that, even with identical genetic deletions at the Xp22.3 locus, the phenotypes may vary. Patients 1 and 2 were similar in that both of them had microphthalmia, sclerocornea, and skin deformities. They also exhibited microphthalmia and a mild cardiac abnormality in the form of SVT. There were, however, several differences in the phenotypes of the twins. Patient 1 had a definite high elevation of IOP, while patient 2 had a borderline elevation of pressure. Patient 2 had a small right eyelid fissure and anophthalmia, neither of which was present in patient 1. Microcornea was present in patient 1, but the corneal diameter of the nonanophthalmic left eye of patient 2 was normal. These phenotypic differences found in identical twin boys with identical karyotypes lend further support to the current hypothesis that the variations found in MIDAS syndrome may be due to different patterns of X inactivation, or lyonization, rather than due to subtle differences in genotypes. Further research should be done to understand more completely how the Xp22.3 deletion is expressed because this research may lead to a better understanding of the role of this chromosomal locus in ocular development.

The clinical definition of MIDAS syndrome continues to be modified as more phenotypic variability is reported. The present study represents one of the largest case studies detailing the ocular findings in patients with an Xp22.3 deletion and serves to expand the current phenotypes associated with this syndrome. Providing additional descriptions of the phenotype should aid clinicians in identifying patients with this genetic syndrome and prompt early genetic testing in appropriate patients. It is apparent that the mnemonics “MIDAS syndrome” and “MLS syndrome” do not fully describe this disease. Therefore, consideration should be given to more accurately and genomically referring to the syndrome as the “Xp22.3 microdeletion syndrome.”

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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.

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 pulsa-tile and neither indurated nor tender. A chest radiograph was unremarkable. The only abnormality on routine laboratory testing was microscopic 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 periarterial 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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