias (36.4% of all admissions), a finding which contradicts some reports from the same area[6]. Acute haemolysis occurred in about 50% of anaemic episodes, but G6PD deficiency was confirmed only in 50% of acute haemolysis, a finding which may support the idea that haemolytic crises may occur in patients with SCD without concomitant G6PD deficiency[6,12,13].

The finding of acute sequestration in 28 admissions (5% of all admissions) contradicts other reports from the same area in which this complication was rarely reported[1,6]. Awareness of this complication and regular measurement of splenic size during follow-up visits may explain this increased reported cases.

Infections were an important cause of morbidity, leading to 15% of admissions (83 admissions). Salmonella infection was the leading cause of septicaemia and osteomyelitis. Although pneumococcal infection had been implicated in SCD patients, none of our patients had this infection in this study[14]. An earlier report from this hospital (QCH) revealed that pneumococcal infection was a cause of significant morbidity[15,16]. The discrepancy between this study and earlier studies can be explained by recent use of pneumococcal vaccines and penicillin prophylaxis in our SCD patients. This prophylactic therapy was found to be very effective in the reduction of pneumococcal infection[14,17].

The presence of confusional state in a patient with severe painful crisis involving lower limbs should alert to the possibility of fat embolism. High index of suspicion of this possibility should be emphasised for early detection and proper management.

CONCLUSION

This study shows that SCD in Eastern Province of Saudi Arabia is not benign in all patients. Many patients suffered from complications of this disease. A special program is badly needed to take care of many patients suffering from the disease. This program should include neonatal screening for early diagnosis of sickle cell disease and appropriate management. Availability of sickle cell centre will optimise the acute and chronic care of these patients.

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