A BANTU CASE OF HAEMOLYTIC ANAEMIA RESEMBLING ACHOLURIC JAUNDICE

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Acholuric jaundice is considered to be a rare condition in American Negroes. Irby[1] reported a total of 20 cases occurring in 10 families. Later Kline and Holman,[2] in a comprehensive search, collected a total of 42 *bona fide* cases with a familial history, representing 13 different family trees.

In the African this condition is considered an even rarer entity. Gelfand[3] has never seen it in an African, although Foy and Kondi[4] in an article on 'Anaemia in the African' quote one typical case with a family history. Merskey and Baskind[5] published a case of 'Chronic Haemolytic Icterus resembling Acholuric Jaundice occurring in an African male'. Their patient presented with a complaint of pain in both legs and on investigation was found to have an anaemia of which the features were identical with those of acholuric jaundice, but a familial study could not be carried out, and comprehensive techniques for the exclusion of antibodies had not been evolved at that time. The case described here also falls short of a proved example of congenital haemolytic anaemia in that a family study could not be made—a common and disappointing problem in dealing with Africans.

The patient, a young Bapedi male of about 20 years, normally resident in the Northern Transvaal, and at present employed on one of the Reef gold mines, presented himself as a donor at one of the weekly bleeding sessions carried out by the South African Institute for Medical Research. He passed the copper-sulphate specific-gravity (1053) test for haemoglobin and 500 c.c. of blood were collected without any obvious ill effects. The following day, during the course of grouping tests, it was noticed that the serum in the 'pilot-tube' appeared to be icteric, and on examination of the bottle of blood it was seen that about a quarter of the volume of settled cells had haemolysed, and after 5 days' storage at 4°C almost the total volume of cells had undergone haemolysis.

The mine medical officer was immediately notified and the patient hospitalized. Further specimens were obtained and the following investigations were carried out 6 days after the donation of blood, with the results shown:

1. Haemoglobin 11·5 g.%. Leucocyte count 5·2 thousand

per c.mm. (neutrophils 61·0%, monocytes 1·5%, eosinophils 2·0%, basophils 1·0%, late normoblasts 1·0%). Normochromia +, anisocytosis ++, poikilocytosis +, diffuse basophilia ++, spherocytosis ++, reticulocytes 23%.

2. *Serum Bilirubin.* Direct 0·6 mg. per 100 ml., total 3·1 mg. per 100 ml.

3. *Antibody Investigations.* Direct Coombs test negative, indirect Coombs test negative, trypsinized cells negative, ficinized cells negative. Cold agglutinins absent. Indirect Donath-Landsteiner reactions negative.

4. *Haemoglobins.* Alkali denaturation, solubility, and paper electrophoresis, all conformed to normal adult haemoglobin.

5. Schumm's test positive.

6. V.D.R.L. negative

7. *Urine, microscopical:* Ova of *S. haematobium* present. Red blood corpuscles. *Chemical:* Bilirubin absent, urobilin +++.

8. Malaria negative.

A fragility test could not be carried out at that time because the blood had commenced to haemolyse in the short time taken to convey the specimens from the mine to the laboratory (a distance of some 30 miles). For this investigation the patient was therefore brought to the laboratory.

He was a well-nourished young adult with no history of ill-health; he had carried out a normal day's work as an underground worker without any complaints and in fact, as he felt quite well, could not understand why he was being hospitalized. Close questioning regarding any family history of illness or jaundice yielded negative results, but as the patient was a poor witness, little importance could be attached to this. He was quite unaware that his conjunctivae were obviously jaundiced.

His liver was found to be about one finger enlarged and the edge of his spleen was just palpable.

He had a large ulcer on the anterior aspect of the lower third of his right leg, which he said had been present for some time, although he could not remember traumatizing the area. Healed scars were present on both legs and he gave a history of previous ulcers which had, apparently, appeared and healed spontaneously.

TABLE I. FRAGILITY TEST

% Saline	Control		Specimen	
	Before incubation	*After 18 hrs. at 37°C*	*Before incubation*	*After 18 hrs. at 37°C*
	%	%	%	%
0·00	100	100	100	100
0·05	100	100	100	100
0·10	100	100	100	100
0·15	100	100	100	100
0·20	99	100	100	99
0·25	98	100	100	96
0·30	95	100	96	91
0·35	91	100	92	84
0·40	85	98	86	77
0·45	71	95	77	67
0·50	35	87	55	56
0·55	10	38	16	40
0·60	4	8	6	25
0·65	2	5	2	17
0·70	0	2	0	9
0·75	0	0	0	4
0·85	0	0	0	0

A screen fragility test was then carried out, which showed haemolysis commencing in 0·72% saline and complete in 0·32%. The result of an incubated fragility test revealed an increase in haemolysis in the higher concentrations of saline as compared to a normal control (Table I). These findings are compatible with congenital spherocytosis.

COMMENT

The patient was undergoing a haemolytic process at the time of his accidental discovery. The anaemia could not be considered symptomatic in nature since no predisposing factors could be found, and treatment for bilharziasis had not been commenced. The anaemia associated with bilharzia is usually mild and of the hypochromic type. In severe cases with marked liver involvement the anaemia may be macrocytic.[3]

An extensive search for auto- and other antibodies yielded negative results; this was in favour of a congenital rather than an acquired condition.

The anaemia closely conformed to that of acholuric jaundice, but owing to the difficulties encountered with migrant African labour, a familial study for the essential confirmatory evidence could not be undertaken.

Up to 1953, 20 cases had been reported in American Negroes, and only 4 years later the total had increased to 42. In South Africa no proven case has been published to date; it is suggested, however, that a number of carriers may go undetected, since the symptoms are usually mild, so that the patient may quite often lead an active life—the only obvious sign being a slightly sallow tinge of the skin (not applicable here) and sometimes an intractable ulceration of the legs or feet the presenting and only symptom.[6]

In this case, as an example, we have a patient with obviously jaundiced conjunctivae and a chronic ulcer of the right leg, who did not consider himself in any way ill and was only discovered because he presented himself as a blood donor.

SUMMARY

The chance discovery of a case of haemolytic anaemia resembling acholuric jaundice in a young adult African is described. This patient shows that cases may go undetected because of the mildness of the symptoms and emphasizes the difficulty of obtaining corroboratory familial evidence in our South African Bantu.

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