Weber-Christian panniculitis

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SUMMARY Two cases of Weber-Christian panniculitis with onset at 7 months and 3 years 6 months are described. Both show evidence of disturbances in immune mechanisms. The family of the first case has a high prevalence of coeliac disease, and the mother of the first patient also suffers from alopecia areata and dermatitis herpetiformis.

Key words: coeliac disease, immunological disease.

Systemic Weber-Christian panniculitis is a disease of unknown aetiology. The patient is seriously ill, suffering crops of sometimes tender subcutaneous lumps, which resolve to leave depressions in the skin contour. Constitutional symptoms may include fever, tiredness, weight loss, abdominal pain, nausea, vomiting, and diarrhoea. Reported cases in children describe a continuing relapsing course or death.

Weber-Christian panniculitis has been described in association with a wide variety of other disease states. Abnormalities of the immune system have been reported and suggested as central to its pathogenesis.

Case reports

CASE 1
Patient No 1 was referred at 7 months with a 2 cm × 3 cm area of induration on the lateral aspect of the right ankle, extending onto the dorsal surface of the foot. The lesion had a pale centre and was surrounded by erythema. A similar 1 cm × 1 cm lesion was noted on the lateral aspect of the left ankle.

During the following three weeks the presenting lesions resolved, with new areas of erythema appearing on the dorsum of the right wrist, the right forearm, left calf, and mandible. New activity was seen in the original sites, with fat atrophy in the hands and forearms, and dimples appearing over the back and buttocks (Fig. 1). He continued throughout to have a low grade pyrexia, rectal temperature 37.8–38.9°C. During his hospital admission persistent loose and bulky stools and failure to gain weight

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Fig. 1 Fat atrophy over back and buttocks.
caused added concern. His family were known to the hospital because of coeliac disease (Fig. 2). His mother, who also has dermatitis herpetiformis and alopecia areata, had been advised to maintain him on a gluten free diet for the first year.

**Investigations**

Haemoglobin 11 g/dl (110 g/l), mean cell volume (MCV) 73 fl, mean cell haemoglobin (MCH) 28 pg, total white cell count 17·4×10⁹/l, erythrocyte sedimentation rate (ESR) 12 mm/h. Serum iron 4·3 μmol/l, total iron binding capacity 78·8 μmol/l.

Plasma urea and electrolytes, liver function tests, amylase, glucose, triglyceride, and cholesterol values were normal.

Blood, urine, and stool cultures were negative. Antistreptolysin titre was 1/50. Serological tests were negative for *Yersinia enterocolitica*, *Y* pseudo-tuberculosis, cytomegalovirus, herpes simplex, rubella, and toxoplasma.

Serum IgG 184 IU/l (normal 64–146), serum IgM 224 IU/l (normal 30–74); serum IgA 42 IU/l (normal 14–38); C3 complement 240 mg% (2·4 g/l). Further analysis of the complement system showed Clq 120 mg/l, C2 72%, C3 0·92 g/l, C4 0·26 g/l, C5 to C9 and CH100 present within normal limits. Immune complexes were not detected. Total lymphocyte, T cell and B cell counts, and helper T/suppressor T cell values were normal.

Nitroblue tetrazolium test unstimulated: 3% (normal 1–10%), stimulated 99% (normal 90–100%).

Tetanus antitoxin level after one immunisation: 0·26 IU/ml (protective level 0·1).

HLA type A2, A11, B7, B8.

Antinuclear and rheumatoid factors, gluten and reticulin antibodies were negative. Smooth muscle antibody was positive.

Biopsy of jejunal mucosa showed moderate villous atrophy and crypt hyperplasia consistent with treated coeliac disease.

Eight weeks after the initial presentation a skin biopsy specimen was taken from an active area on the right foot (Fig. 3), and the diagnosis of Weber-Christian panniculitis made.

He was treated with prednisolone for a total of 6 months. The symptoms were controlled at a dose of 20 mg twice daily. Initial attempts to reduce this failed. Seven months after the onset of the illness, however, he was well. There has been no relapse to date.

**CASE 2**

Patient No 2 was first seen at 3 years 10 months. Four months previously he had complained of pain at the backs of his legs on sitting and was reluctant to walk. Crops of red indurated lesions developed on the anterior aspect of the lower legs and the extensor surfaces of the forearms. There was pr-
gressive muscle weakness and wasting. He was miserable, clinging, and febrile with a tense distended abdomen and episodes of colicky abdominal pain.

**Investigations**

Haemoglobin 9.2 g/dl (92 g/l), MCV 69 fl, MCH 21.1 pg, total white cell count $8.3 \times 10^9$/l, ESR 38 mm/h.

Plasma urea and electrolytes, liver function tests, and creatine kinase were normal. Antistreptolysin titre was 200 U/ml. Urine and stool cultures were negative.

Serum IgG 186 IU/l (normal 97–163); serum IgM 222 IU/l (normal 37–71); serum IgA 5 IU/l (normal 46–84); C3 complement 174 mg% (1.74 g/l). Further analysis of the complement system showed Clq 150 mg/l, C2 95%, C3 1.1 g/l, C4 0.39 g/l, C5 to C9 and CH50 present within normal limits. Immune complexes were not detected. Antinuclear and rheumatoid factors and antibody to smooth muscle were negative.

Electromyography of the right tibialis muscle showed an excess of brief polyphasic potentials during contraction, but the muscle was silent at rest.

A biopsy specimen of skin and subcutaneous tissue taken from an active area on the left thigh showed subcutaneous panniculitis, and a diagnosis of Weber-Christian disease was made. Muscle biopsy was normal.

He was treated with prednisolone. A dose of 25 mg/day with concurrent azathioprine 3 mg/kg/day have been necessary to control the disease activity.

**Discussion**

Disordered humoral immunity has been previously reported in Weber-Christian disease, as have manifestations of cellular immunodeficiency and defects in the alternate complement pathway. The underlying pathology has been described as an autoimmune disease of the adipose tissue, perhaps a component of a more severe and general autoimmune picture. The evidence is, however, inconsistent, with both absent and increased delayed hypersensitivity reactions reported, and both increased and lowered T lymphocyte numbers thought to be important in periods of remission.

Both cases described in this paper had high serum levels of IgG, IgM, and C3 complement at presentation. Further examination of the competence of the classic complement recognition and lytic pathway, however, showed no abnormality. No immune complexes were found. Screening investigations of the first case did not show any defects in humoral or cellular immunity or in neutrophil function.

It is of interest, nonetheless, that disturbances of immune mechanisms are believed to be responsible for the small intestinal damage in coeliac disease and are important in dermatitis herpetiformis and alopecia areata. We suggest that the occurrence of Weber-Christian disease in an infant with such a family history may be more than fortuitous, and that genetic factors and as yet undetected abnormalities of the immune system may be important in its pathogenesis.

**References**