A 23 year old white male with a 4 year history of Crohn's disease presented with an acute two day history of malaise, fever, abdominal pain, vomiting and stomal diarrhoea.
He complained of joint pains affecting the shoulders, elbows, wrists, metacarpophalangeals, knees and ankles.
There was also a rash on the elbows, ankles and feet, which began as erythematous macules and evolved to vesico-pustular lesions followed by crusting.
There was no history of sexual exposure or any intercurrent infection.
He was on no regular medication, but had discontinued Pentasa 4 months earlier.
One month earlier he had undergone a laparotomy to excise a complex ileo-cutaneous fistula with blind tracts, and two weeks earlier a defunctioning ileostomy had been created in view of persistent abdominal pain.
The cutaneous fistula had been present for a year, but was associated with a terminal ileal stricture and ileo-rectal fistula of at least 2 years duration.
His bowel disease had been resistant to immunosuppressive drugs including azathioprine, corticosteroiods and three infusions of Infliximab a year earlier.
There had been no extra-intestinal manifestations.
On admission to the hospital, he was thin, afebrile with a resting tachycardia of 125/minute.
The rest of the cardio-respiratory examination was normal.
The abdomen was minimally tender around the ileostomy without guarding or rebound tenderness.
Examination of the skin revealed some pustules and crusts around the elbows, ankles and feet (Fig 1 and 2).
The buttocks were spared.
There were clinical signs of synovitis of the wrists, proximal interphalangeal and metacarpophalangeal joints, and also both ankles.
Results of the laboratory tests showed a haemoglobin of 13.1 gm/dl, white blood count 15.8 × 109/L, platelets 585 × 109/L, C- reactive protein 37.7 mg/L and erythrocyte sedimentation rate 69 mm/hr.
Urea and electrolytes, complement, urine analysis and microscopy were normal.
Rheumatoid factor, antinuclear and antineutrophil cytoplasmic antibodies, cryoglobulins and Hepatitis B and C serology were negative.
Transthoracic echocardiogram showed no signs of endocarditis, and multiple blood cultures were sterile.
A skin biopsy from the ankle revealed a perivascular lymphohistiocytic infiltrate with prominent neutrophils and associated fibrinoid necrosis of vessels consistent with 'leukocytoclastic' vasculitis.
He was commenced on 60 mg prednisolone per day leading to prompt and complete resolution of all features.
The dose of prednisolone was rapidly tapered over the course of one month by the patient, faster than advised but without any recurrence over the following 3 years.