Pulmonary involvement in systemic lupus erythematosus (SLE) can take the form of pleuritis, interstitial lung disease, alveolar hemorrhage, or pulmonary hypertension, but rarely does it appear as pulmonary vein vasculitis1.

A 19-year-old woman was diagnosed with SLE 6 months before presentation because of malar rash, alopecia, arthritis, leukocytopenia, low complement, and positive anti-DNA antibody, and 30 mg daily prednisolone (PSL) treatment was started. Fever, skin ulcers on the scalp, and dry cough appeared 2 months before presentation, while she was taking 15 mg of PSL. Thoracic computed tomography (CT) scan revealed bilateral multiple nodules distributed along pulmonary veins (Figure 1A).

The patient had jaundice and discoid lesions with ulcers on the scalp on physical examination. Laboratory findings showed acute liver dysfunction, decreased complement titer, positive anti-DNA antibody, negative antiphospholipid antibody, and no kidney involvement. Lung biopsy revealed lymphocytes and foam cell infiltration and fibrosis around pulmonary veins (Figure 2C–E) with disrupted internal elastic lamina (Figure 2F). Methylprednisolone pulse therapy followed by 50 mg of daily PSL was started, and intravenous cyclophosphamide treatment was added. The discoid lesions resolved with scarring, and liver dysfunction improved soon after the initiation of therapy. One month later, thoracic CT scan showed that the pulmonary nodules were drastically reduced in size (Figure 1B).

Our original view was that the patient’s pulmonary vein vasculitis was a manifestation of SLE because the SLE-specific discoid lesions were exacerbated at the same time. Although we sometimes see SLE with vasculitis, this case included the extremely unusual presentation of vasculitis limited to pulmonary veins2. This case highlights the possibility of vasculitis when nodules along pulmonary veins are found.

ACKNOWLEDGMENT
We thank Dr. H. Sugiu for his advice on radiological findings. We also thank Dr. A. Sasaki and Dr. K. Kameyama for useful comments on the pathology.

REFERENCES
Figure 2. Histopathology of the lung tissue. Panels C–E show lung biopsy that revealed lymphocytes and foam cell infiltration and fibrosis around pulmonary veins, but without pulmonary artery lesions or thrombosis (H&E staining, magnification C: ×40, D: ×100, E: ×200). F. Disrupted internal elastic lamina were also seen (arrows, Elastica van Gieson staining, magnification ×100).