Administration of plasma in our C1q-deficient patient has clearly been therapeutically successful over a decade, demonstrating that this is a valid treatment option in this difficult condition.

Rheumatology key message

- Chronic plasma infusion is an effective therapy in complement C1q deficiency.

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Treatment of a patient with remitting seronegative, symmetrical synovitis with pitting oedema using a humanized anti-interleukin-6 receptor antibody, tocilizumab

Sir, RS3PE is an inflammatory disorder of unknown aetiology that affects elderly persons [1]. The clinical features of the disease are characterized by bilateral pitting oedema of the hands and sudden onset of polyarthritis [1]. Although the pathogenesis of RS3PE remains unknown, over-production of IL-6 has been demonstrated to contribute to its development [2, 3]. Corticosteroids constitute the preferred treatment for RS3PE and in most cases of RS3PE the quick and dramatic response to corticosteroids is one of the characteristics of the disease. In our
A 51-year-old female, who presented in 2001 with fever, bilateral pain and swelling of the knee joints, aching in the shoulders and pitting oedema in the hands and feet, was admitted to our hospital. Laboratory test findings were: white blood cell 5990 cells/µl with 87% granulocytes; Hb 7.8 g/dl; platelets 428 000 cells/µl; ferritin 246 ng/ml; and CRP 19.3 mg/dl. Repeated culture test results for blood, urine and sputum were negative. CT and ultrasound examinations showed swelling of the gallbladder, and Ga-citrate scintigraphy showed uptake in the bilateral shoulders and knee and hand joints. ANA, RF and autoantibodies, including anti-DNA, anti-ENA and ANCA, were all negative. These symptoms, laboratory and instrumental findings led to a diagnosis of RS3PE, for which treatment with prednisolone was started. We initially treated the patient with 40 mg/day of prednisolone due to the severe inflammatory findings. Fever and oedema in the bilateral hands and feet promptly disappeared and knee arthralgia and aching in the shoulders diminished. CRP had become normal 10 days later and prednisolone could be tapered. However, when prednisolone was tapered to 7.5–10 mg/day, the disease flared up again with exacerbation of shoulder pain, and oedema in the feet and she had complications with hypertension and osteoporosis. In 2007, this patient underwent a surgical operation for artificial bone replacement of the left femoral head because of bone necrosis. During the clinical course, RF and anti-cyclic citrullinated antibody were negative and the patient did not fulfill the ACR classification criteria for RA. Since IL-6 has been shown to play a role in the development of RS3PE [2, 3] and her serum IL-6 concentration was 7.8 pg/ml, informed consent by the patient and approval by the Ethics Committee of Osaka University Hospital were obtained for the injection of tocilizumab at 8 mg/kg every 4 weeks, starting from November 2008 in conjunction with oral administration of methylprednisolone (6 mg/day; Fig. 1A). Prior to the treatment, the patient felt morning stiffness for 4 h as well as pain in the bilateral shoulders and in the right hand. Serum levels of CRP, serum amyloid A (SAA) and MMP3 were elevated to 0.95 mg/dl (normal value <0.2 mg/dl), 55 µg/ml (normal value <8 µg/ml) and 636 ng/ml (normal value 37–121 ng/ml), respectively. Ga-citrate scintigraphy showed uptake in the bilateral shoulders, hands, and
knee and ankle joints (Fig. 1B). After one injection of tocilizumab, serum CRP and SAA levels became normal and morning stiffness and shoulder pain improved. By March 2009, a total of five infusions of tocilizumab had been administered without any exacerbation of symptoms or any elevation of serum CRP or SAA levels. MMP3 reduced from 508-727 to 334 ng/ml. Ga-citrate scintigraphy also showed a marked reduction of uptake in the bilateral shoulders and hands, and in the left ankle joint (Fig. 1B). However, just before the sixth administration, cholecystitis occurred and tocilizumab treatment had to be stopped. At 3 months after the cessation, the disease activity flared up with shoulder pain and morning stiffness, leading to an increase in the methylprednisolone dose from 6mg/day to 8mg/day.

In this report, we demonstrated the ameliorative effect of tocilizumab on symptoms caused by RS3PE. To the best of our knowledge, this is the first report to evince the efficacy of tocilizumab for RS3PE. A response to low-dose corticosteroids and absence of relapse after 2 years of treatment are characteristics of RS3PE [5] but our patient was refractory to corticosteroids and then the present case was thought to be a rare one. Increased serum concentration of IL-6 has been observed in patients with RS3PE [2, 3], and therefore IL-6 inhibition with tocilizumab might constitute a novel strategy for treatment of RS3PE. Indeed, reported here, tocilizumab treatment resulted in a remarkable suppression of clinical symptoms, accompanied by a reduction in MMP3 levels as well as in Ga-citrate uptake in joints. Although tocilizumab treatment had to be discontinued due to the complication of cholecystitis in the patient, tocilizumab can be considered a viable option for treatment of refractory RS3PE.

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Tropical rheumatology in a UK District General Hospital: a case report of leprosy presenting as acute vasculitis

Sir, In their recent editorial, Adeba et al. highlighted the need for increased awareness of tropical illnesses to avoid problems of misdiagnosis and mismanagement [1]. Leprosy is a medical mimic and should be suspected in patients from leprosy-endemic areas, even in the absence of classical cutaneous features [2]. We wish to report a case of lepromatous leprosy, presenting as primary vasculitis to a UK District General Hospital.

A 23-year-old, previously well, Brazilian woman, presented to the general physicians with a 10-day history of sudden onset of polyarthritis, unresponsive to treatment with ibuprofen, and a diffuse necrotizing rash. She had lived in the UK for the past 4 years, with no recent travel abroad. Examination revealed fever (37.9°C), tachycardia (110 beats/min), tender axillary and inguinal lymphadenopathy and a symmetrical synovitis of elbows, wrists, ankles and MCP joints. She had widespread tender erythematous nodules over her face, arms and legs with truncal sparing (Fig. 1). There was an area of incipient necrosis over her right cheek.

Initial investigations revealed raised CRP of 191.8 mg/l (<6 mg/l), aspartate transaminase 180 IU/l (7–35 IU/l) and γ-glutamyl transpeptidase of 90 IU/l (5–50 IU/l). Routine biochemistry and full blood count was otherwise normal.

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