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Osteomyelitis in a Child with Sickle Cell Anaemia

BY

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The literature on the hereditary haemoglobinopathies is expanding very rapidly and the discovery of new haemoglobins seems to have no end. Already the letters A to Q (with the exception of B) and S have been used to name haemoglobins, and at the eighth International Congress of Haematology, September, 1960, in Tokyo, it was requested that "until the next meeting of the International Congress the letters R to Z (excepting S) should not be allotted to new abnormal haemoglobins, but these should be given names of localities."

Haemoglobin S is by far the commonest of the abnormal haemoglobins and is the one most easily identified. Dr. James B. Herrick, of Chicago (1910), was the first to record a case of sickle cell anaemia when he examined a blood film of a 20-year-old West Indian Negro complaining of cough, fever, dyspnoea, weakness and dizziness, and noted: "The shape of the red cells was very irregular, but what especially attracted attention was the large number of thin, sickle-shaped and crescent-shaped forms."

Such aids to diagnosis as a microscope and slide are available to most practitioners in Central Africa to-day; and even if access to methods of electrophoresis, chromatography and alkali denaturation is difficult, it should still be possible to make a firm diagnosis of sickle cell anaemia from the clinical features and from the examination of a blood film.

Sickle cell anaemia is said to be very rare in Southern Rhodesia, and certainly very few cases have been described. Gelfand (1955) described one case of sickle cell anaemia in a child of Shona origin, but when he and Seymour (1960) carried out electrophoretic studies on 120 Mashona no abnormal haemoglobin could be demonstrated.

Brain (1952) examined blood specimens from 1,387 African men who worked at Shabanie Mine and found the sickle cell trait in 51 of them—an incidence of 3.68 per cent. Six of the sicklers came from Southern Rhodesia, 14 from Northern Rhodesia and 11 from Nyasaland, but the actual tribal distribution is not

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given. No case of sickle cell anaemia was encountered, and one of the conclusions drawn from the survey was that possession of the trait is not a cause of morbidity.

At the Wankie Colliery hospital a number of cases of sickle cell anaemia have been diagnosed—mainly in children of Northern Rhodesian or Nyasaland origin (over 80 per cent. of the African employees come from outside Southern Rhodesia and nearly all of these from countries north of the Zambesi). During the 12 months period ending 31st May, 1961, 1,068 children were admitted to the children's ward and 17 were thought to have sickle cell anaemia.

With the present trend in migrant labour likely to continue, it is important that sickle cell anaemia should be considered in the differential diagnosis of many of our commoner clinical problems—anaemia, oedema of the feet, dactylitis and frank osteomyelitis.

The object of this paper is to describe a case of suppurative osteomyelitis occurring in a child suffering from sickle cell anaemia. This complication of sickle cell anaemia has been found to be common in Western Nigeria, but rare in the Belgian Congo. No case has been reported in Southern Rhodesia. Further observations on sickle cell anaemia found in Wankie will be the subject of a later paper.

CASE REPORT

A 3½-year-old Tonga boy was first seen on 27th October, 1960, with a history of fever and wasting for many weeks. No one seemed to have noticed a large, hot, tense, tender, non-pulsatile swelling of the lower half of his right thigh. He was dyspnoeic at rest and obviously very ill. His temperature was 103° F., the mucosae very pale and the sclerae icteric. There was parietal bossing and proptosis of the eyes (the right being more marked than the left); the heart appeared normal, but examination of the respiratory system revealed coarse crepitations at the left base; the liver was enlarged 4 cm. below the costal margin and the spleen showed a class 2 enlargement (using Hackett's classification), being firm but not tender.

Investigations

Weight, 20 lb.; Hb., 3.7 g./100 ml. (25 per cent.); WBC, 24,800/c.mm. (polymorphs 29 per cent., lymphocytes 19 per cent., eosinophils 1 per cent., monocytes 9 per cent., intermediate normoblasts 8 per cent. and late normoblasts 34 per cent). The sickle cell test was markedly positive, both filamentous and sickle shapes being present. Haemoglobin electrophoresis showed only haemoglobin S, and no foetal haemoglobin was detected on alkali denaturation. Serum bilirubin was 3.10 mg./100 ml. and the serum proteins were 8.75 g. per cent. total; albumin 4.20 g./100 ml. and globulin 4.55 g./100 ml.

The urine contained a trace of albumen, excess of urobilin, and microscopical examination revealed a few pus cells and scanty erythrocytes, together with calcium oxalate crystals; there were no ova. There was nothing of note in the stool examination. Samples of the urine, stool and pus from the leg were repeatedly cultured, but no salmonellae could be grown and the Widal reaction was also negative. *Staphylococcus aureus* was isolated from the pus and was shown to be sensitive to chloromycetin and erythromycin, but resistant to penicillin, streptomycin and aureomycin. He was treated with erythromycin 200 mg. q.d.s. for five weeks.

Chest X-ray examination revealed a segmental collapse in the right upper lobe and no cardiac enlargement. A slight degree of bossing of the skull was apparent. X-ray of the right femur (Fig. 1) showed a thin cortex and a widened medullary cavity with patchy decalcification; there was periosteal reaction forming a false

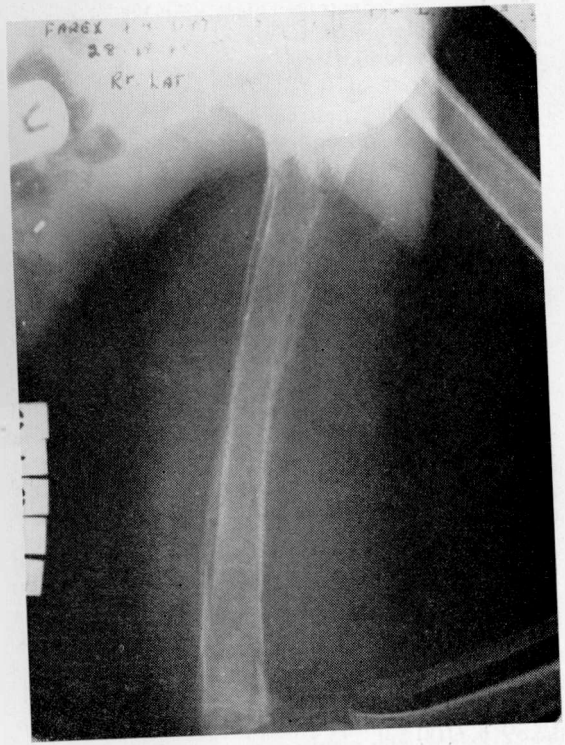

Fig. 1—Note the thinned cortex and widening medullary cavity.

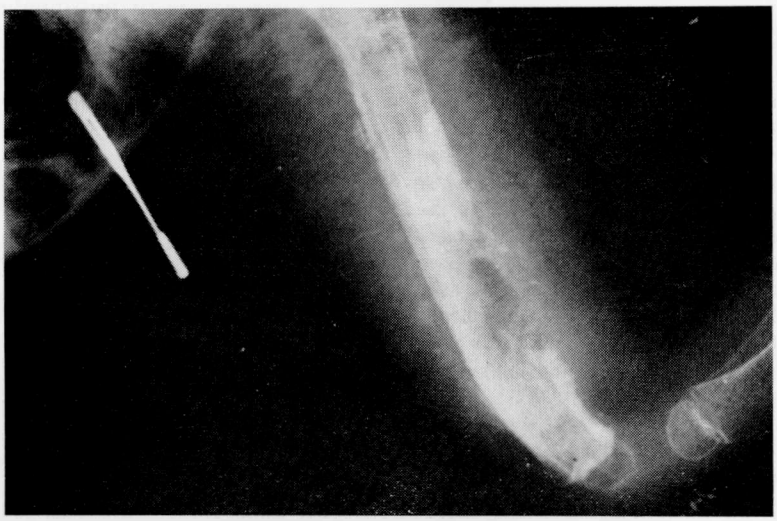

Fig. 2a—Note the periosteal reaction.

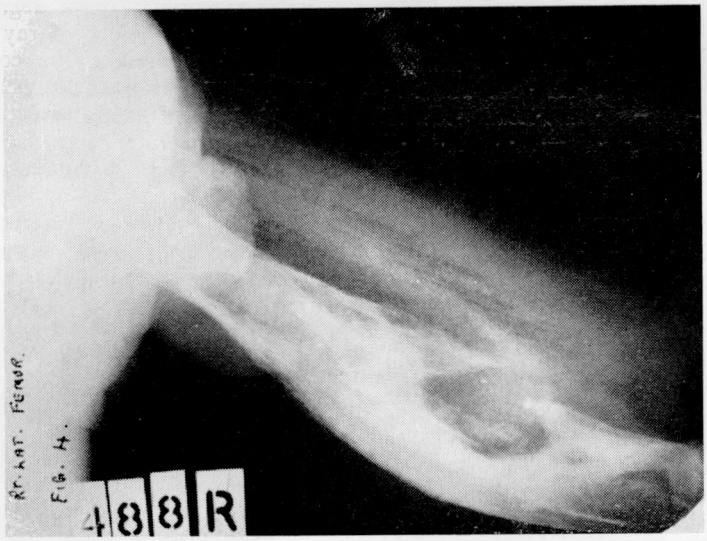

Fig. 2b—The increasing periosteal reaction is clearly seen.

shaft, and the epiphyses of the femur and tibia and the upper end of the tibia were partially decalcified.

The child was transfused with 400 ml. of blood and the haemoglobin rose to 9.59 g./100 ml. (65 per cent.). The superficial tissues were incised two days after admission, and one week later the bone was scraped. By this time the haemoglobin had dropped to 5.15 g./100 ml. and a further 400 ml. blood brought it up to 12.59 g./100 ml.

X-rays of the femur taken on 19th November, 1960 (Fig. 2 (a) and (b)) showed a more marked periosteal reaction with a great deal of bone formation, which extended the whole way up the shaft. Destruction of the underside of the lower third of the femur, a result of the scraping ten days previously, is also apparent.

Fig. 3 shows the X-ray taken five weeks later on 27th December, 1960, when further gross periosteal reaction had taken place, and there were now fractures through the upper ends of both tibia and fibula as well as through the femur. The haemoglobin again fell, and when it had reached 6.0 g./100 ml. (40 per cent.) on 7th January, 1961, a further 400 ml. blood

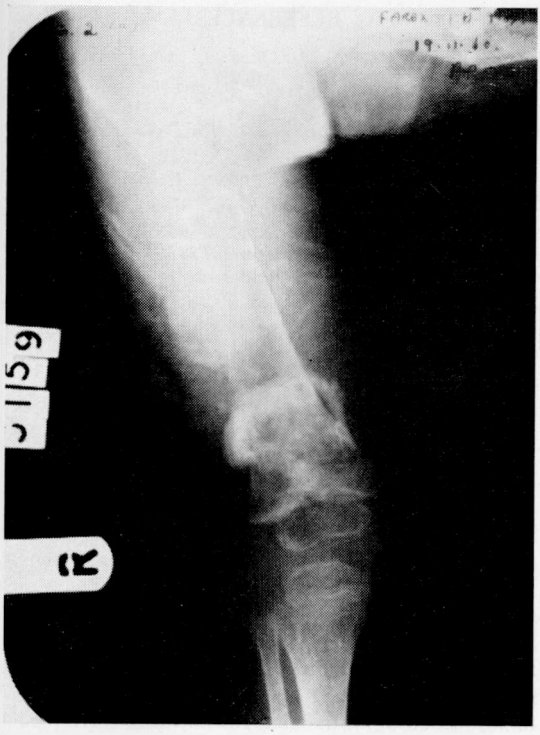

Fig. 3—Showing gross periosteal reaction with fractures through the tibia and femur.

was given. On 21st February an X-ray (Fig. 4) showed the periosteal reaction around the shaft of the femur still very much in evidence, and through it the pattern of the old shaft still visible.

At varying intervals during the following months there were episodes of pyrexia, marked irritability and on one occasion there was a hot, tense, painful swelling over the lower end of the right radius, where some periosteal reaction was visible on X-ray. On another occasion periosteal reaction around the right clavicle was noticed on a chest X-ray.

Jaundice, increasing anaemia and a reticulocytosis (26 per cent. of the total red cells on one occasion being reticulocytes) were also evident during these crises. Following the practice of Smith *et al.* (1953) and Hendrickse (1960), Priscol in a dosage of $6\frac{1}{4}$ mg. t.d.s. was administered and there did appear to be a marked relief of symptoms—the vascular spasm, which is thought to be a factor in the pathogenesis of the crisis, being relieved by the vasodilator drug. A further blood transfusion was required (bringing the total to two litres in six months) before the patient was transferred to Livingstone hospital at the end of April, 1961, where the condition of the femur has improved further but the haemolytic process has continued, making a further blood transfusion necessary.

Family History

The child is the elder of two children, the brother being two years old. Father is a plateau Tonga from Kalomo, Northern Rhodesia, and mother is a Gwembe or valley Tonga from near Binga, on the south bank of the Zambesi. The two children were born and have spent all their lives in the Wankie district of Southern Rhodesia.

Father: A perfectly healthy man who has suffered from no serious illnesses. Hb, 16 g. per cent.; reticulocytes, 0.4 per cent.; sickle cell test markedly positive; electrophoresis showed Hb S only, and alkali denaturation revealed 40 per cent. foetal haemoglobin. A blood film was quite normal and no target cells could be seen. Red cell fragility test was within normal limits. A skeletal survey also failed to reveal any abnormality.

Mother: A healthy woman. Hb, 12.2 g. per cent.; reticulocyte count, 1 per cent.; a moderate amounting of sickling was demonstrated; electrophoresis showed HbA and HbS and no foetal haemoglobin was detected on alkali denaturation.

Brother: Normal child; clinically not anaemic. Unfortunately the result of haemoglobin determination was mislaid; electrophoresis showed HbA and HbS and no foetal haemoglobin was detected on alkali denaturation.

Genotyping therefore supported the diagnosis of homozygous sickle cell anaemia.

COMMENT

Osteopathy in one form or another is a very common feature of sickle cell anaemia. In infants the metacarpal and metatarsal bones and the proximal and middle phalanges are often the sites involved, whereas in older children the long bones are the ones usually affected. Hot, tense, tender swellings occur over the bones at the time of a crisis, and on X-ray examination then or two weeks later a faint periosteal reaction can often be seen. It is thought to be due to thrombi causing an aseptic necrosis. In the case described above a similar periosteal reaction was observed around the right clavicle, whilst the only abnormality in the bones of the hands was a little decalcification and widening of the nutrient arteries of the proximal phalanges. None of the usual skull changes was seen except some slight parietal bossing. There was no suggestion of the "hair-on-end" appearance often described in sickle cell anaemia, but much more commonly found in thalassaemia.

Hendrickse (1960), in his survey of sickle cell anaemia in Nigerian children, found that osteomyelitis is a common complication of the disease. However, the Lambotte-Legrandes (1958), in their account of the disease as it occurs in the Belgian Congo, state that although bone changes resembling those of osteomyelitis are seen, actual suppuration is rare. Both Hendrickse and the Lambotte-Legrandes have found that when there is a superimposed infection it is often due to a salmonella organism. Mercedes Vda-de Torregrosa and colleagues (1960), writing from San Juan city hospital, record three cases of salmonella-caused osteomyelitis associated with sickle cell disease. After a review of the literature they point out that salmonella-caused osteomyelitis is a rare complication of salmonella infections, but that the association of salmonella-caused osteomyelitis with sickle cell disease is frequent. They suggest that tests for sickle cell anaemia should be performed on patients with salmonella-caused osteomyelitis and a search for salmonella organisms be conducted in patients with sickle cell anaemia.

Although the use of a tourniquet to assist the surgical treatment was not contemplated in this case, it is well to bear in mind the warning given by Hendrickse (1960) against its use. He reports that two cases of gangrene occurred at University College Hospital, Ibadan, due to the use of a tourniquet during the surgical treatment of patients with osteomyelitis complicating sickle cell anaemia. It would seem that the anoxia causes gross sickling of the red cells in the blood vessels of the limb, and this is followed by thrombosis and subsequent gangrene.

Dr. P. E. S. Palmer was invited to comment on the series of X-rays and he writes: "The decalcification and widening of the bone marrow can occur in any disease in which there is an active marrow. However, in sickle cell anaemia bone infarcts occur frequently, and in this case are well demonstrated in the upper end of the tibia and fibula as well as in the femur itself. There is a tendency throughout the series to maintain the ghost of the original shaft, whereas in a simple chronic osteomyelitis, although there may be just as much periosteal reaction, it is unusual for a complete sequestrum of the shaft to form as occurs with sickle cell anaemia. It is, however, the associated changes in the upper end of the tibia and fibula, as well as the condition of the shaft on the original film of the femur (Fig. 1), which would lead to suspicion of sickle cell disease."

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