An 86-year-old man presented with double vision and bitemporal ulcerative secreting skin lesions that had developed over the preceding 3 weeks (Figure A). His medical history included a 3-month episode of new headache and jaw claudication, and a common cardiovascular risk profile (cerebral ischemic stroke 10 years previously, hypertension, atrial fibrillation, and vascular surgery involving the aorta and right carotid artery), as well. The patient denied experiencing fever or weight loss; his drug regimen included antihypertensive, anticoagulant (phenprocoumon), and topical corticosteroid agents. Clinical examination revealed mild anisocoria and palphypesthesia, resulting in a slight impairment in gait, and a prominent, indurated, but painless left superficial temporal artery (STA), as well.

High-resolution color duplex ultrasonography of the STA revealed an atypical echogenic endothelial layer and a periluminal hypoechogetic halo sign (Figure B and C). Extracranial ultrasonography identified moderate generalized arteriosclerosis. The erythrocyte sedimentation rate (ESR) was mildly elevated: 28 mm after 1 hour and 58 mm after 2 hours.

The halo sign, masticatory claudication, and patient’s age prompted the diagnosis of temporal arteritis (TA), whereas the echogenic intimal layer of the STA was interpreted as a consequence of the prolonged or subacute course of the disease. A biopsy of the left STA was performed with surprisingly little loss of blood and yielded histological findings of lymphomonocytic inflammatory infiltration within the vessel wall, intimal fibrosis, and polymuclear giant cells (Figure D and E). The pattern of skin erosions perfectly fit the route of the STA. We treated the patient with oral glucocorticoid medication (initial dose of 100 mg prednisolone tapering to 7.5 mg over 6 weeks), which resulted in rapid normalization of the ESR, and elimination of the patient’s headaches and improvements in his skin lesions and double vision, as well. At the 5-month follow-up, the patient was free from symptoms.

TA, also known as Morbus Horton or giant cell arteritis, is acute vasculitis of medium to large arteries; it is found in elderly patients in whom there is granulomatous inflammation of the vessel wall. Typical symptoms consist of headache, jaw claudication, weight loss, and fever; of greater concern are symptoms of vision loss and diseases of cardiac or cerebral blood vessels. The diagnostic gold standard for TA is represented by the American College of Radiology criteria of age ≥50 years, localized headache with (sub)acute onset, ESR elevated to >50 mm/h (Westergren), palpable STA, and histological proof of vasculitis. Recently, high-resolution color duplex ultrasonography has been proposed as an alternative to temporal artery biopsy, because the appearance of the halo sign, a hypoechogetic signal indicating a thickened vessel wall, has a high predictive value for vessel wall infiltration.

This case illustrates an unusual presentation of TA with the following characteristics: (1) necrotic skin lesions; (2) double vision instead of loss of vision; (3) prolonged course of the disease; (4) concurrent echogenic intimal and hypoechogetic perivascular appearance on high-resolution color duplex ultrasonography; and (5) only mild elevation of the ESR.

Fewer than 100 cases of TA with bitemporal affliction of the scalp have been reported to date. This specific disorder consists of vasculitic occlusion of supplying small arteries and insufficient collateral routes, leading to deficient circulation and necrosis. The mean time from initial symptoms to beginning scalp necrosis is 2.9 months, whereas the mean time to diagnosis is 4.0 months. Interestingly, elevated ESR—a primary criterion for TA—is not always found in cases of TA with scalp necrosis. The fact that the ESR in this case was only mildly elevated may indicate the prolonged course of the disease before diagnosis. This may be supported by the high reflectivity of the STA vessel wall often observed with chronic inflammation. Another aspect of the disorder is that preexisting antiplatelet or anticoagulatory therapies, such as in this case, seem to prevent or attenuate major ischemic complications such as amaurosis or stroke. Double vision attributable to eye muscle paresis is a rare symptom of TA and may be observed in vasculitic ischemia of the eye muscles.

Unusual symptoms of the neck, head, and brain possibly related to ischemia with only mild ESR elevation in elderly patients should prompt investigation for TA including high-resolution color ultrasonography of the STA.

Disclosures

None.

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References


Figure. A, Necrotic skin lesion on the right temporal scalp at treatment initiation. B and C, High-resolution color duplex sonograms of the STA showing hypoechoic periluminal halo signs (∗) and echogenic intimal layers (arrows). D and E, Photomicrographs showing a vascular lumen that is narrowed owing to the proliferation of intima cells and intima fibrosis (D) and intima proliferation and severe transmural granulomatous inflammation (E). STA indicates superficial temporal artery.
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