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A Case of Myositis Preceding Neuropathy in Eosinophilic Granulomatosis with Polyangiitis

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Dear Editor,

Eosinophilic granulomatosis with polyangiitis (EGPA; also called Churg-Strauss syndrome) is a necrotizing vasculitis affecting small-to-medium-size vessels of multiple organs. Most patients exhibit respiratory involvement, such as pulmonary infiltrates, allergic rhinitis, or asthma. The nervous system is also commonly affected, most frequently as a mononeuritis multiplex.[1] Myalgia may appear during the course of EPGA,[2] but it is rarely a major complaint. Muscle weakness is typically attributed to peripheral neuropathy rather than to primary myositis, resulting in muscle biopsies rarely being performed. However, we recently experienced a case of EGPA that showed inflammatory myopathy that preceded the onset of peripheral neuropathy.

A 78-year-old male presented with myalgia and muscle weakness in the thighs and calves that had first appeared 3 weeks previously, and had been accompanied by fever for the previous 1 week. He was unable to walk without support. The power in hip flexion was Medical Research Council (MRC) grade 3 (of 5), while it was MRC grade 4 for ankle dorsiflexion on both sides. Myalgia was most prominent in the posterior thighs and calves. He complained of paresthesia over his legs, but did not show any primary sensory deficit.

He had been treated for asthma for 5 years with leukotriene-receptor antagonist, xanthine, inhaled corticosteroid, beta-2 agonist, and muscarinic antagonist. He had suffered an exacerbation of asthma with eosinophilia 2 years previously, which was alleviated by a short course of systemic corticosteroid.

His peripheral white blood cell count was high, at 37.96×10^9/L (normal, $4.0-11.0\times10^9$/L) with marked eosinophilia at 59% (normal, 0–6%). Abnormal serologic results included elevated creatine kinase (559 U/L; normal, <171 U/L), rheumatoid factor at 90 U/mL, and immunoglobulin E at 1,500 U/mL (normal, <87 U/mL). There was positivity for perinuclear antineutrophil cytoplasmic antibody. Blood, stool, and urine analyses for parasites produced negative findings.

Muscle MRI revealed diffuse symmetric T2-weighted signal intensities with mild gadolinium enhancement in the muscles of both thighs (Fig. 1A and B). Chest CT revealed diffuse emphysema in both lungs, small areas of subpleural consolidation, and ground-glass appearance in the lateral segment. Multiple foci of temporary pulmonary infiltrates had been detected in chest CT images over the past 4 years (Fig. 1C). Brain CT revealed maxillary sinusitis.

A nerve conduction study was unremarkable despite significant leg weakness, with electrophysiologic parameters of the superficial peroneal and sural nerves being within the normal limits. Needle electromyography revealed abnormal spontaneous potentials in the first dorsal interosseous, biceps brachii, medial gastrocnemius, and tibialis anterior muscles, which were suggestive of an active myopathy.

A biopsy sample from the right vastus lateralis showed marked perivascular infiltrations of inflammatory cells that extended to the nearby endomysium (Fig. 1D). Moderate varia-

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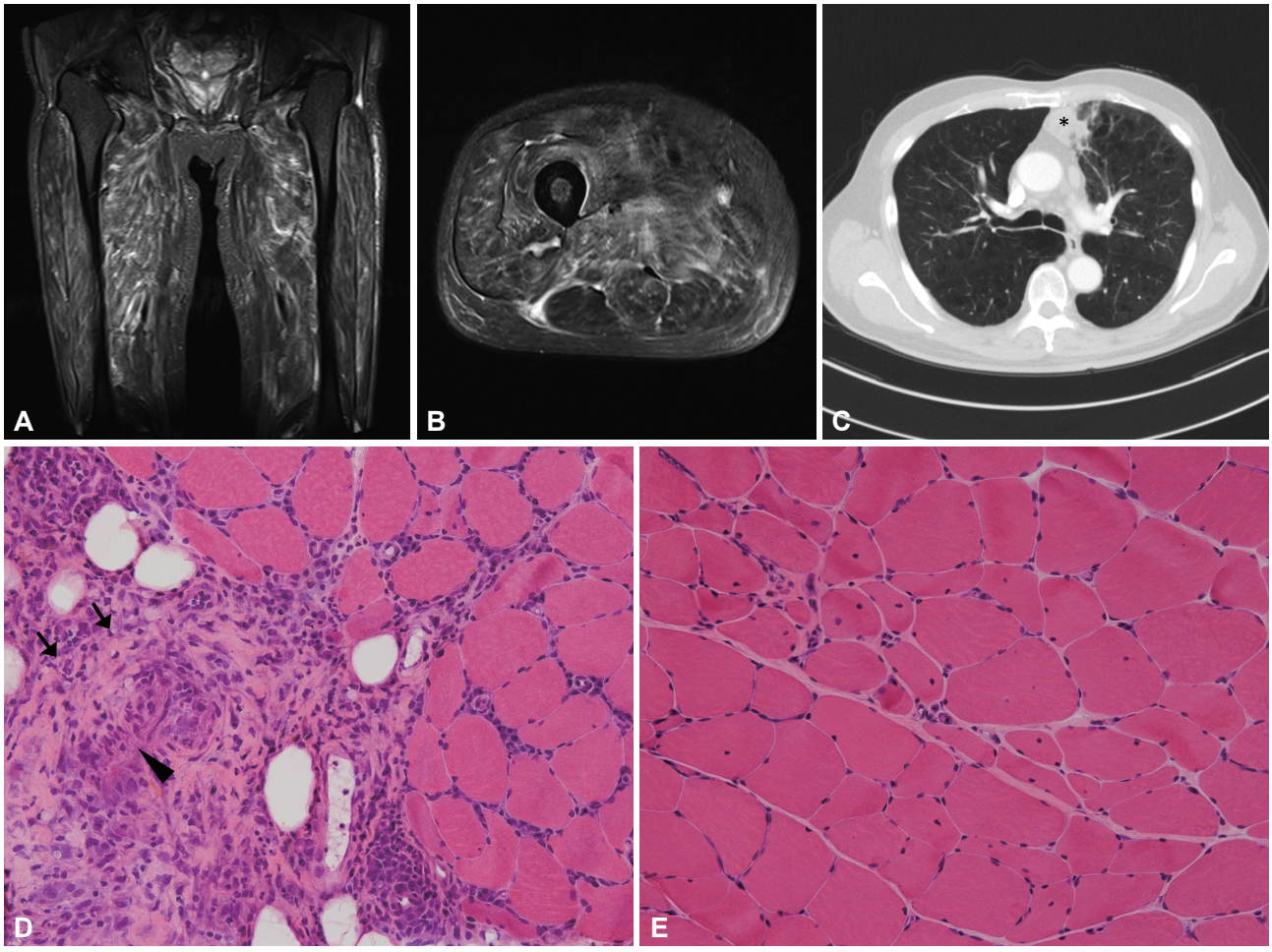

Fig. 1. Muscle imaging (A and B), chest CT (C), and muscle pathology (D and E: hematoxylin and eosin stain; magnification, ×200). A and B: T2-weighted MR images of the thigh muscle showing diffuse inflammation (A, coronal; B, axial). C: Subpleural consolidation is evident in the left lingular segment (asterisk). D: Muscle pathology reveals predominantly perivascular infiltration of inflammatory cells, an obliterated vessel lumen (arrowhead), and the thickening of vessel walls (arrows). E: Regions away from the perimysium display nonspecific myopathy with scattered small atrophic fibers and increased internalized nuclei.

tion in the muscle fiber size was accompanied by the random appearance of small atrophic fibers and internalized nuclei (Fig. 1E). Most of the muscle tissue that was away from vasculitic foci appeared normal.

He was treated with high-dose intravenous methylprednisolone and 1 g daily for 5 days, followed by oral prednisolone at 60 mg daily. The myalgia was alleviated from the fourth day of treatment, while the muscle weakness progressed to the ankles. Weekly oral methotrexate was added thereafter.

After 2 months, the laboratory findings of eosinophilia and the elevated creatine kinase and the erythrocyte sedimentation rate were normalized. However, ankle dorsiflexion and plantarflexion did not improve. A nerve conduction study at that time revealed marked axonal polyneuropathy affecting all of the tested nerves of the upper and lower limbs.

The present case depicts a rare episode of EGPA presenting as a myositis with proximal-dominant weakness, followed by typical peripheral neuropathy. Diagnosis of EGPA was fulfilled based on the presence of asthma, eosinophilia, pulmonary infiltrates, paranasal sinusitis, and eosinophilic vasculitis. There have been few case reports of inflammatory myopathy in EGPA, and only one recurrent myositis case included a detailed description on muscle pathology.[3]

MR images together with muscle pathology in our case confirms the presence of vasculitis in skeletal muscle. It is interesting that one retrospective study found proximal limb weakness in 6 of 24 EGPA patients with peripheral neuropathy had proximal limb weakness, which might have been concomitant skeletal muscle involvement that was overlooked clinically.[4] The wider use of noninvasive imaging may help to detect more cases of EGPA myositis either with or without peripheral neuropathy.

Author Contributions

Data curation: Young-Eun Park. Validation: Doosoo Jeon. Writing—original draft: Kyoungnam Woo. Writing—review & editing: Jin-Hong Shin.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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