of children with EN of undetermined aetiology were in the upper part of the normal range for healthy children and their mean value was also slightly higher (Table). Statistical analysis was not attempted because of the small number of cases.

Discussion

Clinical symptomatology, aetiological diagnosis, age, and sex incidence in our series of cases do not differ in any respect from other reports (Lorber, 1958; MacPherson, 1970).

Specific allergic antibodies long known as reagins belong to the IgE fraction. As already stated, it has been shown that not all IgE are reagins (Havnen *et al.*, 1973), though it is not clear what other role IgE plays in the immune response. In the present study children with poststreptococcal EN had very high levels of serum IgE reaching over 10 times that of controls. We are studying whether IgE increases in other sequelae of streptococcal infections such as rheumatic fever, acute glomerulonephritis, or even during the actual course of streptococcal infection.

Joint involvement, extension of the rash, and recurrence, which are characteristic of streptococcal EN, could well represent an extension of an immune response. So could IgE production, but the two should not have a cause and effect relationship, as higher levels of IgE do not directly correlate with the severity of clinical manifestations.

Summary

Twenty-two children aged from 3 years 6 months to 12 years with erythema nodosum (EN) were grouped according to aetiology into streptococcal, tuberculous, and those whose aetiology was undetermined. Serum IgE levels were determined in all by the radioimmunosorbent technique. Levels were higher (mean value 991 units/ml) in streptococcal EN than in those of the other two groups (mean value 68 units/ml and 97 units/ml, respectively) and healthy age-matched controls (mean value 60 units/ml). Increase of IgE levels in individual cases of streptococcal EN did not correlate with severity of the clinical manifestations characteristic of that group.

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Vitamin E deficiency and thrombocytosis in Caffey's disease

Infantile cortical hyperostosis, described first by Caffey and Silverman in 1945, affects both sexes equally and onset is always before the age of 5 months. It is characterized by soft tissue swellings overlying cortical thickening which has been observed in all tubular bones except phalanges and vertebral bodies. It also affects flat bones. Infants usually exhibit irritability, pallor, and fever. There may be unpredictable remissions and relapses. Cases have presented with periorbital swellings and proptosis (Minton and Elliott, 1967). Associated thrombocytosis has been observed (Pickering and Cuddigan, 1969), but to our knowledge vitamin E deficiency in Caffey's disease has not previously been described.

Case report

A female, the fourth child of healthy parents, was born by normal delivery at term after a normal pregnancy. She was small-for-dates with a birthweight of 2640 g (<10th centile). After initial feeding difficulty she progressed well until 2 months when she presented with irritability, nasal obstruction, slow feeding, and intermittent swelling around the right eye with dropping of the eyelid on that side.

FIG.—Oblique view of mandible showing typical periosteal layering of Caffey's disease.

Examination showed fever, right-sided ptosis, and slight periorbital swelling, but no other abnormal signs. Length and weight lay on the 75th and 50th centiles, respectively. She initially recovered spontaneously but required readmission 3 weeks later with a recurrence of the same symptoms. She was intermittently febrile while on the ward. No evidence of sepsis was found but she subsequently developed left-sided mandibular swelling. X-ray showed lamellar new bone formation affecting the whole of the mandible but there was no evidence of cortical hyperostosis affecting the orbits (Fig.).

Investigations showed hypochromic anaemia Hb 9.9 g/dl and low serum iron 19 μ g/100 ml; leucocytosis 19 500/mm³ (polymorphs 34%, lymphocytes 56%); platelets 860 000/mm³ rising to 910 000/mm³. Bone marrow was very cellular with abundant megakaryocytes; haemoglobin electrophoresis normal. Urea, electrolytes, uric acid, cholesterol, proteins, serum glutamic-oxaloacetic transaminase, bilirubin, calcium, alkaline phosphatase, inorganic phosphorus were normal. IgM raised at 120 mg/100 ml (normal range at age 3–6 months 19–86 mg/100 ml). Leucocyte transformation normal. Viral studies on throat swab, stool, urine, and paired sera were all normal. Vitamin E level 0.1 mg/

100 ml (mean of 21 normal children aged 6 months–13 years $\pm$ 1 SD 0.8 $\pm$ 0.2 mg/100 ml); repeated after 3 months, 0.4 mg/100 ml.

The left-sided facial swelling subsided after one week but was followed by similar swelling on the right side. Feeding continued to be erratic until the age of 5 months when all symptoms gradually disappeared. X-ray then showed that the lamellar new bone had been modelled into the mandible. The platelets had also returned to normal.

Discussion

The diagnosis of Caffey's disease in our patient was made 6 weeks after the onset of ptosis and periorbital swelling, the significance of which was not appreciated at the time. The ocular manifestations of infantile cortical hyperostosis have received scant mention in published reports and are probably more common than realized. Minton and Elliott (1967) reported 24 cases, of which 8 had periorbital swellings and in 6 of these the periorbital swelling was noted before mandibular swelling became apparent. Iliff and Ossosky (1962) reported a case

in which there was rapidly progressive unilateral exophthalmos. In the report of Galyean and Robertson (1970), the patient presented in much the same way as ours with puffy eyes, irritability, and poor feeding. This child had glaucoma which resolved along with the hyperostosis over a period of 4 months.

The occurrence of thrombocytosis in Caffey's disease is now well recognized. Pickering and Cuddigan (1969) described 3 cases, all with raised platelet counts. In their review of reports published in English they found 3 cases with platelet counts $> 500\,000/\text{mm}^3$, this measurement having been made in only 9 of 132 cases. Our patient's platelets rose to $910\,000/\text{mm}^3$ but she suffered no thrombotic complications and no anticoagulant or antimegakaryocytic agents were used.

The reason for the thrombocytosis in Caffey's disease is not known. The low vitamin E levels shown in our patient may be a fortuitous finding but it is tempting to postulate a possible association between the thrombocytosis and the vitamin E deficiency in our patient. Ritchie *et al.* (1968) described a syndrome of oedema, haemolytic anaemia, and thrombocytosis in association with vitamin E deficiency. Thrombocytosis was also noted by Hassan *et al.* (1966) in their description of a syndrome in premature infants associated with low plasma vitamin E levels and a high polyunsaturated fatty acid diet. Our patient was fed on Cow and Gate 'Premium', then changed to Ostermilk II. She received Abidec drops (vitamins A, B, C, D) but no vitamin E. The plasma vitamin E level rose spontaneously to reach the lower level of normal after 3 months. There was no evidence of haemolysis.

Whether vitamin E deficiency is a factor in the pathogenesis of Caffey's disease remains to be seen. It is hoped that this report will stimulate further studies of vitamin E levels in infantile cortical hyperostosis.

Summary

An infant is described who presented with ptosis and periorbital oedema and was found to have infantile cortical hyperostosis with thrombocytosis, raised IgM, and vitamin E deficiency.

I am indebted to Dr. A Moosa for his advice in preparing this paper and to Dr. D. P. R. Muller, Institute of Child Health, The Hospital for Sick Children, for the measurements of vitamin E levels.

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Gastric pneumatosis in infancy

Gastric pneumatosis (defined as gas within the wall of the stomach) may be easily detected by radiography. It presents as a fine lucent stripe conforming to the contour of the stomach and enveloping any intraluminal gas and fluid content. The finding, though quite rare, is of utmost clinical importance. In adults, gastric pneumatosis is caused either by gas-forming bacterial infection (emphysematous gastritis), or in association with bullous emphysema (Holgersen, Borns, and Srouji, 1974). In infancy, isolated gastric pneumatosis has been seen very rarely in gastric outlet obstruction. It has also been reported in neonatal necrotizing enterocolitis, in association with intestinal pneumatosis (Bell, Graham, and Stevenson, 1971; Santulli *et al.*, 1975) though it may rarely be isolated (Robinson, Grossman, and Brumley, 1974). Thus, radiographic detection of gastric pneumatosis indicates serious underlying disease and determination of its cause will depend on the associated clinical findings, a point illustrated by the 2 cases presented here.

Case reports

Case 1. A 6-week-old girl developed vomiting, constipation, and oliguria 5 days before admission. She was a well-nourished infant, mildly dehydrated, and irritable. There was moderate epigastric distension but no visible peristaltic waves. No pyloric 'tumour' could be palpated. Laboratory studies including serum electrolytes were normal, except for slightly raised blood urea nitrogen ($18.7\text{ mg}/100\text{ ml}$). Radiographic examination of the abdomen showed gastric distension with a long air fluid level in the stomach seen on the upright film. The findings suggested gastric retention.