Netherlands
The Journal of Medicine

Skin lesions depicting a systemic disease

L.A.A. Moonen[1]*, H. van den Bosch[2], T.B.J. Demeyere[3], B. Bravenboer[1]

Departments of [1]Internal medicine, [2]Radiology, [3]Pathology, Catharina Hospital, Eindhoven, the Netherlands, *corresponding author: tel.: +31 (0)40-239 91 11, fax: +31 (0)40-245 50 35, e-mail: linda.moonen@cze.nl

CASE REPORT

A 38-year-old woman with a previous medical history of diabetes mellitus type 2, and a hysterectomy, presented to our hospital because of acute renal failure for which temporary dialysis was necessary. She did not have any physical complaints. The patient was haemodynamically stable, and had no fever.

Physical examination revealed a large ulcerative process in the abdominal scar (*figure 1*) and multiple painful red nodules on the lower extremities (*figure 2*, this is also the biopsy site). Laboratory investigations showed the following results: haemoglobin of 9.8 mmol/l, white blood cell count 24.3 /nl, C-reactive protein 180 mg/l, creatinine 1100 µmol/l, urea 27 mmol/l, modification of diet in renal disease (MDRD) 3.0 ml/min, potassium 2.8 mmol/l, sodium 133 mmol/l, amylase of 59 U/l, and no antinuclear antibodies. Routine urine analysis showed no protein, erythrocytes or cylinders. Fractional sodium excretion was 0.2%. The chest X-ray was normal. A CT scan of the abdomen showed pseudocysts indicating chronic pancreatitis (*figure 3*). On repeated imaging, there were no signs of a pancreatic carcinoma. Biopsy of one of the painful nodules on her legs revealed a septal and lobular panniculitis (*figure 4*).

WHAT IS YOUR DIAGNOSIS?

See page 42 for the answer to this quiz

Figure 1. *Large ulcerative process in the abdominal scar*

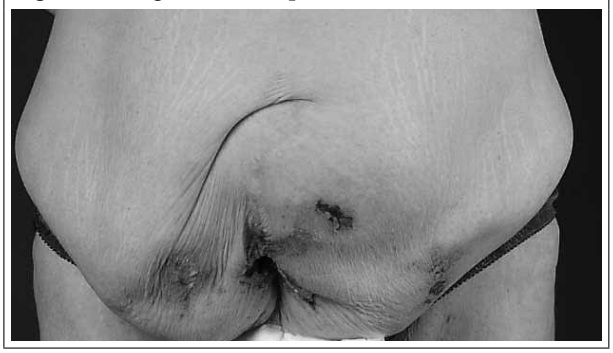


Figure 2. *Multiple painful red nodules on the lower extremities (biopsy site)*

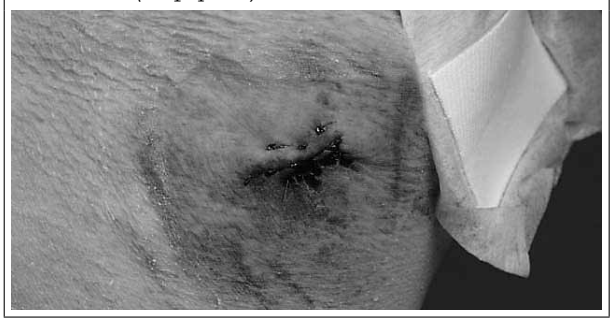


Figure 3. *CT scan showing signs of chronic pancreatitis*

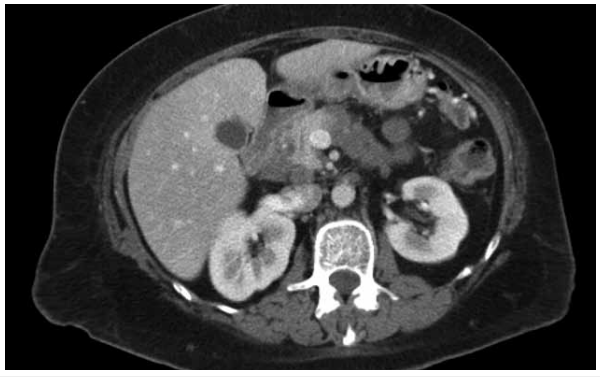
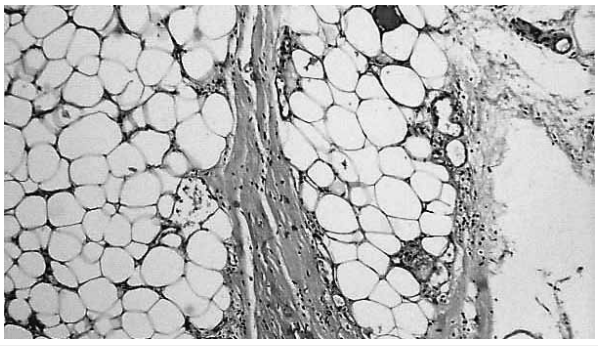


Figure 4. *Biopsy from one of the nodules revealing septal and lobular panniculitis*



ANSWER TO PHOTO QUIZ (PAGE 41)

SKIN LESIONS DEPICTING A SYSTEMIC DISEASE

The combination of the above-mentioned findings led us to the diagnosis of systemic panniculitis or Weber-Christian disease. Weber-Christian disease was initially described by Pfeiffer in 1892, and was established by Weber and Christian in the 1920s. It is an infiltrative inflammatory disease of fat tissue that usually occurs in young white females. It is characterised by tender skin nodules that are often but not always associated with constitutional symptoms, such as fever, arthralgias and myalgias.[1] Patients with Weber-Christian disease typically present with subcutaneous nodules on the extremities, but skin lesions can also occur over the posterior thorax, abdomwinal area, breasts, face or buttocks, and the panniculitis can also affect other organs. The disease can then present as a severe systemic illness leading to death from panniculitis involving the heart, lungs, liver, pancreas or kidneys.[1,2] The diagnosis is confirmed by excisional biopsy of a nodule. The panniculitis is typically lobular, although it may be both lobular and septal. This patient responded well to corticosteroid therapy.

REFERENCES

1. Wang HP, Huang CC, Chen CH, Lin HY. Weber-Christian disease presenting with intractable fever and periorbital swelling mimicking angioedema. Clin Rheumatol. 2007;26(6):1002-4.
2. Panush RS, Yonker RA, Dlesk A, Longley S, Caldwell JR. Weber-Christian disease. Analysis of 15 cases and review of the literature. Medicine. 1985;64(3):181-91.