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Erythropoietic (Congenital) Porphyria in a Bantu Girl

BY

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The case here presented is of congenital porphyria occurring in a three-year-old Bantu girl of the Bapedi tribe. This is the second case of this rare type of porphyria recorded in the Bantu of Southern Africa.

ILLUSTRATIVE CASE

The patient was admitted to the Jane Furse Memorial Hospital from a district clinic on 6th December, 1955, with a provisional diagnosis of scurvy. The history was that she had had measles a few weeks previously, and two weeks before admission developed weakness and tenderness of the legs and could not walk.

The patient was the fourth child of healthy parents, had a straightforward delivery and was normal except that, from birth, she passed red urine consistently, had a "patchy" skin and was considered a weakling. A year before the present admission the child developed weak legs and blebs on the skin. These bled if scratched or produced water. The illness resembled the present one and she was admitted to Jane Furse Hospital. After two months she was discharged well, and remained so until the present episode. The parents, the father's other wife and all the patient's siblings were healthy and free from similar symptoms. The paternal grandmother is healthy; the other grandparents had been well during their lifetime.

On examination the child was well grown and plump and looked about 3½ to 4 years. She was extremely lethargic, lay curled up on her side all day, but would co-operate in her examination. The skin was very dark and powdery, as though rubbed in coal-dust. There were several healing blebs on her face and limbs. The hair was normal, but the scalp showed patches of desquamation thought at that time to be healing impetigo. The teeth were foul, buff-coloured beneath a layer of debris which obscured the tooth outlines. The gums were hypertrophic and bled on touch. The nails were dull, thickened and raised off the nail bed at the tips; later a toe nail was shed completely. The mucous membranes were rather pale.

The skeletal system showed no abnormality apart from marked tenderness of the lower legs and ankles. The child refused to take weight on the legs.

The tympanic membranes showed what appeared to be collections of blood within the layers of the drum below the tip of the malleus on both sides, reminiscent of influenzal bullous haemorrhagic otitis media. The heart and lungs were normal. The abdomen was normal apart from a four fingers' breadth spleen. The liver edge was not felt. The superficial structures of the eye and the ocular fundi were normal when seen by the hospital ophthalmologist, Dr. J. H. W. Wessels. The diagnosis of scurvy seemed most likely, but a syphilitic periostitis was considered with a multiple vitamin deficiency in addition. The Mantoux 1/1000 was negative at 72 hours. X-ray of the lower limbs was normal.

At this point the nurse reported that the urine was deep red and stained the sheets. A specimen of urine was sent to Mr. Barnes at the Biochemistry Department of the South African Institute for Medical Research, Johannesburg, who reported (22nd December, 1955): "Porphyrin present (in the urine) in fair quantity; urobilin and a little urobilinogen present." Porphobilinogen was never detected in fresh specimens of urine. The child's general condition improved on hospital diet and hygiene, vitamins and haematinics; notwithstanding the last, the anaemia progressed. Two doses of chloral hydrate were administered without upset.

The findings were in keeping with a diagnosis of porphyria and the child was transferred on 17th January, 1956, to Edenvale Hospital, under the care of Dr. G. Lange, for further investigation. On re-examination the following points were apparent:

Hypertrichosis was present, with excess of fine hair over face, neck, back and extensor aspect of the limbs. An ulcer, almost punched out, was present in the region of the left deltoid, with a scar of a healed bulla over the lateral aspect of the left wrist, and a bulla containing milky fluid on the right forearm. A firm liver edge was palpable one finger's breadth below the costal margin. The patient was able to stand, but not able to walk.

On demonstration under ultra violet light, using a Wood's filter, deep pink fluorescence was seen in the teeth and vulva (urine contamination) and the urine and faeces (19th January, 1956).

While in hospital the blood picture consistently showed evidence of a haemolytic anaemia, which was borne out by the marrow puncture. Porphyrin excretion in stool and urine was very high. Liver function tests suggested a mild impairment of liver function.

The porphyrin excretion in the urine and faeces before the operation is summarised in Table I:

The bone marrow count on the 19th January, 1956, was reported as follows:

Reticulum cells, —; myeloblasts, 0.8 per cent.; promyelocytes, 0.4 per cent.; neutrophil myelocytes, 0.4 per cent.; eosinophil myelocytes, 4.4 per cent.; eosinophils, —; neutrophil metamyelocytes, 2.0 per cent.; staff cells, 4.8 per cent.; segmented neutrophils, 5.6 per cent.; segmented eosinophils, 0.8 per cent. monocytes, 0.4 per cent.; polymorphocytes, —; lymphocytes, 5.6 per cent.; pronormoblasts, 2.4 per cent.; early normoblasts,

Table I
PORPHYRIN EXCRETION

URINE		FAECES	
Total Porphyrin. mg. per day.	Coproporphyrin. mg. per day.	Coproporphyrin. µg./g. dry weight.	Protoporphyrin. µg./g. dry weight.
Jan. 19		1,550	57
20: 14.3	3.5	1,540	47
21: 5.5	1.9	932	26
22: 10.5	2.8	740	60
23: 10.5	2.4	800	58
25: 18.4	3.8		
29: 9.6	2.5		
30: 8.7			
Feb. 1: 14.4			
3: 7.6		1,240	113
4: 3.7			
6: 13.4			
8: 6.7			
9: 5.6			
13: 1.1		43	8
15: 4.8			
20: 11.5			
22: 14.7			

The blood counts from 17th January, 1956, to the 20th February, 1956, are given in Table II.

COMMENTS

19.1.56: The picture is that of an haemolytic anaemia.

7.2.56: There is still diffuse basophilia, anisocytosis and macrocytosis of red cells.

14.2.56: There is a fall in haemoglobin level with moderate anisocytosis and moderate increased diffuse polychromasia.

20.2.56: Little change since 14.2.56. Prothrombin index 88 per cent.

7.2 per cent.; intermediate normoblasts, 32.8 per cent.; late normoblasts, 28.4 per cent.; total nucleated cell count, 200,000 per cu./mm.; myeloid/erythroid ratio, 1:3.4.

SUMMARY

This is a very hypercellular marrow having intense erythroid hyperplasia which is normoblastic in type. Myelopoiesis and platelet genesis appear relatively hypoplastic. There is an eosinophilia present. On examination by ultra-violet light the majority of the normoblasts were observed to show reddish fluorescence.

COMMENT

The erythroid hyperplasia is consistent with an haemolytic anaemia and the fluorescence observed

Table II
DETAILS OF BLOOD COUNTS

	19.156	7.256	14.256	20.256
Haemoglobin (g. per 100 ml.)	9.0	9.4	7.2	7.7
ESR mm./hour (normal 1-15 child)	43	61	70	63
PCV per cent.	29	30	22	24
MCHC per cent.	31	31	33	32
MCV cu.u.	89			
White cells—				
Per c.mm.	6,600	4,800	7,300	8,800
Neutrophils	28.0		44.0	
Monocytes	3.0		2.5	
Lymphocytes	58.0		47.0	
Eosinophils	11.0		6.0	
Basophils	—		0.5	
Late normoblasts	—		—	
Platelets	Normal Nos.	Normal in number and size	Normal in number	
Reticulocytes	16%	9.5%		
Hypochromia	±			
Anisocytosis	++			
Poikilocytosis	+			
Macrocytosis	+			
Diffuse basophilia	+			
Target cells	+			

in the normoblasts is in keeping with the diagnosis of congenital porphyria (*Porphyria erythropoietica*).

Red Cell Fragility (19.156): The test showed resistance of red blood cells to be normal. Haemolysis commenced in 0.5 per cent. saline and was complete in 0.3 per cent. saline.

Liver function tests (7.256): Thymol turbidity, 5.5 units; thymol flocculation, positive +++; colloidal red, positive +++++; cephalin cholesterol flocculation, negative takata ara reaction, positive ++; zinc sulphate turbidity, 25.6 units; total lipid, 392 mg. per cent.; alkaline phosphatase (K.A.), 16.0 units; Van den Bergh reaction, delayed direct; bilirubin, direct, 0.3 mg. per cent.; bilirubin, total, 0.9 mg. per cent.; total protein, 7.6 gm. per cent.; albumin, 3.3 gm. per cent.; globulin, 4.3 gm. per cent.; gamma globulin, 1.61 gm. per cent.; cholinesterase, 93 per cent. of average normal activity.

COMMENT

When compared with the findings in "normal" Bantu children of the same age, these results suggest mild impairment of liver function (Dr. I. Bersohn).

Blood Chemistry (8.256; Dr. M. M. F. Fitzpatrick): Urea = 24 mg. per cent.; potassium = 3.9 meq./litre

= 15.5 mg. per cent.; plasma chloride = 100 meq./litre = 585 mg. per cent. (as NaCl); CO₂ combining power = 21 meq./litre; sodium = 122 meq./litre = 280 mg. per cent.; Schumms test negative, 9.256; serum inorganic phosphorus, 5.0 mg./100 ml. Serum calcium 4.7 meq./L. Direct Coombs test was negative, 11.256. Prothrombin index = 88 per cent. of normal; 20.256 control clotted in 11.1 seconds; specimen clotted in 12.6 seconds.

On these findings a diagnosis of congenital porphyria was firmly established, and since amelioration of symptoms in this condition has been reported after splenectomy (Aldrich *et al.*, 1951), this operation was suggested and the consent of the parents obtained.

Medication before operation consisted of atropine gr. 1/100 by hypodermic injection. An intravenous infusion of 5 per cent. dextrose in water was set up. Splenectomy was performed by Mr. Norwich on 23rd February, 1956. At operation, lasting 20 minutes, an enlarged spleen

aged about 48 years. This was Trowell's first case in East Africa in over 26 years of practice there. The patient enjoyed a diet rich in meats and eggs, butter and animal fats for eleven years. He was a portly figure and weighed 204 lb. In 1947 he experienced severe renoster-nal pain on effort and it recurred in August, 1955. His blood pressure in 1955 was 210/110. On 2nd January, 1956, he developed an acute attack of severe pain, lasting some 10 minutes, for which he was admitted to the Mulago Hospital. The blood cholesterol was 260 mg. per cent. The cardiographic changes were more typical of left heart strain than of a coronary infarction, showing depression of the ST segments in leads V₄ and 6. The T waves were inverted T₁ and ₂ and V₄ and V₆. In this case the pain was perhaps of rather shorter duration than one would expect in coronary infarction and the E.C.G. was not characteristic, as it could be attributed to the hypertension.

The infrequency of coronary artery disease in the Bantu has led to valuable research being done by the Cape Town Medical Department under Prof. Brock, Bronte-Stewart, Keys and Brock studied the serum cholesterol and lipoprotein levels in the three main racial groups in Cape Town, viz., Bantu, Cape Coloured and European. Highly significant differences in the total cholesterol level, particularly in the P lipoprotein fraction of the serum were found, the lowest levels being found in the Bantu and at the other extreme came the European. The fat consumption of the Europeans was more than twice that of the Bantu, with the Coloured group being intermediate. The results of these interesting investigations would support the hypothesis that the dietary intake of fat influences the level of serum cholesterol, especially that in the P lipoprotein.

Yet another cause given for the apparent rarity of coronary artery disease amongst the Bantu was made by Brink. He found that in the Bantu

heart there appeared to be an extra large branch of the left coronary artery arising close to the origin of this artery. He named it the third primary division of the left coronary artery and it ran posteriorly over the surface of the heart and its posterior border. Brink further remarked that the right coronary artery supplied a greater part of the posterior surface of the left ventricle in the Bantu as compared with that of the European. Thirdly, Brink observed that the anterior descending branch of the left coronary artery sometimes passed over the outer border of the heart and ascended posteriorly for a very long distance.

It is felt that within the next few years many more cases of coronary infarction will be published. So far there can be no doubt that occasionally one does meet a case admitting to anginal pains and which would fit in more nearly with coronary thrombosis or angina pectoris than with any other disorder. It is true, however, to state that so far the majority of cases recorded are atypical in one or more features, and it is for this reason that it might be preferable at this stage to refer to them as cases of "Bantu coronary insufficiency."

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