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Mental Retardation, Cataracts, and Unexplained Hyperphosphatasia

We report a 4-year-old boy with a persistently raised serum alkaline phosphatase, 90% of which was of bone origin. There was no evidence of bone disease.

Case History

This boy was referred to the Department of Child Health of the Children's Hospital, Sheffield, for investigation of mental retardation associated with convulsions and bilateral cataracts at 4 years and 4 months of age. He was the first child of healthy, non-consanguineous parents. His mother had one fit in early childhood. There was a history of mild toxaemia, but not of rubella, during the pregnancy which terminated at 36 weeks' gestation. Birthweight was 1800 g., head circumference 39·7 cm. Though covered with thick offensive liquor at birth the child's condition was satisfactory till the fourth day of life, when he had a cyanotic attack. He made a good recovery after a course of penicillin given for presumed inhalation pneumonia.

The first year of life was uneventful, though his milestones were delayed. At 20 months of age he had his first convulsion; the CSF was normal and there was no hypoglycaemia. Anticonvulsant therapy was started. At 28 months he was found to have bilateral cataracts. During the third year of life he had two further convulsions. The seizures have always been right-sided with loss of consciousness and transient hemiparesis. During his third seizure the blood glucose was 15 mg./100 ml. The convulsions were becoming more frequent but with an increase in the dose of anticonvulsant they are now less severe, consisting of right-sided twitching, with no loss of consciousness or paresis.

Examination. Height 97·5 cm., weight 15·4 kg. Skull circumference 50 cm. His intelligence, assessed by the educational psychologist, was recorded as low normal. Hearing was normal, but vision poor due to cataracts. The cardiovascular, respiratory, and neurological systems were normal. The liver was firm with a smooth surface; a sharp border was palpable 3 cm. below the costal margin.

Blood. Hb, WBC, plasma urea, sodium, potassium, bicarbonate, and amino nitrogen, serum proteins, calcium, inorganic phosphates, bilirubin, thymol turbidity, thymol flocculation, glutamic oxaloacetic transaminase, glutamic pyruvic transaminase, 5 nucleotidase, galactose-1-phosphate uridyl transferase, and bromsulphalein excretion were normal. Alkaline phosphatase (on three separate samples) 376, 88, and 77 KA units/ml., respectively. Further analyses were done on the third sample. Alkaline phosphatase isoenzyme electrophoresis showed one band (Rf 0·56 ± 0·02). After incubation at 56 °C. for 15 minutes 90% was destroyed, indicating osseous origin.

Urine. No protein or reducing substances present. 24-hour urinary excretion of calcium, creatinine, and hydroxyproline were normal.

EEG. Gross abnormalities suggestive of *grand mal* epilepsy.

X-ray. Full skeletal survey showed no radiological abnormality.

Discussion

Normal adult serum contains predominantly alkaline phosphatase of liver origin, with a small but variable quantity of intestinal enzyme and little or no bone enzyme. In childhood and infancy there is in addition a bone iso-enzyme. Views differ as to the normal value for serum alkaline phosphatase in children; the range given by Gray (1965) was 15–20 KA units/ml., and that by King (1951) 10–30 KA units/ml. for children aged 1–3 years. Increases in the serum alkaline phosphatase may be secondary to bone or liver disease. The commoner bone diseases causing such increases are rickets, healing fractures including the battered baby syndrome, hyperparathyroidism, neoplasia with bone involvement, infantile cortical hyperostosis (Caffey's disease), and congenital hyperphosphatasia (Caffey, 1961).

The increase in the alkaline phosphatase level in this patient was chiefly attributable to bone isoenzyme. In spite of this, a full radiological skeletal survey showed no bony lesions. Though it is appreciated that enzyme changes may appear before bone change becomes evident, one would certainly expect some bone change and alteration in the

hydroxyproline level when the alkaline phosphatase level is of the order found in this patient. Further evidence of normal bone metabolism was obtained from the normal hydroxyproline excretion. There was no laboratory evidence to suggest hyperparathyroidism.

The normal galatose-1-phosphate uridyl transferase level ruled out galactosaemia. Though a diagnosis of glycogen storage disease was considered, no confirmatory laboratory evidence was found. The high serum alkaline phosphatase level in this boy has not been explained.

Summary

A 4-year-old boy with mental retardation and cataracts was found to have a grossly raised level of serum alkaline phosphatase, of which 90% was of osseous origin. There was no evidence of bone disease. The association with cataracts, convulsions, and mental retardation is as yet unexplained.

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Anaphylactoid Purpura with Cardiac Involvement

Anaphylactoid purpura (Schönlein-Henoch syndrome) is characterized by involvement of the skin, gastro-intestinal tract, joints, and kidney. Gairdner (1948) linked this syndrome clinically, pathologically, and aetiologically with rheumatic fever, acute nephritis, and polyarteritis nodosa, and described the co-existence of these diseases. However, serious cardiac involvement seems to be uncommon, and this report describes such a case.

Case Report

A 14-year-old girl was first seen medically two weeks after she had had a purpuric rash on the fronts of the legs, backs of the hands, buttocks, and abdominal wall. The eruption was soon followed by arthralgia and oedematous swelling of the major joints. She complained of colicky abdominal pain. There had been no previous sore throat.

Though her abdominal pain subsided promptly after administration of oral corticosteroid, her skin and joint symptoms gradually increased in severity. Frequent tarry stools and gross haematuria without hypertension were noted, and a few days later she developed a systolic cardiac murmur with low grade fever. Seven weeks after the onset of her illness she was transferred to this hospital.

On admission she was seriously ill, dehydrated, and undernourished. Temperature 37·8 °C., pulse regular 110/min., BP 124/70 mm. Hg. There was a widespread skin eruption with purpuric and erythematous lesions and pigmentation, especially on the back of the hands and fronts of the legs and feet. There was oedematous and painful swelling of the major joints. A high-pitched systolic cardiac murmur was heard at the apex.

Hb 7·4 g./100 ml., WBC 12,600/cu. mm., with 78% neutrophils and 2% eosinophils; platelets 210,000/cu. mm.; ESR 80 mm./hour; Serum urea nitrogen 17 mg./100 ml. Tourniquet test strongly positive. Tests for LE cells negative. ASO titre 1250 Todd units. C-reactive protein positive. Albumin 2·1 g./100 ml., total globulin 4·0 g./100 ml. (α_2-globulin 1·2 g./100 ml., γ-globulin 1·3 g./100 ml.). The immunoelectrophoretic studies showed slightly increased IgA and β_{1C}. The stools contained blood. Chest *x*-ray showed moderate enlargement of the left heart with clear lung fields (cardiothoracic ratio 0·53). ECG normal.

Three days after admission, she suddenly developed chest pain and fever, followed by a productive cough, dyspnoea, and cyanosis. Marked dullness was present at the lung bases. In addition, a gallop rhythm was heard at the apex, and moist râles were heard over the entire lung fields, indicating congestive cardiac failure. Chest *x*-ray showed conspicuous enlargement of the heart with a configuration suggestive of left heart failure and pulmonary congestion (cardiothoracic ratio 0·60).

The patient was promptly given deslanoside intravenously, followed by blood transfusion, oral corticosteroid, and penicillin. The clinical signs of congestive heart failure had mostly disappeared in a week but she still had the cardiac murmur and an abnormal ECG (see Fig. 1). She also had persistent gross haematuria and generalized purpuric eruption.

Her general condition improved over the next two weeks, the purpuric eruption, joint manifestations and gross haematuria subsiding. The cardiac sounds became normal.