In this report, we describe a 61-year-old man that presented with isolated pulmonary vasculitis and a positive anti-cyclic citrullinated peptide (CCP) antibody. Within a few months, the patient developed the symmetric polyarthritis consistent with rheumatoid arthritis (RA). Because the anti-CCP antibody is highly specific for RA and vasculitis is a known association of RA, we suspect the pulmonary vasculitis in this patient was the first manifestation of underlying RA. This case extends on previous reports that have shown that lung disease may predate the development of articular RA and that anti-CCP positivity and lung disease may represent a pre-RA phenotype. To our knowledge, this is the first case report of pulmonary vasculitis as the first manifestation of RA.

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improving (Fig. 3). Follow-up autoantibody testing three and six months after initiation of methotrexate was notable for persistent high-positive anti-CCP and low-titer positive RF, and negativity to anti-proteinase-3, anti-myeloperoxidase and C- and P-anti-neutrophil cytoplasmic antibody (ANCA).

### 3. Discussion

Vasculitis is a well recognized extra-articular manifestation of RA; however, it is usually considered to be associated with long-standing, severe, erosive, nodular, and sero-positive disease. RA-vasculitis often manifests in the skin with pyoderma gangrenosum, nervous system with mononeuritis multiplex, or may involve other organs such as the lungs. Because the current era offers more effective disease modifying anti-rheumatic drug therapies, RA-vasculitis is less commonly encountered.

We believe this case represents pulmonary vasculitis as the presenting manifestation of RA. A few factors argue in favor of such a diagnosis: the presence of the high-titer anti-CCP; an autoantibody known to be highly specific for RA. The growing body of evidence that anti-CCP antibody positivity and lung disease may predate the articular manifestations of RA, and that such patients may reflect a pre-RA phenotype. Our patient developed the symmetric inflammatory polyarthritis and synovitis consistent with RA. Finally, pulmonary vasculitis is a well known manifestation of RA. Although the order of presentation — lungs before joints — is atypical, the overall clinical scenario favors anti-CCP positive RA with pulmonary vasculitis as the best unifying diagnosis.

We can not fully discount the possibility that this case represents an atypical presentation of granulomatosis with polyangiitis (GPA) (formerly Wegener’s). Indeed, inflammatory polyangiitis is a common finding in patients with GPA. There are several aspects of this case that argue against a diagnosis of GPA: the patient did not have sinusitis or renal involvement; two features commonly encountered in GPA. The patient was ANCA, proteinase-3, and
myeloperoxidase antibody negative. The patient had a high-titer RA specific autoantibody (anti-CCP).

In conclusion, this case reinforces the concept that a wide spectrum of lung disease may be the presenting manifestation of RA – including pulmonary vasculitis.

Conflict of interest statement

None of the authors have any financial interests to disclose.

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