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Infantile Cortical Hyperostosis (Caffey's Disease) in a Nigerian Child

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Summary

Azubuike, J. C. (1976). *Nigerian Journal of Paediatrics*, **3** (1), 26. **Infantile Cortical Hyperostosis (Caffey's Disease) in a Nigerian Child.** A case of infantile cortical hyperostosis (Caffey's disease), in a female infant is reported. She presented with a history of irritability and cold, tender swelling of both lower limbs. The diagnosis was confirmed radiologically. Haematologically, there was thrombocytopenia.

INFANTILE cortical hyperostosis (Caffey's disease), is a well recognised clinical and radiological entity of unknown actiology (Caffey and Silverman, 1945). The condition consists of hyperplasia of the sub-periosteal bone over which there is soft tissue swelling. Occasionally, there may be an overlying skin discoloration. The skeletal changes occur in several bones—mandible, clavicle, calvarium, scapula, ribs and the long bones of the extremities. Though the clinical symptoms may vary, they are as a rule, not severe. This communication reports a case of this disease in a Nigerian infant.

Case Report

C.I. (UNTH 74/073675), a female infant, aged two-and-a-half months, was admitted with the main complaints of swelling of both legs and excessive crying for four weeks. There was also a history of mild fever, cough and nasal stuffiness. Appetite was good.

Antenatal care was at a private maternity hospital. Pregnancy was normal. Delivery was at term, and the baby cried immediately at birth, necessitating no resuscitation. Birthweight was 3 Kg. There was no past illness of significance. The baby was the only child of young healthy parents.

On admission, the temperature was 37.5°C. The infant was irritable but well nourished and well developed. She weighed 6 Kg; the respiratory rate was 30 per minute and pulse rate 120 per minute. The head circumference measured 38.5 cm and the anterior fontanel was patent and not bulging. The legs were swollen and tender but not warm. The infant cried constantly during the examination of the legs. There was also a small, tender and cold swelling over the distal end of the clavicle.

Laboratory investigations revealed: Hb., 12.7 gm per cent; PCV (haematocrit) 36 per cent. White blood count was 3,650 per cmm with 32 per cent neutrophils, 3 per cent eosinophils, 52 per cent lymphocytes and 6 per cent monocytes; platelets 58,000 per cubic millimetre;

erythrocyte sendimentation rate 13 mm 1st hour (Westergren). Haemoglobin genotype was A; VDRL was negative, serum calcium was 9 mg. per cent, phosphate 5.5 mg per cent; alkaline phosphatase 200 i.u./L. Immunoglobulins were qualitatively normal.

Radiographs of the legs (Fig. 1) showed cortical hyperostosis of both tibiae and a soft tissue swelling. There were similar radiological changes in the left clavicle. Although corticosteroid therapy has been recommended for the treatment of this disease (Caffey, 1967), the present case was successfully treated symptomatically using acetylsalicylic acid. Follow-up radiographs four months later showed a marked decrease in the hyperostosis in both tibiae (Fig. 2).

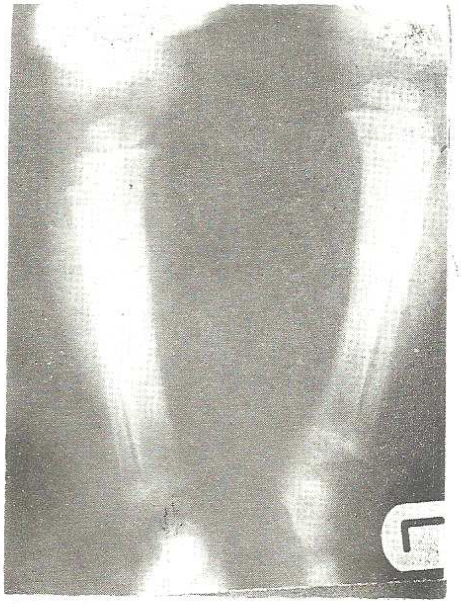

Fig. 2. Repeat radiograph at the age of 6½ months showing almost complete disappearance of the hyperostosis.

Discussion

This patient was a previously healthy infant who suddenly developed multiple bony swellings with tender induration of the overlying soft tissues. The clinical and radiological features were consistent with the diagnosis of infantile cortical hyperostosis as originally described by Caffey and Silverman (1945). Although most of the cases so far reported have had polyostotic lesions, few European and non-European workers have reported monostotic forms (Krause, 1973; Berio, 1973; Mazur, 1973; Sudjarwo, 1972). Because of the presence of acute inflammatory changes in the periosteum, Caffey (1967, postulated an infective basis (either prenatal or postnatal) for this condition. A recent report by Temperly, Douglas and Reeves (1972), gives this postulation some support because their patient had persistently raised Ig A, Ig M and IgG. In viral infections, persistently raised IgM has been reported. In the present case there was neither clinical, haematological nor immunological evidence to suggest an infective basis for the condition.

There has been only one previous report (McEnery and Nash, 1973) which mentions thrombocytopaenia in association with Caffey's syndrome. The two patients reported by these authors also had Wiskott-Aldrich syndrome. Our

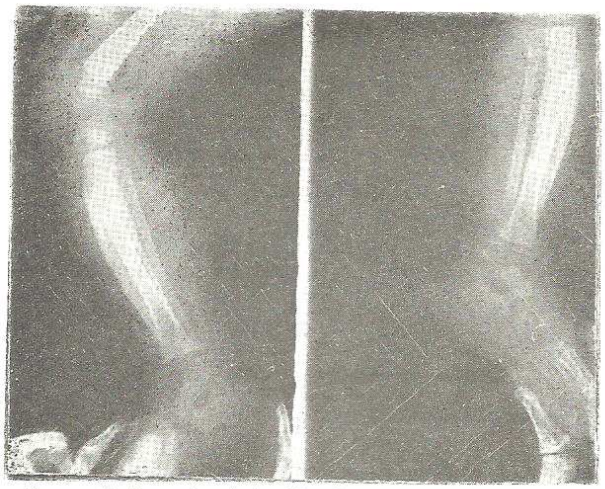

Fig. 1. Radiographs of the tibia-fibula of the patient aged 2½ months. Note the hyperostosis of the bones.

patient with a platelet count of 58,000 per cmm. had no clinical features of Wiskott-Aldrich syndrome. In contrast, Temperly, Douglas and Reeves (1972) found thrombocytosis in their case. The significance of thrombocytopaenia or thrombocytosis in association with Caffey's disease requires further study.

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