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Sickle Cell Anaemia, A Study from the Capital Area of Oman

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One hundred and six Omani children with sickle cell anaemia (SCA) aged between 10 months and 16 years were studied by reviewing their medical notes and following their clinical course. All the cases were diagnosed on clinical presentation. Eighty-five percent were diagnosed below the age of 3 years. The clinical presentations and complications were compared with studies from Saudi Arabia and some other tropical countries. The frequency of hospitalization due to the complications of the disease is higher than that reported in other parts of Arabian Peninsula. The incidence of serious complications such as vaso-occlusive, aplastic, hemolytic and sequestration crisis were high (91%, 1.9%, 59%, 6.7% respectively). Infection is more frequent. However, pneumococci were not the commonest isolated organism. G6PD deficiency was reported in 32% of cases which might explain the higher incidence of hemolytic crisis. Our study shows that SCA has a severe clinical course in Omanis. Because of the intermarriage of Omanis with Africans and Arabs, the nature of the SCA gene needs to be identified in this population. Bahrain Med Bull 1995;17:

Sickle cell anaemia SCA is prevalent throughout many parts of the world particularly in tropical Africa, the Middle East, the Mediterranean and parts of India and America[1,2]. This disease constitutes one of the most frequent causes of hospitalization of children in Oman. The purpose of this study was to evaluate the epidemiology and clinical presentation of SCA in Omani children.

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METHODS

The study represented a combined retrospective and prospective studies of all of 106 children with SCA seen at the outpatient clinics and/or admitted to the Royal and Sultan Qaboos University Hospitals in Muscat, the capital of Oman during 1992. The medical records of the patients were reviewed for the clinical presentations, complications and laboratory investigations.

Routine hematological parameters were measured on a Coulter counter model S plus. Sickledex screening test (Ortho diagnostics) was used for sickle screening. Hemoglobin electrophoresis was done for all sickle cell positive samples using Gelman agarose gel at pH 8.6 (barbitone buffer) and scanning of the electrophoretic strip. Hemoglobin S and Hemoglobin F were quantified on the scanner[3]. It was not possible to rule out sickle cell/beta thalassemia with absolute certainty as globin chain synthesis could not be done at that time. However all patients with SCA with low MCV and/or high hemoglobin A2 had the diagnosis confirmed by family study. Glucose-6-phosphate dehydrogenase (G6PD)

activity was determined in fresh hemolysate using commercially available kits from Boehringer Mannheim GmbH.

RESULTS

All of the 106 patients in this study were diagnosed on clinical presentation. The patients had equal sex distribution. Of these 76 (72%) lived in Muscat, the capital area while 30 (28%) patients were referred from other regions. The age range was between 10 months to 16 years old with a mean age of 2.1 years old (Table 1).

Table 1
Comparison of age at first diagnosis of patients with Sickle cell anaemia

Age	Oman Capital Area 1992		Saudi Arabia West & South-West Areas 19914	University College Hospital Ibadan Nigeria 19825
	No	%	%	%
< 12 months	38	35.8	33	9.9
13-36 months	52	49.1	40	32.9
4-6 years	11	10.4	21	25
> 6 years	5	4.7	5	31.4

Forty-two (40%) patients had positive family history with two affected family members in 64%, three affected members in 34%, and more than three in 2% of cases.

At the time of diagnosis the hemoglobin level of < 6 gm/dl was found in 7 (6.6%) patients, a level between 6 to < 9 gm/dl in 78 (73.6%), and between 9 and 12 gm/dl in 20 (18.8%) patients. Only one (1%) case had hemoglobin of 14 gm/dl. The mean percentage of HbS was 72.9% (Table 2).

Table 2
Hemoglobin levels in patients with SCA

Variable	Mean	S D	Minimum	Maximum
HB (gm)	8.2	1.5	3.4	14.0
HBS (%)	72.9	16.6	20.4	98.3
HBF (%)	10.8	8.5	0.2	35.0
HBA2(%)	3.4	0.9	1.6	5.5

About 77% of cases (82 patients) were hospitalized. Forty-one patients (39%) had 2 to 4 hospitalizations per year. Only 48 (45%) patients received blood transfusion, 9 (20%) of whom were transfused more than three times per year.

Table 3
Clinical presentation of children with SCA in Oman as compared to those of Eastern Saudi Arabia6 and Jamaica8

Clinical picture	Oman		Eastern Saudi Arabia	Jamaica
	No	%	%	%
Vaso occlusive crisis	96	91		
Hand-foot syndrome	41	43	14	40
Painful crisis	96	90	21	18
Abdominal crisis	85	80	5	
Acute splenic sequestration	7	7	1	25
Aplastic crisis	2	2	2	9
Hyperhemolytic crisis	63	59	2	-
Hypersplenism	6	6	1	-
Splenomegaly	40	38	33	80
Hepatomegaly	25	24	21	-

Table 3 shows the clinical presentation of patients with SCD. Vaso-occlusive crisis occurred in 96 (91%) patients of whom 41 (43%) had Hand-foot syndrome, 96 (90%) painful crisis and 85 (80%) abdominal crisis. Acute sequestration crisis was seen in 7 (7%) patients, four of whom were below 2 years of age and the other were between five and ten years. Aplastic crisis was seen in 2 (2%) patients. Hyperhemolytic crisis occurred in 63 (59%) patients. Of the 106 patients in this study 34 (32%) patients also had G6PD reduced activity. Only two patients had cerebro-vascular accidents with subsequent neurological handicap. Biliary stones were also found in 2 (1.9%) patients.

Severe infection that required hospitalization had been registered in 22 (21%) patients. Thirty-nine episodes of infection have been reported. Six patients had more than one episode. Among these infection episodes pneumonia was seen in 17 (55%) patients, septicemia in 9 (23%) and CNS and urinary tract infections in 5 (16%) patients.

Pneumococci were isolated from two septicemic patients. Klebsiella, E. Coli and pseudomonas were found in the others. The two patients with pneumococcal septicemia were below 5 years of age. One of them died from septic shock, and the second developed meningitis which was complicated with hydrocephalus and severe motor disabilities.

DISCUSSION

All the patients in this study were diagnosed on clinical presentation because there were no screening programme available at the time of the study in 1992. Eighty-five percent of patients were diagnosed before the age of 3 years old. This may reflect the severity of the disease in early life. The figures for West and Southwest Saudi Arabia in Nigeria were 73% and 43% respectively[4,5] (Table 1).

The morbidity of SCA can be estimated from the frequency of patient hospitalization. The hospitalization rate of 77% in this study is higher than the 20% reported from Eastern Saudi Arabia[6]. The rates for black American and Jamaican patients were less than the Saudi figure[7,8]. However, such comparison does not take into account the different admission policies of the hospitals.

In addition, the difference may be due to parental concern, socio-economic status, hospital access etc.

Vaso occlusive crisis was the commonest cause of hospitalization in Oman (91%), a very high incidence compared to Saudi Arabia, Jamica, South London and Africa[4,6,8,9,10,11.] Ninety percent of these crisis involved bones, a very high figure compared to Eastern Saudi Arabia6 and Jamaica[8]. The incidence of dactylitis was also higher in our study. Abdominal crisis occurred in 80% of our cases compared to 5% in Saudi patients[6] (Table 1). This may be considered as an evidence of the disease severity. In two American studies, lung disease was reported as the commonest cause of admission[7,12]. Only seven (6.6%) patients in this study were admitted due to acute chest syndrome.

The relatively high incidence of hyperhemolytic crisis (59%) compared to Saudi Arabia may be explained by the reported high incidence of G6PD deficiency which was found in 32% of our cases[4,6]. Splenic sequestration crisis occurred more frequently compared to Eastern (1%) and Southwest areas of Saudi Arabia (2.2%)[4,6]. Twenty-five percent of SCA patients in Jamica had sequestration crisis[8]. This may exist as an example of variable clinical presentation of the disease in different ethnic groups. The incidence of aplastic crisis in this study is more than that reported in Eastern Saudi Arabia but markedly lower than the incidence in Jamaica[6,8].

The most frequent reported infection was pneumonia followed by septicemia. The frequency of infections in our cases were higher than those reported from Eastern Saudi Arabia but as severe as in black American and Jamaican studies[6-8]. The incidence of pneumococcal meningitis in Saudi patients with SCA was 7% in one study giving an attack rate of 12.8 per 1000 patients years while in another study from Dhahran it was found to be 1.48 per 1000 patients years[4'13]. Pneumococci were isolated from two septicemic cases. Klebsiella, E. Coli and Pseudomonas were found in the other.

A change in the pattern of septicemia in SCA following pneumococcal vaccine has been observed and Haemophilus influenzae found to be the predominant organism[14]. The lower incidence of pneumococci as a causative organism of septicemia in our cases may be due to the policy of giving pneumococcal vaccine and penicillin prophylaxis at the time of diagnosis. This agrees with Bailer's study who found that Gram negative organisms are the most common isolate from septicemic patients with SCA[15]. The results regarding this association from two different regions of Saudi Arabia were different. Although there was no significant increase in the rate of infection between Eastern Saudi patients with SCA when compared to a control group, a high incidence of severe infection had been reported in Southwestern Saudi Arabia[4,6]. Our patients may be similar to those of the Southwestern Saudi region and this requires further study.

Blood transfusion in SCA should be done only as absolute necessity. Its indications include splenic sequestration, aplastic crisis, fulminant infection, and elective or emergency surgery operations. Since most of these complications were frequent in Omani children, the incidence of blood transfusion is high compared to Saudi children[4,6]. Furthermore, the difference in hospital policy for the correction of low hemoglobin level may explain the high incidence of blood transfusion in our cases compared to others. Splenomegaly was observed in 40 (38%) patients compared t0 33% in the Eastern Saudi Arabia and 80% in Jamaica[6,8].

Splenomegaly was not associated with splenic dysfunction in Saudi patients compared to Jamaica[8] and black American[7] patients. However, splenic function was not assessed in our study. Hepatomegaly was found in 24% compared to 21% in Saudi children[6]. The lower incidence of biliary stones may be explained by the fact that most of our patients were below six years of age while gallstones were reported in 30% of sicklers over the age of 10 years[16].

CONCLUSION

This is the first study on sickle cell disease in Oman and further studies are required. Although the study represents the capital area of Muscat, it highlights how the disease is severe in Omanis. A large number of the Omani population had lived in Africa for several generations and intermarried with Africans. Whether they may have the African type of SCD gene also needs further study. Since 1993 more recent diagnostic approach of hemoglobinopathy using DNA study has been started and in 1994, a national working group for sickle cell disease was established to study this health problem.

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REFERENCES

1. Luzzatto L. Sickle cell anaemia in tropical Africa. Clin Haematol 1981;3:757-84.

2. Serjeant GR. Sickle cell disease. Oxford: Oxford Medical Publications, 1988,7.

3. White JM, Frost BA. Investigation of haemoglobinopathies. In: Dacie JV, Lewis SM, Eds.
Practical Hematology. Edinburgh: Churchill Livingstone, 1984:179-98.

4. Mulik R, Butikofer A, Paed F, et al. A clinico- epidemiological study of sickle cell anaemia in Saudi Arabia. J Trop Pediatr 1991;37:100-5.

5. Fleming AF. Sickle cell disease. Edinburgh: Churchill Livingstone, 1982:58.

6. Al-Awamy BH, El-Mouzan MI, Niazi GA, et al. Sickle cell anaemia in early childhood in Eastern Province of Saudi Arabia. King Abdul Aziz City for Science & Technology: Riyadh, 1991;49:37-48.

7. Powar DR. Natural history of sickle cell disease: The first ten years. Semin Haematol 1975;12:267-85.

8. Bainbridge R, Higgs DR, Maude GH, et al. Clinical Presentation of homozygous Sickle cell disease. J Pediatr 1985;106:881-5.

9. Murtaza LN, Stoud CE, Davis LR, et al. Admissions to hospital of children with Sickle cell anaemia: a study in South London. Br Med J 1981;282:1048-51.

10. Konotey-Ahulu FID. The sickle cell diseases. Arch Intern Med 1976;133:611-9.

11. Ndugawa CN. Observations of sickle cell anaemia in Uganda. J Trop Pediatr 1978;24:152-4.

12. Barrett-Connor E. Pneumonia and pulmonary infarction in sickle cell anaemias. J Am Med Ass 1973;224:997-1000.

13. Perrine RN, Pembrey ME, John P, et al. Natural history of sickle cell anaemia in Saudi Arabs. Ann Intern Med 1978;88:1-6.

14. Addiego E, Mandel M, Vichinsky EP, et al. Changing pattern of septicemia in sickle cell disease: Possible effect of pneumococcal immunization. Pediatr Res 1981;15:606.

15. Babiker MA. Prophylaxis of pneumococcal infection in sickle cell disease by the combined use of vaccination and penicillin. Ann Trop Pediatr 1986;6:179-81.

16. Vichinsky EP, Lubin BH. Sickle cell anaemia and related Haemoglobinopathies. Pediatr Clin North Am 1980;2:247-9.