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CORRESPONDENCE

Earlier continuous administration of mepolizumab for eosinophilic granulomatosis with polyangiitis based on cutaneous findings

Treatment of eosinophilic granulomatosis with polyangiitis (EGPA) remains a challenge because currently available therapies, corticosteroids, and immunomodulators, do not always control symptoms and are often associated with significant morbidity and relapse. We previously proposed that the initial cutaneous clinical and histopathologic findings based on skin biopsy and peripheral blood findings, as hallmark manifestations of EGPA, could prompt dermatologists to consider this diagnosis at an earlier stage.[1] Based on these cutaneous findings, we also reported that earlier adjunct administration of mepolizumab and intravenous immunoglobulin therapy (IVIG) led to significant improvement in EGPA symptoms.[2] Since those initial reports, we have been able to maintain good control of the disease, allowing us to taper their prednisolone dosage without any negative impact on the improvement of these symptoms or any significant adverse effects related to the drugs during a 3-year follow-up.

A 55-year-old man presented with erythematous nodules and livedo racemosa with purpura on his leg edema. There were clinical signs of mononeuritis multiplex as evidence of peripheral neurologic involvement. Microscopic examination of the indurated nodules revealed necrotizing vasculitis in the lower dermis and subcutaneous fat. There was a predominance of eosinophil infiltration into the dermis around the vascular walls and nerve fiber. He had been treated with oral prednisolone and intravenous cyclophosphamide pulse therapy (IVCY). The therapy did not resolve his symptoms, including the associated multiple mononeuritis. We administered IVIG, 400 mg/kg for 5 days, and mepolizumab 300 mg subcutaneously every 4 weeks to address concerns of exacerbation of complications such as peripheral neuropathy. The symptoms gradually improved, with resolution of the peripheral eosinophilia and normalized IgE levels. Azathioprine was additionally administered from the 6th month as prednisolone was tapered smoothly. To date, he has not developed any additional vasculitis symptoms during the 3-year follow-up (Figure 1A).

A 30-year-old woman presented with slightly purpuric skin lesions on her lower extremities. Nerve conduction tests revealed a mononeuritis multiplex on her lower extremities. Microscopic examination of skin biopsy specimens obtained from the purpura revealed

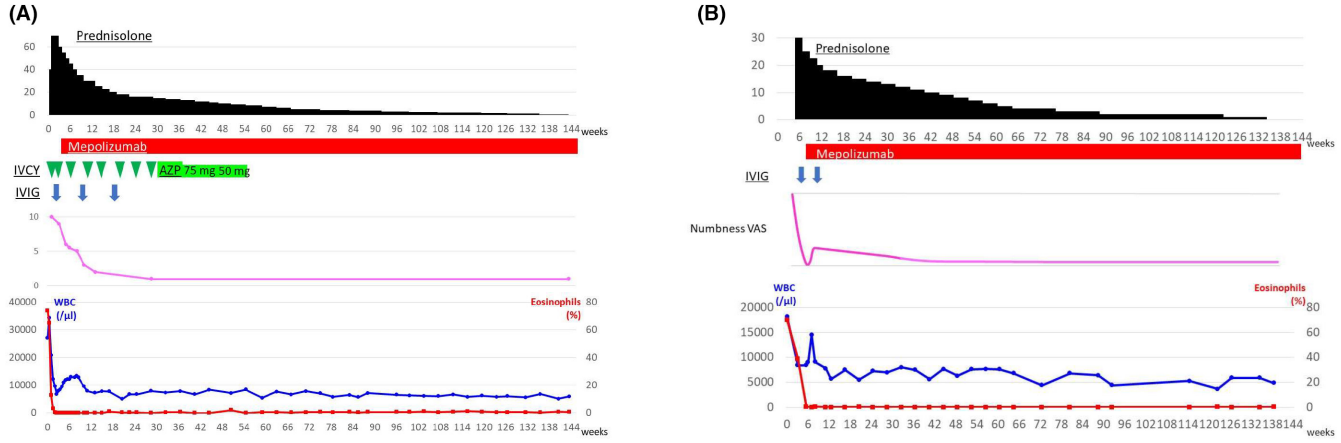

FIGURE 1 Manifestation of the patient's clinical symptoms and laboratory findings (A: 55-year-old man; B: 30-year-old woman). AZP, azathioprine; IVCY, intravenous cyclophosphamide therapy; IVIG, intravenous immunoglobulin therapy; VAS, visual analog score; WBC, white blood count.

leukocytoclastic vasculitis. There was a predominance of eosinophils infiltrating the dermis around the nerve fiber. We applied an adjunct combination therapy of IVIG and mepolizumab. After two courses of this adjunct therapy, the cutaneous manifestations, as well as the arthritis and mononeuritis multiplex had been completely resolved along with the normalization of peripheral eosinophilia, without any remarkable adverse effects related to prednisone such as opportunistic infections. Prednisolone was subsequently tapered smoothly. After 3 years, there has been no evidence of clinical recurrence or adverse effects of the systemic corticosteroids (Figure 1B).

The most important objective in the treatment of EGPA is to induce long-term remission and reduce the burden of systemic glucocorticoids and immunosuppressive therapies. Some studies have reported on the ability of mepolizumab to induce remission, prevent relapses, and allow a reduction in glucocorticoid dose.[3–5] We propose that earlier continuous administration of mepolizumab could be useful in controlling relapse and refractory disease, and can reduce the need for less desirable treatment protocols such as long-term corticosteroids.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

ETHICS STATEMENT

Approval of the research protocol: Yes. N/A?
Informed Consent: Yes.
Registry and the Registration No.: N/A.
Animal Studies: N/A.

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