Leg Gangrene in Granulomatosi s With Polyangiitis

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Granulomatosi s with polyangiitis (GPA) is an antineutrophilic cytoplasmic antibody (ANCA)-associated vasculitis characterized by necrotizing granulomatous inflammation of the small vessels. Primarily, the ears, upper respiratory tract, lungs, and kidneys are affected; gangrene occurs in less than 1% of cases.1,2

A 53-year-old man with no prior medical history visited the emergency room with severe left leg pain, redness in the eye, and hearing loss over the past month. He underwent endovascular treatment for suspected acute arterial occlusion, but no thrombus could be aspirated, resulting in gangrene in his toes (Figure 1). Multiple pulmonary nodules were also detected; septic pulmonary embolism was suspected at that time. Angiographic findings and antimicrobial drug-resistant inflammation led to a suspicion of vasculitis, and he was referred to our hospital. Blood
tests revealed positive proteinase 3 (PR3)-ANCA, and the presence of hearing loss, pulmonary nodules, and inflammation of the nasal/paranasal sinuses (Figure 2) collectively contributed to the GPA diagnosis.3

Methylprednisolone pulse therapy and rituximab (375 mg/m²/week) immediately resolved the symptoms and lung lesions, but not the gangrene, leading to below-knee amputation 3 months after the induction of remission. Pathological examination of the amputated specimen revealed intimal thickening, medial dissection, and vascular occlusion in the anterior and posterior tibial arteries, but no atherosclerotic changes.

Leg gangrene typically manifests in patients with conditions such as diabetes mellitus, atrial fibrillation, and peripheral vascular disease.4 Although not a typical sign of small-vessel vasculitis, GPA should be considered if other clinical findings suggest it, as prompt treatment may help avoid amputation.

REFERENCES

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Figure 2. (A) Noncontrast CT image shows soft-tissue opacification of the sinuses, in the mastoid air cells and middle ear. (B) Noncontrast CT image shows multiple pulmonary nodules in a random distribution. CT: computed tomography.