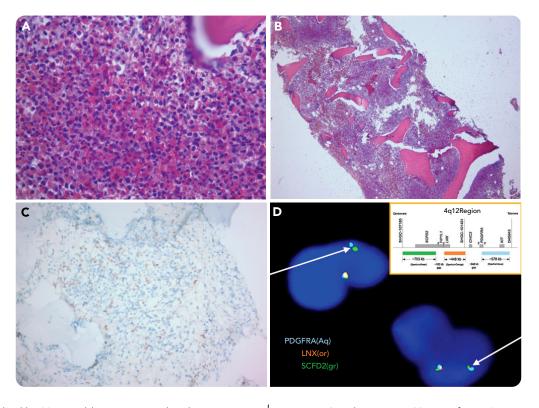


Myocarditis, kidney injury, and hypereosinophilia: *FIP1L1-PDGFRA*-positive myeloid/lymphoid neoplasm with eosinophilia

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A previously healthy 28-year-old man presented to the emergency department with chest pain and petechiae on his extremities. Labs revealed elevated troponin level (6.86 ng/mL), acute kidney injury (Cr, 1.95 mg/dL), and eosinophilia (WBC, 22 400/μL, 57% eosinophils). Transthoracic and transesophageal echocardiograms were negative for valvular pathology/endocarditis, but a cardiac magnetic resonance image was consistent with myocarditis. Fluorescence in situ hybridization (FISH) from peripheral blood revealed *PDGFRA* gene rearrangement with *FIP1L1-PDGFRA* fusion in 75% of interphase cells (panel D; tricolor rearrangement FISH probe, peripheral blood). The serum tryptase level was elevated to 30.9 ng/mL. A bone marrow biopsy demonstrated hypercellular marrow (panel B; hematoxylin and eosin [H&E]-stained bone marrow, 2× objective, ×20 magnification) with granulocytic hyperplasia, increased eosinophilic precursors with maturation (panel A; H&E-stained bone

marrow, $40\times$ objective, $\times400$ magnification), increased mast cells (panel C; tryptase-stained bone marrow, $20\times$ objective, $\times200$ magnification), increased reticulin network (MF-1 to MF-2 fibrosis), and PDGFRA gene rearrangement. He was diagnosed with hypereosinophilic syndrome secondary to FIP1L1-PDGFRA-positive myeloid/lymphoid neoplasm with eosinophilia and was started on steroids and imatinib treatment. After 2 weeks of therapy, he achieved a complete hematologic response with an undetectable eosinophil count. His troponin eventually normalized, and his creatinine improved to 1.75 mg/dL.

The FIP1L1-PDGFRA fusion protein is a tyrosine kinase that results from a deletion of CHIC2 on chromosome 4q12 and causes transformation of hematopoietic stem cells. Patients with this mutation typically respond well to tyrosine kinase inhibitors, such as imatinib.



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