ANCA-associated vasculitis and actinomycosis: a case report

Dear Editors,

Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a group of autoimmune diseases characterised by necrotising small-vessel vasculitis, including microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA) and eosinophilic granulomatosis with polyangiitis (EGPA) (1). Infection is both the leading cause of mortality in AAV and a potential trigger or exacerbating factor. This article presents a rare case of AAV complicated by infection with *Actinomyces bouchesdurhonensis*, a previously undocumented strain. In addition to the well-known issues of infection-mimicking vasculitis and infection-triggered vasculitis, this case provides diagnostic and therapeutic experience for the coexistence of chronic occult actinomycosis and primary AAV. It also offers new insights into the differential diagnosis and staged treatment of patients with suspected AAV and chronic lung lesions.

A 65-year-old male patient was admitted to our hospital with a 3-month history of bilateral ankle swelling and pain, with intermittent nonproductive cough and recurrent fever (peak temperature: 39.5°C). Anamnestic records showed a history of otitis media in childhood and left lacrimal duct obstruction 10 years prior, without recent hearing loss, ear discharge, or otalgia. Nasal examination revealed no deformity, discharge, or paranasal sinus tenderness.

Laboratory findings showed: white blood cell count $6.02x0^9$ /L, haemoglobin 88 g/L, platelets $248x10^9$ /L, and erythrocyte sedimentation rate (ESR) 90 mm/h. Renal and hepatic function tests were within normal ranges. C-reactive protein (CRP) was elevated at 116.65 mg/L. MPO-ANCA and p-ANCA were positive. Electromyography (EMG) demonstrated peripheral neuropathy in both lower limbs, predominantly involving sensory fibres. Maxillofacial CT suggested left maxillary sinusitis and a right maxillary sinus cyst. Ankle ultrasound showed joint effusion, posterior tibial tendon tenosynovitis, and retrocalcaneal bursitis in both ankles.

Chest CT showed multiple nodules, linear opacities, consolidations, pleural thickening, as well as reactive mediastinal and hilar adenopathy (Fig. 1). A review of chest CT scans over the past five years indicated that the pulmonary lesion had been progressing very slowly. Bronchoscopy was performed, and next-generation sequencing (NGS) of bronchoalveolar lavage fluid detected *Actinomyces bouchesdurhonensis*, but conventional bacterial culture did not grow *Actinomyces* or other pathogens. Given that NGS had already identified the pathogen and the patient's respiratory symptoms were stable, the incremental benefit of invasive biopsy was considered limited relative to its potential risks. Histopathological examination was not performed, and sulphur granules could not be observed. Although colonisation or contamination could not be completely excluded, the slowly progressive pulmonary lesions over five years and the CT morphological features strongly supported that *Actinomyces bouchesdurhonensis* was pathogenic infection rather than simple colonisation.

The diagnosis of microscopic polyangiitis (MPA) was established according to the 2022 ACR/EULAR criteria, with a total

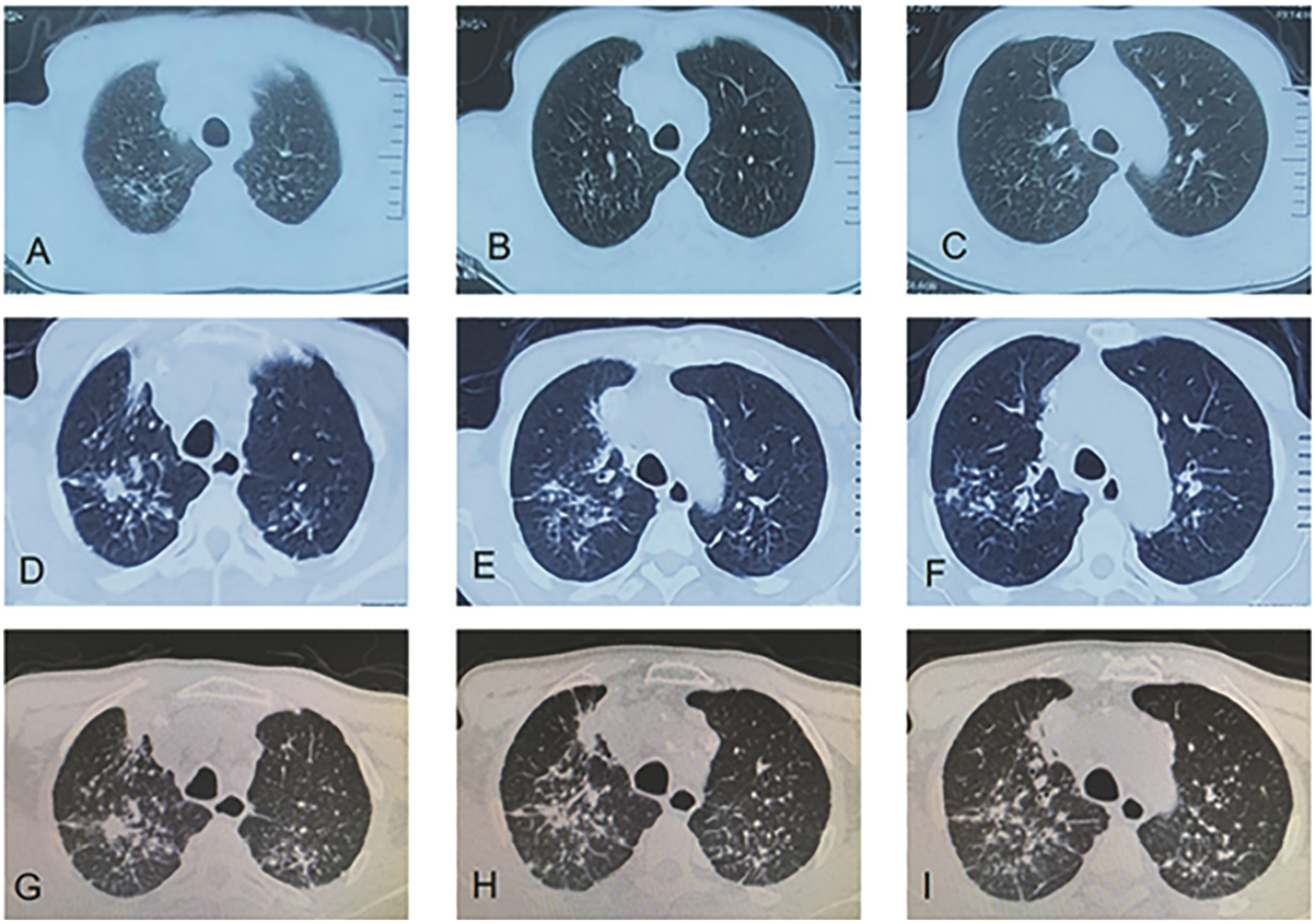


Fig. 1. Chest CT images of a male patient at different times. Chest CT in November 2018 (**A, B, C**) revealed multiple small nodules. Chest CT in August 2023 (**D, E, F**) and October 2023; (**G, H, I**) showed localised consolidation with partial patchy fusion, findings more consistent with actinomycosis. The absence of diffuse ground-glass opacities further argues against AAV.

score of 6 points based on positive MPO-ANCA (or p-ANCA) and peripheral neuropathy without other identifiable cause (2). The patient's ankle symptoms initially improved with non-steroidal anti-inflammatory drugs (NSAIDs) and antibiotics, and reactive arthritis secondary to actinomycosis was considered. However, symptoms recurred one month later, accompanied by persistently elevated inflammatory markers, supporting arthritis as a manifestation of MPA. Considering that the patient had received standardised anti-infective therapy for one month and the respiratory symptoms were initially stable, immunosuppressive therapy was initiated. At the same time, a six-month tapering antibiotic course for presumed chronic actinomycosis was continued. The patient's symptoms resolved and inflammatory markers normalised within one month. Although follow-up imaging was unavailable for an objective comparison due to relocation, clinical remission has been maintained for two years with prednisone (5 mg/day) and leflunomide (20 mg/day).

The reports of AAV combined with infection are not uncommon, with pathogens including *Klebsiella*, *Staphylococcus*, among others (3). However, the coexistence of AAV with actinomycosis is extremely rare, and the case caused by *A. bouchesdurhonensis* is first reported. Actinomycosis is a rare chronic granulomatous disease caused by the Gram-positive anaerobic *Actinomyces* (4). It can affect multiple organ systems and present with diverse clinical manifestations that mimic malignancies or other inflammatory diseases, posing diagnostic and treatment challenges (5).

A. israelii is the predominant pathogenic species in the genus *Actinomyces*. Other common human pathogens include *A. naeslundii, A. viscosus*, *A. odontolyticus*, *A. meyeri*, and *A. gerencseriae* (6). *A. bouchesdurhonensis* was first isolated in March 2016 from a gastric lavage sample of a 60-year-old man with iron-deficiency anaemia (7). To our knowledge, this is the first documented case demonstrating the potential pathogenicity of *A. bouchesdurhonensis* in humans.

On imaging, the patient's lung lesions progressed slowly for over five years, which is clearly inconsistent with typical AAV lung involvement. AAV lung lesions are mostly acute or subacute, with migratory infiltrates, ground-glass opacities, alveolar haemorrhage, as well as vascular and bronchial changes including signs of vasculitis and bronchial wall thickening (8). Actinomycosis mostly manifests as chronic peripheral nodules, consolidation, and adjacent pleural thickening, with a prolonged and progressive course (9). The patient had core vasculitis manifestations (ANCA positivity and peripheral neuropathy) along with imaging and microbiological evidence of chronic lung infection. Thus, the final diagnosis was coexistence of two diseases, rather than one mimicking or secondary to the other.

This case highlights that for patients with suspected AAV who also exhibit chronic and indolent lung lesions, it is necessary to actively investigate the possibility of chronic infections (such as actinomycosis). Integrating disease course, imaging, and microbiological findings is essential. A sequential approach, with antibiotics followed by immunosuppressants after infection control, proved safe and effective and minimized the risk of infection flare. As a newly discovered species, *A. bouchesdurhonensis* warrants clinical attention regarding its pathogenicity. Nevertheless, a limitation of this case is the lack of post-treatment chest imaging follow-up, which precluded objective radiographic assessment of lesion resolution, although clinical remission has persisted for two years.

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Competing interests: none declared.

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