enlarged. Multiple enlarged lymph nodes were present in the hilus of the liver and spleen and in the retroperitoneum. The weight of the removed spleen was 3450 g. Histopathological findings showed atypical paracortical immunoblastic and plasma cell proliferation in the lymph nodes, with follicular hyperplasia, sinus histiocytosis, dilated lymphatic sinuses and hilar haemorrhage. Liver biopsy showed reactive B- and T-cell proliferation. The molecular genetic investigation of liver, spleen and lymph nodes revealed no dominant monoclonal band. T-cell receptors, both gamma and beta, and immunoglobulin heavy-chain polymerase chain reaction showed no dominant monoclonal band. Bcl-2 staining of all samples was negative. The final diagnosis was plasma cell variant and multicentric subtype of Castleman's disease.

Standard X-ray of hips was normal and revealed no exostoses.

Following splenectomy, the skin lesions significantly regressed, but subsequently reappeared and ascites progressed. Prednisone 1 mg/kg was started, which resulted in complete regression of the skin lesions and regression of the volume of ascitic fluid. Later, azathioprine was combined with low-dose prednisone and the skin changes and ascites disappeared completely.

Our patient had many features of Schnitzler's syndrome, i.e. urticarial vasculitis, hepato-splenomegaly, lymphadenopathy, and a polyclonal gammopathy.[2] However, neither bone pain nor radiological bony exostotic changes were present. Six of the 72 patients with urticarial vasculitis analysed by Mehregan *et al*.[3] had a haematological disease, two of whom had myelodysplastic syndrome, one had Hodgkin's lymphoma in remission, one, acute myeloid leukaemia in remission, one, acute myelogenous leukaemia, and one had idiopathic thrombocytopenic purpura.

Castleman's disease (angiofollicular lymph node hyperplasia or giant lymph node hyperplasia) is part of a group of rare and distinctive types of benign atypical lymphoproliferative disorders sharing characteristic histopathological and clinical features.[4]

The skin symptoms of our patient disappeared temporarily when the large mass of lymphatic tissue was removed by splenectomy. This suggests that the occurrence of urticarial vasculitis in our patient might be related to Castleman's disease. Twenty-three of the 72 patients with urticarial vasculitis presented by Mehregan *et al*.[3] had hypocomplementemia, as did our patient. It can be hypothesized that plasma cells were triggered by unknown antigen(s) to produce polyclonal immunoglobulins, which were then deposited in the vessel walls in the skin, causing activation of the complement cascade and leading to the development of vasculitis.

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References

1 Toussaint S, Kamino H. Non-infectious erythematous, papular and squamous diseases. In: Elder D, Elenitsas R, Jaworsky C, Johnson B Jr, eds. *Lever's Histopathology of the Skin*, 8th edn. Lippincott-Raven publishers, Philadelphia, 1997: 151–152.

2 Puddy P, Cianchini G, Girardelli CR *et al.* Schnitzler's syndrome: report of a new case and review of the literature. *Clin Exp Rheumatol* 1997; **15**: 91–95.

3 Mehregan DR, Hall MJ, Gibson LE. Urticarial vasculitis: a histopathological and clinical review of 72 cases. *J Am Acad Dermatol* 1992; **26**: 441–448.

4 Martino G, Cariati S, Tintisona O *et al.* Atypical lymphoproliferative disorders: Castleman's disease. Case report and review of the literature. *Tumori* 2004; **90**: 352–355.

DOI: 10.1111/j.1468-3083.2006.01941.x

Treatment of bullous pemphigoid with enteric-coated mycophenolate sodium

Editor

Mycophenolate mofetil (MMF) is an immunosuppressive drug that has recently been used to treat autoimmune and inflammatory skin diseases.[1,2] Mycophenolate sodium is enteric coated and has been suggested as a potential method to reduce the gastrointestinal adverse events seen with mycophenolate mofetil.[3] We herein report the first case of bullous pemphigoid (BP) successfully treated with enteric coated-mycophenolate sodium (EC-MS), treatment of which failed with adjuvants such as azathioprine and IVIG.

A 78-year-old female patient presented with a 1-year history of intense blisters on the limbs and abdomen (fig. 1). Nikolsky's sign was negative. Conjunctiva, scalp and mouth were normal. Previous systemic steroid treatments (methyl prednisolone 48 mg/day for 1 month, deflasocort 90 mg/day for 2 weeks) resulted in either no improvement or transient, mild improvement. The results of routine laboratory analysis and IgE level were within normal limits. Systemic review revealed no obvious malignancy. A skin biopsy specimen revealed subepidermal cleavage with a mixed dermal infiltrate of polymorphonuclear leucocytes,

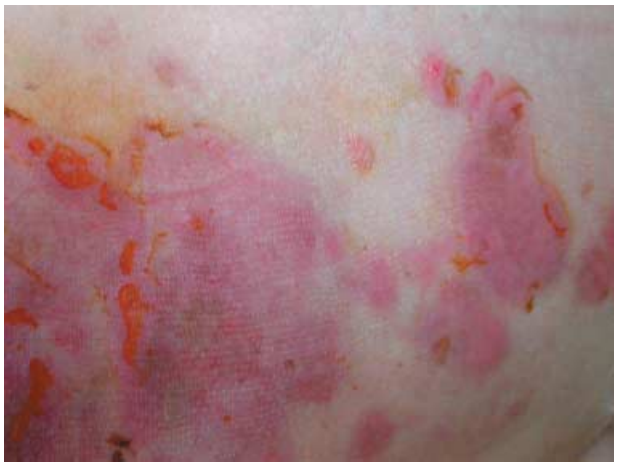

fig. 1 Pruritic erythematous lesions and erosions on the buttock before treatment.

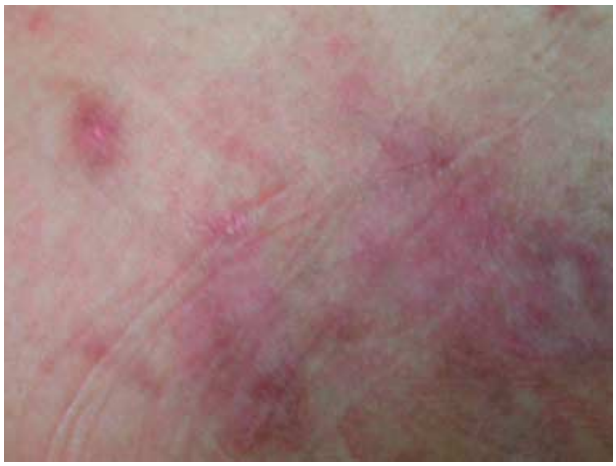

fig. 2 Clinical healing at the third month of treatment.

lymphocytes and eosinophils. Histopathological findings were in accordance with BP. Direct immunofluorescence of perilesional skin showed smooth linear deposits of C_3 and IgG along the dermoepidermal junction. Based on clinicopathological and laboratory findings, a diagnosis of BP was made. A treatment with prednisolone, 100 mg (1 mg/kg) and azathioprine 150 mg daily was started. After 3 weeks, although previous lesions started to heal, new blisters continued to erupt. Therefore, high-dose intravenous immunoglobulin (IVIG) at a dose of 0.1 g/kg daily for five consecutive days in a week was started, but new blister formation was still present after 2 weeks. Azathioprine and IVIG were discontinued. EC-MS (Myfortic®, Novartis Pharma AG, Basel, Switzerland) 360 mg twice a day was commenced in addition to prednisolone, 100 mg/day. By the eighth week, the lesions had completely resolved. Steroid therapy was gradually discontinued after 2 months. EC-MS was continued and tolerated without any side-effects for 3 months. She has now been clear of bullae for more than 6 months (fig. 2).

BP is a subepidermal autoimmune disease presenting as papulovesicles and intense blisters.[4] Current treatments include systemic and topical steroids, dapsone, IVIG, topical tacrolimus, minocycline, tetracycline and nicotinamide, immunosuppressive agents such as azathioprine, MMF, cyclophosphamide, and methotrexate.[5–7] MMF is the morpholinoethylester prodrug of mycophenolic acid, an agent that inhibits the proliferation of B and T lymphocytes through non-competitive, reversible inhibition of inosine monophosphate dehydrogenase, a key enzyme in the de novo synthetic pathway of guanine nucleotides.[1,2] Some experiences suggest that MMF monotherapy or combination with systemic steroids may be effective for patients with bullous pemphigoid.[8,9]

Recently, it has been shown that mycophenolate sodium is effective in preventing acute rejection in renal transplant recipients.[3,10] Enteric-coated mycophenolate sodium (EC-MPS; Myfortic®, Novartis Pharma AG, Basel, Switzerland) is an advanced formulation delivering mycophenolic acid (MPA). Optimal MMF therapy may be limited by gastrointestinal (GI) intolerance, which may result in the need for MMF dose reduction, interruption, or discontinuation, leading to an increased risk of acute rejection. EC-MPS is a new formulation delivering mycophenolic acid, developed with the aim of improving upper GI tolerability.[3] To our knowledge, our patient illustrates the first case of BP successfully treated with EC-MS. The treatment of our patient demonstrates a novel therapeutic option for patients with autoimmune bullous dermatoses; EC-MS treatment may be preferable to azathioprine and MMF treatment because it has a safer adverse effect profile. However, larger studies must be performed to establish the risk–benefit ratio of various therapeutic dosages of EC-MS for these patients.

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References

1 Lipsky JJ. Mycophenolate mofetil. *Lancet* 1996; **348**: 1357–1359.

2 Epinette WW, Parker CM, Jones EL, Greist MC. Mycophenolic acid for psoriasis: a review of pharmacology, long-term efficacy, and safety. *J Am Acad Dermatol* 1987; **17**: 962–971.
3 Gabardi S, Tran JL, Clarkson MR. Enteric-coated mycophenolate sodium. *Ann Pharmacother* 2003; **37**: 1685–1693.
4 Walsh SR, Hogg D, Mydlarski PR. Bullous pemphigoid: from bench to bedside. *Drugs* 2005; **65**: 905–926.
5 Yeh SW, Ahmed B, Sami N, Razzaque AA. Blistering disorders: diagnosis and treatment. *Dermatol Ther* 2003; **16**: 214–223.
6 Ko MJ, Chu CY. Topical tacrolimus therapy for localized bullous pemphigoid. *Br J Dermatol* 2003; **149**: 1079–1081.
7 Goon AT, Tan SH, Khoo LS, Tan T. Tetracycline and nicotinamide for the treatment of bullous pemphigoid: our experience in Singapore. *Singapore Med* 2000; **41**: 327–330.
8 Ratnam KV. Adjuvant therapy of bullous pemphigoid with mycophenolate mofetil: old drug, new use. *Singapore Med J* 2001; **42**: 482–484.
9 Grundmann-Kollmann M, Kaskel P, Leiter U *et al.* Treatment of pemphigus vulgaris and bullous pemphigoid with mycophenolate mofetil monotherapy. *Arch Dermatol* 1999; **135**: 724–725.
10 Budde K, Curtis J, Knoll G *et al.* Enteric-coated mycophenolate sodium can be safely administered in maintenance renal transplant patients: results of a 1-year study. *Am J Transplant* 2004; **4**: 237–243.

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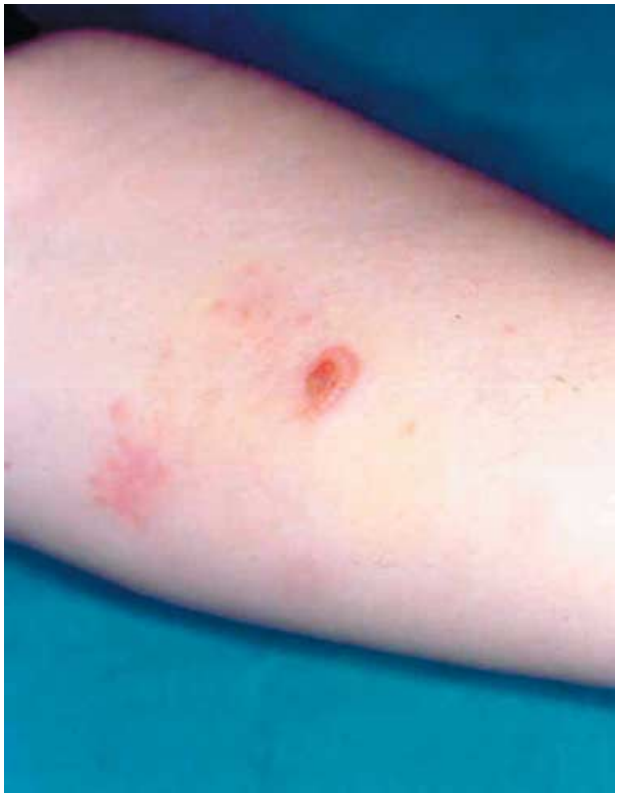

fig. 1 Erythematous plaque with vesicles on the right forearm.

Localized linear IgA disease associated with monoclonal gammapathy of undetermined significance

Editor

Linear IgA bullous dermatosis (LABD) is a rare subepidermal disorder characterized by linear IgA deposits. LABD as a solitary localized lesion has only been described twice.[1,2] We present a 46-year-old woman with a 3-month history of an asymptomatic plaque of slow enlargement on her right forearm. She was diagnosed with hypertension 2 years earlier and had been receiving enalapril since then. Physical examination revealed an erythematous plaque located on her right forearm, 6 × 3 cm in diameter, with vesicles (fig. 1). A biopsy revealed a subepidermal blister with a dermal infiltrate composed of neutrophils, scattered eosinophils and some lymphocytes (fig. 2). Direct immunofluorescence showed a linear IgA deposition at the basal membrane zone. Enalapril was discontinued for 1 month without improvement. She then received dapsone 100 mg a day with rapid improvement after

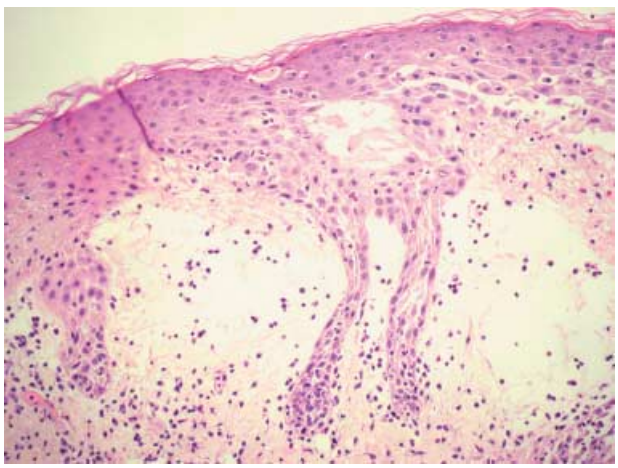

fig. 2 Subepidermal blister with an inflammatory infiltrate.

1 month's treatment and complete healing of the lesion 11 months later, after which dapsone was discontinued. Full blood count, biochemical analysis, urine analysis and erythrocyte sedimentation rate were normal. Anti-ANA and anti-DNA were negative. Serum protein electrophoresis showed normal gamma globulin levels but immunoelectrophoresis revealed the presence of a monoclonal component