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 galactose-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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References


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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. (α1-globulin 1·2 g./100 ml., γ-globulin 1·3 g./100 ml.). The immunoelectrophoretic studies showed slightly increased IgA and β1C. 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.
Two years after the onset of her illness she has remained well, except for persisting microscopical haematuria and proteinuria.

Biopsy of her skin showed hyperkeratosis of corium, perivascular infiltration of small vessels with mononuclear cells, and deposition of fibrinoid materials on the capillaries and small vessels (Fig. 2). A renal biopsy specimen showed abnormal glomeruli with endothelial hypercellularity, fibrinoid deposition and hyaline degeneration findings similar to those of Bergstrand, Bergstrand, and Bucht (1960) and Vernier et al. (1961). The lesions were occasionally associated with adhesions of the glomerular tuft to the capsule.

Discussion

Anaphylactoid purpura has been considered to link with rheumatic fever, acute nephritis, polyarteritis nodosa, and other so-called collagen diseases (Gairdner, 1948; Derham and Rogerson, 1952). However, judging from the literature, serious cardiac involvement in the syndrome seems uncommon. The ECG of our patient, showed progressive atrioventricular conduction disturbance and myocardial injury compatible with an acute carditis, such as may be seen in rheumatic fever. The raised ASO titre was also noteworthy.

Gairdner (1948) found clinical features characteristic of rheumatic carditis in two male cases with Schönlein-Henoch syndrome, one aged 15 years and the other aged 40 years. The former, 9 days after the onset of the syndrome, developed a typical rheumatic carditis and was left with an enlarged heart, mitral stenosis, and aortic insufficiency.
The latter had been at home convalescing from the syndrome; he then developed acute pharyngitis, and 18 days later he also developed rheumatic fever. Since this report, however, no other example of anaphylactoid purpura with cardiac involvement has been presented.

Summary
Anaphylactoid purpura was complicated by cardiac involvement in a girl of 14.

We are indebted to Professor N. Yamada for criticism and advice.

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Anaphylactoid purpura with cardiac involvement.

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