Cortical Hyperostosis
Infantile and Juvenile Manifestations in a Boy

Cortical hyperostosis was recognized as an entity by Roske (1930), and again by Caffey and Silverman (1945). This condition is characterized by its occurrence in infancy with fever and irritability, and the development of cortical new bone in the mandible and other bones. The features generally regress completely in infancy and very early childhood, but there may be some tendency to relapse during this period.

Case Report

This report gives details of an Iraqi boy, now 19, who had had signs of cortical hyperostosis from 1951, when he was 4 months, which had persisted to the present time. The condition began with involvement of the mandible and subsequently changes were observed in the right ulna, right tibia, ribs, right femur, and left fibula (Fig. 1–3). The condition relapsed and remitted at increasingly long intervals, and, until the age of 14 years, steroids were effective in controlling symptoms. In the past 5 years this treatment has been less successful and there has been protracted swelling and hyperostosis in the left leg, and in the right forearm where there is limited rotation without any radioulnar synostosis (Caffey, 1952).

Comment

Cortical hyperostosis may present in several members of the same family (van Buskirk, Tampas, and Peterson, 1961), but all relatives of this Arab boy are unaffected by the condition. There are a number of causes of diaphysial new bone formation, but all other entities have been carefully excluded as diagnostic possibilities in this patient with cortical hyperostosis. Investigations in this patient have included biopsy from the right ulna at age 2 years, and from right fibula at 3 years, which showed a histological picture consistent with Caffey's disease. Since there is no specific diagnostic test for cortical hyperostosis as described by Caffey, and because the aetiology of the condition remains in doubt, it must be presumed that this Arab boy has the condition.

The duration of the condition in this patient is exceptionally long. It is clearly inappropriate to append the adjective 'infantile' to cortical hyperostosis which, though having typical features, has continued from infancy through childhood and into adolescence.

REFERENCES


Raised Serum IgM Levels in Neonatal Endemic Diarrhoea

For over 2 years cases of diarrhoea occurred intermittently on the Special Care Nursery of the Bolton District General Hospital, and no cause was found on routine investigation. During the period of this study, from December 1968 to April 1969, 209 babies were admitted into the unit and 44 of these developed diarrhoea, but only 25 of these were selected for special study because the other 19 cases had idiopathic respiratory distress syndrome (6), asphyxia neonatorum (1), haemolytic disease of the newborn (3), hypoglycaemia (6), intracranial haemorrhage (2) or urinary infection (1) as well as the diarrhoea.

Of the 209 admissions, 102 were of low birthweight (2.5 kg or less) and 68 were small-for-dates. Of the 44 cases with diarrhoea, 35 were of low birthweight, and 24 were small-for-dates. Of the 25 cases under study, 18 were of low birthweight and 15 were small-for-dates. The incidence of diarrhoea was significantly higher in small-for-dates infants (χ² = 11.27 with 1 D.F., 0.001 > P > 0.005). Male infants showed a striking preponderance over female in susceptibility to the illness (χ² = 4.18 with 1 D.F., 0.05 > P > 0.025).

The clinical features were fairly similar in all cases. The affected infants had either lost weight or failed to gain weight shortly before the onset of diarrhoea. Lethargy and anorexia were common preceding symptoms. The stools were frequent, explosive, and watery.

The affected babies were isolated locally and barrier nursed. The milk was discontinued for 24 hours and was replaced by clear fluids (glucose and electrolytes mixture) given orally. Adequate fluid intake was ensured by tube feeding when necessary. Intravenous fluid was given in one patient who was moderately severely dehydrated. Milk feeding was reintroduced after 24 hours in gradually increasing concentration. 19 patients responded to this regimen, but in 6 oral neomycin sulphate was given because the response to clear fluids was delayed. There was no mortality.

The diarrhoea occurred in small outbreaks and relatively more cases tended to occur when the nursery was fully occupied. The symptoms developed between the 5th and 11th day of life with a peak incidence between the 8th and 10th day. The birthweight did not appear to affect the age of onset of diarrhoea. The pattern of feeding and the calorie intake of the affected cases was not significantly different from that of the unaffected babies. Only 3 babies were fed completely on human milk and none of them developed diarrhoea.

For a standard of comparison the investigations which were done on the affected cases were also carried out on a large number of unaffected babies of similar birthweight and age on the unit over the same period.

In 9 of the 25 cases the faecal pH was acid (pH between 5.5 and 6) but it quickly rose to 7 or above after 1 to 2 days of treatment. In about 12 per cent of the unaffected babies the faecal pH was also acid (pH between 6 and 6.5).

Two viral cultures of stools were done on each case with negative results. Three specimens of