High-Resolution Color-Coded Sonography in Angiolymphoid Hyperplasia With Eosinophilia Presenting as Temporal Arteritis

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A 37-year-old white man presented with extensive and mildly painful swelling of both superficial temporal arteries (STAs) (Figure, upper left). The condition was progressive for 4 weeks, and the patient had mild night sweats. He noticed a circadian rhythm with less prominent STA swelling in the morning. No other deficits, including claudicatio masticatoria, visual symptoms, or (any) neurological symptoms, were reported. High-resolution color-coded sonography (Siemens Sequoia, 15L transducer, Mountain View, Calif) revealed highly enlarged STA bilaterally, with a diameter of up to 9 mm and sinusoidal intraluminal vessels with arteriovenous blood flow velocities (Figure, middle, and Movie in the online-only Data Supplement) and some areas with a hypoechoic rim ("halo" sign). Furthermore, swelling of the occipital arteries and enlarged cervical lymph nodes were found. Clinical chemistry revealed eosinophilia of 17% with an almost unremarkable blood sedimentation rate (20 mm/2 hour), whereas other lab parameters, including leucocyte count, c-reactive protein, and total immunoglobulin E, were normal. Exposure of the left STA showed the grossly enlarged and convoluted vessel with the surrounding connective tissue appearing more prominent than usual and altogether thickened (upper right). The histological examination in cross-sections showed STA with nearly complete occlusion of the vascular lumen due to irregular proliferation of small capillary-size vessels (Figure, lower left, hematoxylin & eosin staining, magnification ×25). Most of the vessels were lined by distinct epithelioid endothelial cells admixed/interspersed with chronic inflammatory cells, predominantly eosinophils. Staining for the endothelial cell marker CD31 revealed multiple small vessels correlating to the sonographic appearance of the arteritis (Figure, lower right. CD31 staining, magnification ×100). No giant cells were found. At the periphery, there was a prominent/dense lymphocytic infiltrate arranged like a rim around the STA. Magnetic resonance imaging ruled out any intracerebrovascular involvement but showed enhancement of the STA. We recommended corticosteroid therapy; however, the patient improved spontaneously over the next 5 months without any therapy.

Temporal arteritis in the young is a rare entity. Differential diagnosis includes angiolymphoid dysplasia with eosinophilia, Kimura disease, and juvenile temporal arteritis. The common features of these entities are nodules in the head and neck adjacent or within the vasculature. Epidemiological differences between the 3 exist but partially overlap or are inconsistent in the literature. Angiolymphoid dysplasia with eosinophilia often occurs in young women in the third and fourth decade with multiple lesions, no regional lymphadenopathy, mild eosinophilia and immunoglobulin E elevation, arteriovenous malformations from exuberant angiomatoid proliferation, and masses of uncanalized epitheloid or histiocytoid endothelial cells. However, only a halo sign had been previously described in high-resolution sonography. Kimura disease is more often seen in young men with involvement of vessels in the head and neck, frequent elevated serum immunoglobulin E and blood eosinophilia, and extensive capillary proliferation with large thick-walled vessels. Finally, juvenile temporal arteritis occurs in boys and young men regardless of ethnic background, and the prominent histological features occurring in all vessels are vascular proliferation, eosinophilic infiltration, and, to a lesser degree, fibrinoid necrosis. All 3 diseases appear to be benign although pathogenesis remains widely unknown. Therapeutic strategies include local photodynamic therapy, local and systemic corticosteroids, and 3-cis-retinoic acid, but recurrences have been reported in all 3. No systemic vascular inflammation has been reported so far.

Our case illustrates the value of high-resolution sonography revealing extensive arteriovenous flow consistent with CD31-positive intraluminal vessel proliferation. The case presented here might present an overlap syndrome with features of all 3 entities: angiolymphoid dysplasia with eosinophilia, Kimura disease, and juvenile temporal arteritis.
In conclusion, high-resolution sonography may be an additional noninvasive tool to further characterize and separate angiolymphoid dysplasia with eosinophilia, juvenile temporal arteritis, Kimura disease, and overlapping syndromes.

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Disclosures
None.

References
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