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A “Negative” Temporal Artery Biopsy, Positive for Arteritis

Ophthalmologists often participate in the diagnosis and treatment of patients with giant cell arteritis (GCA), typically when the diagnosis is heralded by a central retinal artery occlusion or ischemic optic neuropathy. However, even in the absence of eye symptoms or signs, ophthalmologists may be asked to examine the patient and perform a biopsy of the temporal artery. The microscopic findings in the patient described herein bear on the technique of temporal artery biopsy.

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Report of a Case. A previously healthy 81-year-old woman developed mandibular pain with chewing, low-grade fevers, night sweats, and proximal limb myalgias. She denied headache or visual changes. There were no abnormalities on results of physical examination, and her temporal arteries were pula-ntile and neither indurated nor tender. A chest radiograph was unremarkable. The only abnormality on routine laboratory testing was microscop ic hematuria. Her condition did not improve after a trial of antibiotics for the presumed diagnosis of sinusitis. Three weeks after onset of symptoms, an erythrocyte sedimentation rate of 131 mm/h and a C-reactive protein level of 7.9 mg/dL prompted referral for neuroophthalmic consultation and biopsy. Although no abnormalities were found on the examination results, we performed a biopsy of the left temporal artery.

The biopsy specimen was fixed and serially sectioned. Microscopic examination (Figure) showed a normal temporal artery in all sections, but several small adjacent arteries included in the biopsy specimen showed granulomatous angiitis.

The patient’s symptoms improved with oral prednisone therapy, but further investigations prompted by persistent hematuria resulted in the diagnosis of Wegener granulomatosis.

Comment. As demonstrated by this case, symptoms, signs, and even histologic findings classically associated with GCA can be found with other systemic vasculitides. Features attributed to GCA, such as jaw claudication, amaurosis fugax, and an erythrocyte sedimentation rate greater than 100 mm/h, have been seen in patients with Wegener granulomatosis.1-3 Like patients with GCA, patients with Wegener granulomatosis can improve with corticosteroid treatment, at least initially. However, patients with Wegener granulomatosis who are misdiagnosed as having GCA and who are treated only with corticosteroids may develop acute renal failure and respiratory tract disease.4 When faced with a “positive” tem-

poral artery biopsy, the physician must always consider a diagnosis of a systemic vasculitis other than GCA, as the patient may experience a fulminant and downhill course if appropriate therapy is not instituted.

From personal experience with a patient whose diagnosis of GCA was established from angiitis in small vessels adjacent to a normal temporal artery, one of us (S.L.) routinely includes the periarterial tissue in temporal artery biopsy specimens. Had this tissue been stripped from the specimen in our patient, the biopsy would have been interpreted as negative. Temporal artery biopsy specimens from other patients with Wegener granulomatosis have also shown only small-vessel involvement.1-3

When a temporal artery biopsy is performed, the surgeon should refrain from removing the periar-terial tissues except to the extent necessary to ensure accurate identification and secure ligation of the vessel. Nor should the pathologist strip the artery before embedding.

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The Role of Midface Lift and Lateral Canthal Repositioning in the Management of Euryblepharon

Euryblepharon is a congenital eyelid anomaly characterized by horizontal enlargement of the palpebral fissure. The eyelid is shortened vertically compared with the horizontal dimension, with associated lateral canthal malpositioning and lateral ectropion.1,2 It may be an isolated finding or associated with ocular anomalies such as lateral displacement of the proximal lacrimal drainage system, a double row of meibomian gland orifices,2 telecanthus, and strabismus.3 In severe cases, it may result in lagophthalmos and exposure keratopathy2 and may require surgical treatment. We report the results of 2 patients with hereditary disorders and euryblepharon treated successfully with midface lift and lateral canthal repositioning surgery. The surgical technique is described.

Report of Cases. Case 1. A 17-year-old girl with Noonan syndrome (Online Mendelian Inheritance in Man 163930) and bilateral lower eyelid euryblepharon since birth was seen with eye irritation and nocturnal lagophthalmos. The parents were also concerned about the aesthetic appearance of their child. An examination revealed upper eyelid ptosis, bilateral inferolateral displacement of the lateral canthus, lateral ectropion (Figure 1), and lagophthalmos with mild punctuate keratitis.

She was treated with bilateral midface lift via a swinging eyelid