ably, vasculitis might lead to capsule and ligament deformity and, thereby, to Jaccoud's deformity. This possibility deserves to be studied. We suggest the acronym "JAVS" to designate this syndrome.

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Sustained 3-year remission after rituximab treatment in a patient with refractory Wegener's granulomatosis

Sirs,
A 37-year-old woman was admitted in March 2001 because of purpuric rash, diarrheas, arthralgias and edema of the lower legs. Laboratory examinations revealed: Hb: 10.2 g/dl, WBC: 9000/mm^3, PLT: 295000/ mm^3, ESR: 68 mm/h, CRP: 2.5 mg/l (normal range 0-0.5), creatinine: 1.1 g/l, liver enzymes: normal. Urine tests showed 30-40 RBC/hpf and 4-5 WBC/hpf. A 24h urine collection yielded 6.2 gr of protein. Blood, urine and stool cultures were negative. Antinuclear and anti-DNA antibodies, ANCA by indirect immunofluorescence and cryoglobulins were negative. Chest X-Ray was normal. A kidney biopsy showed segmental glomerulonephritis with necrotic and crescentic changes and subtle IgA and IgM deposits. Skin biopsy revealed leukocytoclastic vasculitis. A preliminary diagnosis of small vessel vasculitis was made and the patient received treatment with intravenous cyclophosphamide 1 gr and 3 pulses of methylprednisolone 1 gr daily followed by oral methylprednisolone 24 mg daily. Fever, diarrheas and skin rash improved. One month later, treatment with mucophenolate mofetil 2 gr daily was started. In August 2001 the patient was readmitted because of fever, cough and purpuric rash. Chest X-Ray showed pulmonary nodules bilaterally and a 4-cm cavitary lesion in the right lower lobe. Infectious causes were excluded. Mucophenolate mofetil was stopped and monthly pulses of cyclophosphamide 1gr/m^2 in combination with oral methylprednisolone 32 mg daily were started. Two months later she complained again for fever and was admitted to our hospital. Physical examination showed a deep mouth ulcer, palpable purpura on both legs and 3 skin ulcers with a diameter of 2-3 cm. Anemia, high ESR and CRP values, microscopic hematuria and 24-h proteinuria of 1 gr were detected. ANCA were found negative by indirect immunofluorescence as well as by ELISA. A diagnosis of Wegener's granulomatosis was made on the evidence of lung nodules and cavitary lesion, necrotizing glomerulonephritis, mouth ulcers and leukocytoclastic vasculitis of the skin. The patient received therapy with oral cyclophosphamide 100 mg daily, methylprednisolone 40 mg daily and trimethoprim-sulfamethoxazole 960 mg twice daily with gradual improvement of her symptoms.
Five months later, she relapsed with nephrotic range proteinuria, red blood cell casts and a 3-cm diameter skin ulcer of the left leg. Cyclophosphamide treatment was discontinued. She received 4 weekly infusions of 375 mg/m^2 of rituximab as previously described (1) while the dosage of methylprednisolone remained stable at 16 mg daily. No side effects were noticed. Two months later she was in complete remission regarding the Birmingham Vasculitis Activity Score modified for Wegener (2). Currently, 3 years after rituximab treatment, she remains in full remission receiving treatment with methylprednisolone 1 mg every other day and trimethoprim-sulfamethoxazole 960 mg daily.
This is a case of refractory Wegener's granulomatosis treated effectively with anti-CD20 therapy. Our patient had segmental necrotizing glomerulonephritis, lung nodules and cavitary lesions, mouth ulcers and leukocytoclastic vasculitis but negative ANCA. Cytoplasmic ANCA and proteinase 3 reactions have 98% specificity in Wegener's granulomatosis, however, their sensitivity varies from 30 % to 99 % depending of the extent, severity and activity of the disease.
Anti-CD20 therapy has been used in several autoimmune diseases the last years (3-5). The successful use of B lymphocyte depletion therapy in Wegener's granulomatosis has also been recently described (1, 6-8). Remission was maintained as long as B lymphocytes remained undetectable. In almost all the above cases, the duration of follow-up period was approximately 1-1.5 years (1, 6-8). In our patient with refractory Wegener's granulomatosis and negative ANCA, remission persisted for 3 years after rituximab treatment. This case suggests that the use of B cell depletion therapy may induce sustained remission in refractory Wegener's granulomatosis.

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