CASE REPORT

Sickle Cell Anemia; First Case Reported from Egypt

By A. S. Abbasy, M.B., D.Ch., M.D.

SICKLE CELL ANEMIA, which was first described by Herrick[15] in 1910, occurs almost exclusively in the Negro race. For hundreds of years intermarriages have occurred among inhabitants of the equatorial border of Egypt and Negroes of the neighboring countries. Despite this, not a single case of sickle cell anemia has been reported in Egypt thus far.

Only a few cases have been reported from the African continent, where the Negro race is especially predominant in the central and southern parts: one from Sudan,[2] one from the Gold Coast,[22] and a third from South Africa.[1] Four more cases have recently been reported from West Africa[10, 11] and Rhodesia.[3, 4] The apparent rarity of the disease is perhaps due to the lack of adequate blood studies in many cases of "anemia," since Trowell,[20] by routinely examining fresh blood for sickling, discovered 21 cases in six months among Negroes of East Africa. Unlike sickle cell anemia, sickle cell trait has been well recognized in Africa, where from 9 to 28.3 per cent of the population harbor the trait.[9] Sickle cell anemia, although sought for, has never been found in Egypt. The case reported here is the first to be discovered in this area. It also adds to the few cases of sickle cell anemia described in white families of Greek, Italian or Sicilian ancestry.[6–8, 12–14, 17, 18]

Case Report

A 13 year old white girl from Alexandria, Egypt, was admitted to the Children's Hospital with the chief complaints of pallor and weakness.

For the past three years the girl had suffered from recurrent crises, consisting of anorexia, fever and colicky abdominal pains. During the crises, which lasted from seven to twenty days, the patient complained of extreme fatigue and exhaustion, tenderness of her limbs and joints and jaundice. Such crises occurred five to seven times a year without any apparent precipitating cause. The girl had not yet menstruated, although the average Egyptian girl matures sexually at the age of twelve. The patient belonged to a family which migrated from Algeria to Alexandria three hundred years ago. None of the ancestors is known to have been a Negro. The father and mother, who are second cousins, and five siblings of the patient are in good health. Her paternal uncle died at the age of 18 of an undiagnosed disorder. According to his brother's report, he usually appeared weak and pale and became easily fatigued.

On admission, the girl appeared thin and pale and showed a definite, though slight, greenish-yellow tinge of the sclerae. She was of pale-brownish color, of average height but underweight for her age (76 pounds). She showed no definitely Negroid features (figs. 1 and 2). Her skeletal measurements in inches were as follows: height, 58; sitting height, 27.5; span, 60.5; lower limbs, 30.75; upper limbs, 26; circumference of the skull, 21.

The temperature was normal. The eye grounds showed congestion of the veins. The apical impulse was localized at the fourth intercostal space just outside the midclavicular line; the right margin was two fingerbreadths outside the parasternal line. A blowing systolic

From the Children's Hospital, Alexandria, Egypt.

FIG. 1.

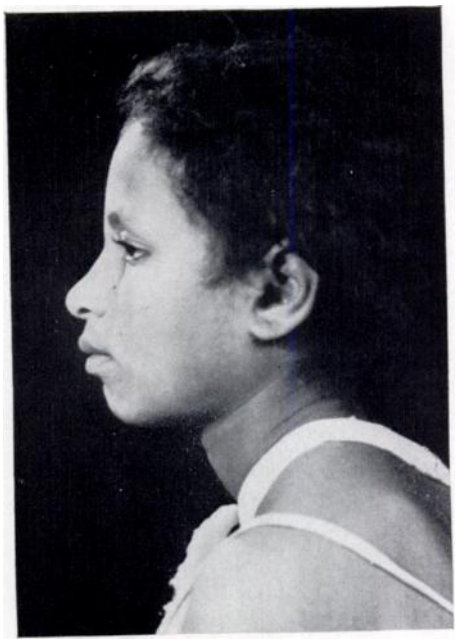

FIG. 2.

murmur was heard, with maximum intensity over the pulmonic area. There were no thrills. The second pulmonic sound was accentuated. The rhythm was regular with a rate of 90. The blood pressure was 118/80. The liver was not enlarged. The spleen was just palpable and not tender. No ulcers were observed on the legs or elsewhere. There were no palpable lymph nodes.

X-ray examination of the chest showed mitral configuration of the heart and increased markings at the base of both lungs. An electrocardiogram was normal.

Laboratory Studies

The blood studies were as follows: red blood cells, 2.50 M.; Hgb, 45 per cent; reticulocytes, 0.1 per cent; platelets, 210,000; white blood cells, 7,600; polymorphs, 54 per cent; lymphocytes, 35 per cent; monocytes, 3 per cent; eosinophils, 8 per cent.

The icterus index was 7 units with a negative van den Bergh reaction. There were no malarial parasites in the smears. The bleeding time was two and a half minutes; the coagulation time was two minutes. The hypotonic fragility of red cells showed initial hemolysis at 0.36 per cent saline and complete hemolysis at 0.22 per cent saline.

Examination of the urine and the cerebrospinal fluid was unremarkable. The stool showed Ascaris ova.

Bone marrow examination showed normoblastic hyperplasia. No sickling of the normoblasts was seen.

Sickling preparations on the patient's red cells were positive. Investigation of all other available members of the family, i.e., 5 brothers and sisters of the patient, her father and mother, her 3 maternal uncles and 8 of their siblings showed sickling in the father, but in no one else.

It was found that the washed red cells of the patient would sickle again when suspended in their own serum, a finding which contradicts the observations of Bell et al.[5]

Course

Since the discovery of this case two and a half years ago, the patient had become accustomed to come to the hospital whenever she felt ill. Sometimes she had minor attacks but at other times she presented herself in crisis which necessitated hospitalization. Her seventh admission was on September 15, 1950. At that time she had mild but definite jaundice, temperature ranging between 37.5 and 38.5 C., anemia, joint and back pains, abdominal colic and palpitations.

The urine contained an excess of urobilin but no bile. The icterus index was 25 and the van den Bergh reaction was positive indirect. Examination of the blood revealed a moderate anemia with increase in reticulocytes. Sickling of red cells was present. The sedimentation rate was 6 mm. in one hour and 15 mm. in two hours after venous stasis of six minutes duration had been produced by a tourniquet. After aeration of the same blood sample the sedimentation rate was 11 mm. in one hour and 50 mm. in two hours. These results support the idea that the abnormal shape of sickled cells prevents rouleaux formation, thus leading to a slow sedimentation rate despite the presence of anemia.[21]

Physically, the patient was in better condition than previously. She had gained 20 pounds in two and a half years. Her skeletal measurements in inches were: height, 60.5; sitting height, 31.5; span, 63; lower limbs, 31.5; upper limbs, 27; circumference of skull, 2.15.

She became sexually mature at the age of 15.

Discussion and Summary

Rheumatic fever, rheumatic arthritis, acholuric jaundice were excluded in this case on the clinical and laboratory findings. The history, physical examination and the laboratory studies all supported the diagnosis of sickle cell anemia.

This case presents certain points of interest. It is the first case of sickle cell anemia reported from Egypt. The disease was found in the patient and her

father and excluded in the other members of the family. It is, however, possible that the paternal uncle had also suffered and died from the disease.

The patient is a white girl and admixture of Negro blood was reasonably excluded through 6 ancestral generations. This case, therefore, adds to those already described in subjects of the white race from the Mediterranean area. It will be noticed that the patient's family originates from Algeria, where 3 cases were diagnosed in natives by Smith[19] on the basis of the anatomical changes observed in the spleen.

REFERENCES

[1] ALTMAN, A.: Sickle cell anemia in a South African born European. Clin. Proc. *4:* 1, 1945.

[2] ARCHIBALD, R. G.: A case of sickle cell anemia in the Sudan. Tr. Roy. Soc. Trop. Med. & Hyg. *19:* 389, 1929.

[3] BEET, E. A.: Sickle cell anemia in Balovale district of Northern Rhodesia. East African M. J. *23:* 75, 1946.

[4] —: Sickle cell disease in Northern Rhodesia. East African M. J. *24:* 212, 1947.

[5] BELL, A. J., KOTTE, R. G., MICHELL, A. G., COOLEY, T. B. AND Lee P.: Sickle cell anemia. Am. J. Dis. Child. *34:* 923, 1927.

[6] CLARKE, F.: Sickle cell anemia in the white race with report of two cases. Nebraska M. J. *18:* 376, 1933.

[7] COOKE, J. V. AND MACK, J. K.: Sickle cell anemia in a white American family. J. Pediat. *5:* 601, 1934.

[8] COOLEY, T. B. AND LEE, P.: Sickle cell anemia in a Greek family. Am. J. Dis. Child. *38:* 103, 1929.

[9] EVANS, R. W.: The sickling phenomenon in the blood of West African natives. Tr. Roy. Soc. Trop. Med. & Hyg. *37:* 281, 1944.

[10] —: Anemia associated with the sickle cell trait in British West African natives. Tr. Roy. Soc. Trop. Med. & Hyg. *39:* 207, 1945.

[11] ROBERTSON, W. M. AND FINDLAY, G. M.: Sickle cell anemia in West Africa. Tr. Roy. Soc. Trop. Med. & Hyg. *40:* 435, 1947.

[12] GREENWALD, L. AND BURRETT, J. B.: Sickle cell anemia in a white family. Am. J. M. Sc. *199:* 768, 1940.

[13] —, SPIELHOLZ, J. B. AND LITWINS, J.: Sickling trait in a white adult associated with hemolytic anemia. Endocarditis and malignancy. Am. J. M. Sc. *206:* 158, 1943.

[14] HADEN, R. L. AND EVANS, F. D.: Sickle cell anemia in the white race. Arch. Int. Med. *60:* 133, 1937.

[15] HERRICK, J. B.: Peculiar elongated and sickle shaped red blood corpuscles in a case of severe anemia. Arch. Int. Med. *6:* 517, 1910.

[16] LAWRENCE, J. S.: Human elliptical erythrocytes. Am. J. M. Sc. *181:* 240, 1931.

[17] ROSENFELD, S. AND PINCUS, J. B.: The occurrence of sicklemia in the white race. Am. J. M. Sc. *184:* 674, 1932.

[18] SIGHTS, W. P. AND SIMON, S. D.: Marked erythrocytic sickling in a white adult associated with anemia, syphilis and malaria. J. Med. *12:* 177, 1931.

[19] SMITH, E. C.: Sickle cell anemia. Tr. Roy. Soc. Trop. Med. & Hyg. *27:* 201, 1933.

[20] TROWELL, H. C.: Sickle cell anemia. East African M. J. *22:* 34, 1945.

[21a] WINSOR, T. AND BURCH, G. E.: Diagnostic physico chemical blood tests in sickle cell anemia. Am. J. M. Sc. *207:* 152, 1944.

[b] — AND —: Sickle cell anemia, "a great masquerader." J.A.M.A. *129:* 793, 1945.

[22] WINTROBE, MAXWELL: Clinical Hematology, ed. 2, Philadelphia, Lea & Febiger. p. 501, 1948.