result of maternofetal blood group incompatibility is relatively common, persistent hyperbilirubinaemia is much less frequent. Because the other features of hypothyroidism may be minimal at this age, it would be ideal to perform thyroid function studies in all babies in whom unexplained hyperbilirubinaemia persists more than 2 or 3 weeks. These tests should certainly be done whenever slight inactivity, slowness with feeding, or constipation is present in addition to jaundice.

Summary

Twelve patients are described in whom hypothyroidism was associated with significant neonatal jaundice which persisted for as long as 7 weeks in some babies. The fact that these babies were seen at one paediatric centre during an 11-year period indicates that this association is not uncommon. Recognition that hyperbilirubinaemia can be the only obvious symptom of hypothyroidism in the neonatal period is diagnostically important.

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References


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Infantile Cortical Hyperostosis

with Lytic Lesions in the Skull

A case of infantile cortical hyperostosis is described in which lytic changes in the skull vault were a prominent feature and which posed a diagnostic problem.

Case History

A female child, the second child of healthy unrelated parents, was born at term, birthweight 3617 g. For a chest infection in the neonatal period antibiotics were given. After a period of good progress she fell ill at the age of 4 weeks with a cold followed by bronchitis, for which she was given a course of antibiotics at home. One week later swelling developed around the left eye and a few days later spread to both periorbital regions. Aged 9 weeks, she was seen in hospital because the swelling had persisted. No other abnormality was noted. Skull x-rays at this stage were unremarkable, but Hb was only 7.9 g/100 ml and there was a leucocytosis (white blood cells 21,700/mm³). Despite this anaemia and a further cold she remained well but feeding became a little slow.

At the age of 3 months because of the persisting periorbital swelling she was transferred to this hospital with a diagnosis of possible neuroblastoma. She was a pale baby on the 75th centile for weight. Both upper eyelids were swollen but there was no proptosis and the fundi were normal. There was no general oedema and no clinical evidence of cardiovascular or renal disease.

Investigations confirmed her anaemia, Hb 7.3 g/100 ml with anisocytosis, poikilocytosis, and a low serum iron of 21 µg/100 ml. Reticulocytes were 2.8%. Hb electrophoresis was normal. WBC 11,800/mm³ with a normal differential. The platelet count was increased at 955,000/mm³ ESR 80 mm/1 hour. Bone-marrow examination showed a slight shift to the left with morphologically healthy megakaryocytes. No blood loss was detected in the stools. The alkaline phosphatase was 25 KA units/100 ml. 24-hour urinary VMA was 0.25 mg. Other biochemical investigations were normal. Skull x-rays (Fig. 1) showed patchy lytic lesions in the frontal bones bilaterally and early hyperostosis of the mandible. An IVP showed no evidence of a neuroblastoma and the rest of the skeletal system appeared normal.

Shortly after admission her anaemia was corrected by transfusion since when she has remained clinically well, without further anaemia, and gained weight satisfactorily. No further therapy has been given.

Her face became progressively more deformed, and between 4 and 6 months of age she developed a heavy jowl appearance with squaring of the lower face. At the age of 7 months further skull x-rays (Fig. 2) showed a mandible which had the characteristic features of infantile cortical hyperostosis but the lytic lesions previously seen in the frontal bone had disappeared. At this time the ESR had fallen to 10 mm per hour, the platelets were normal, and Hb was being maintained at a normal level. When seen at 10 months of age there had been some clinical improvement in the appearance of the lower face.

Discussion

The present case is of particular interest both because of the radiological features and because of the light it may throw on the pathogenesis of Caffey's disease. Radiologically evident bone erosions are not a well-known feature of the condition and seem to have been described previously only recently by Neuhauser (1970). In the absence of other osseous lesions, differentiation of the skull
FIG. 1. (a) and (b).—Aged 3 months: lytic areas in frontal region and early hyperostosis of the mandible.

FIG. 2.—Aged 7 months: mandibular hyperostosis remaining.

changes here described from the appearance of secondary neoplasm, particularly neuroblastoma, may be difficult, and the diagnosis may only become clear with evolution of more typical radiological appearances.

Eversole, Holman, and Robinson in 1957 described focal resorption of cortical bone as an early microscopical feature in the evolution of infantile cortical hyperostosis. The relative vascularity and the thinness of the cortical bone of the skull vault may be the reason why this histological change has not until recently been reported radiologically.

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Renal Cortical Necrosis in an Infant

Renal cortical necrosis is a rare and usually rapidly fatal disease, death resulting from acute renal failure. However, in recent years with the increasing availability of improved supportive measures, including peritoneal dialysis and haemodialysis, some of the patients survive long enough to allow calcification of the necrotic renal cortex which can be detected radiologically.

Up to 1968, 17 cases of renal cortical necrosis radiologically diagnosed ante mortem had been reported in adults, but only 3 in infants (Rémy et al., 1968; Whelan, Ling, and Davis, 1967). Mauer and Nogrady (1969) reported a newborn infant in whom papillary and cortical necrosis was docu-