Giant-cell temporal arteritis in a 17-year-old male

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Temporal arteritis, particularly in its classic form, is exceedingly rare in individuals <50 years old. We report the youngest case of biopsy-proven giant cell temporal arteritis. A 17-year-old male presented with a progressively expanding and pulsatile but otherwise asymptomatic mass in his forehead. The patient's medical history was significant for uveitis since the age of 3, and severe allergic rhinitis, mild asthma, and juvenile rheumatoid arthritis as a young adolescent. Admission laboratory values included a mildly elevated erythrocyte sedimentation rate and C-reactive protein level. A computed tomography evaluation demonstrated aneurysmal degeneration of the frontal branch of the right superficial temporal artery and confirmed no other cerebrovascular changes. Histologically, the aneurysmal arterial segment demonstrated subacute temporal arteritis. The arterial wall had a primarily lymphoplasmacytic infiltrate with rare giant cells and focally marked medial destruction. Additionally, severely obstructive intimal hyperplasia with chronic adventitial and periadvential dense fibrosis was noted. The diagnosis of classic giant cell temporal arteritis was established from the biopsy result. Postoperatively, the patient was treated with prednisone for 3 months. Three years after surgery, the patient remains well and reports no recurrence of temporal artery disease. (J Vasc Surg 2006;43:1053-5.)

Temporal arteritis is a vasculitis that is extremely rare in patients <50 years old. In the young, temporal arteritis exists in two forms: juvenile temporal arteritis and classic giant cell arteritis. Juvenile temporal arteritis was first reported in the English literature in 1975.[1-6] It has a benign prognosis, and to date, only nine cases have been reported in the English literature. The classic form is even more uncommon: only four cases have been reported,[7-10] one of which is disputed.[11] We report the youngest case of biopsy-proven giant cell temporal arteritis.

CASE REPORT

A 17-year-old male was referred to the vascular surgery service at the University of Nebraska for evaluation of a localized pulsatile mass in his forehead It was initially noted by the patient 3 months earlier as a mild, painless swelling, but it had acutely hardened and expanded over the previous 2 days. The patient denied fever, malaise, headache, visual changes, or jaw claudication. On examination, the easily noticeable mass was pulsatile, nontender, oblong, and located on the frontal belly of the right epicranius muscle. It started midway between the right eyebrow and the hairline and extended in serpentine fashion (Fig 1).

The patient's medical history was significant. Idiopathic uveitis had developed at age 3, with a positive antinuclear antibody (ANA) (1:320). A recent ophthalmology evaluation demonstrated bilateral aphakia, aphakic glaucoma, and postoperative changes from a previous corneal transplant with chronic inflammation and calcium deposits. Severe allergic rhinitis and mild asthma developed when he was young adolescent. At age 15, juvenile rheumatoid arthritis was diagnosed in the right ankle. His medications at the time of presentation included fexofenadine, nasal steroids, sulfasalazine, naproxen, and allergy immunotherapy.

Admission laboratory values included an erythrocyte sedimentation rate of 11 mm/h, a C-reactive protein concentration of 3.0 mg/dL, an ANA titer of 1:640, and negative double-stranded DNA profile. Peripheral blood demonstrated no anemia, a white cell count of 10,600/μL, with 2% eosinophils. Serum electrolytes were within normal limits.

The patient had normal vital signs, and the rest of his vascular exam was within normal limits. An ophthalmologic evaluation showed no new changes and no active ocular vasculitis.

A computed tomography evaluation demonstrated aneurysmal degeneration of the frontal branch of the right superficial temporal artery and confirmed no pathologic changes in the aortic arch, supra-aortic trunks, or extracranial and intracranial cerebral vasculature.

The preliminary diagnosis was aneurysmal dilatation of the frontal branch of the right superficial temporal artery. The patient was taken to the operating room and under local anesthesia with intravenous sedation underwent exploration of the diseased artery and resection of its aneurysmal segment.

Grossly, the arterial segment removed was 1.2 cm long, with asymmetric aneurysmal dilatation resulting in a diameter that ranged from 0.4 to 0.7 cm. On cross section, the aneurysmal portion had marked luminal narrowing due to a thickened tan-gray intima. Microscopically, the resected artery demonstrated subacute (mildly active and healing) temporal arteritis. The infiltrate in the artery was composed primarily of lymphocytes and plasma cells and extended transmurally from intima to adventitia (Figs 2 and 3) with large areas of internal elastic lamina destruction. Rare, multinucleated giant cells were present. Eosinophils were not present.

Marked intimal hyperplasia was present and included large numbers of smooth muscle cells, as confirmed by immunophenotypic marker for smooth muscle actin, and caused severe luminal stenosis. Luminal thrombus focally contributed to the obstruction. Medial destruction was focally prominent. Dense adventitial fibrosis focally extended into adjacent soft tissue and skeletal muscle and

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Competition of interest: none.

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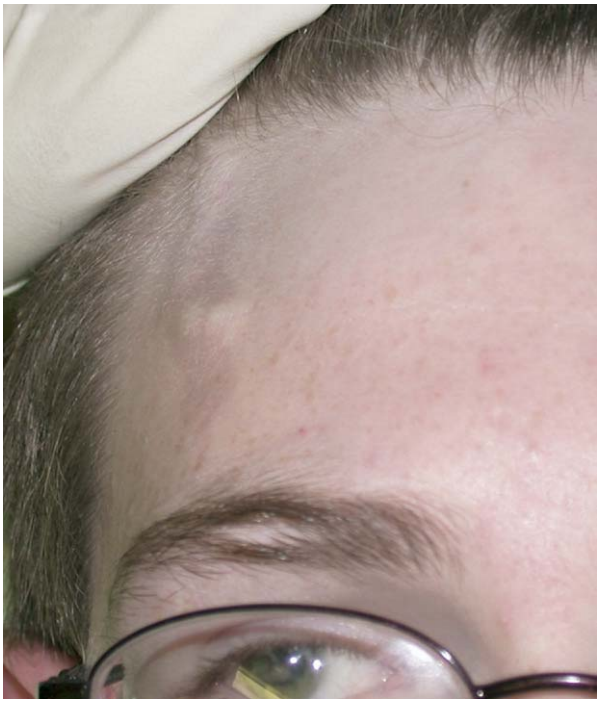

Fig 1. Right temporal artery of patient.

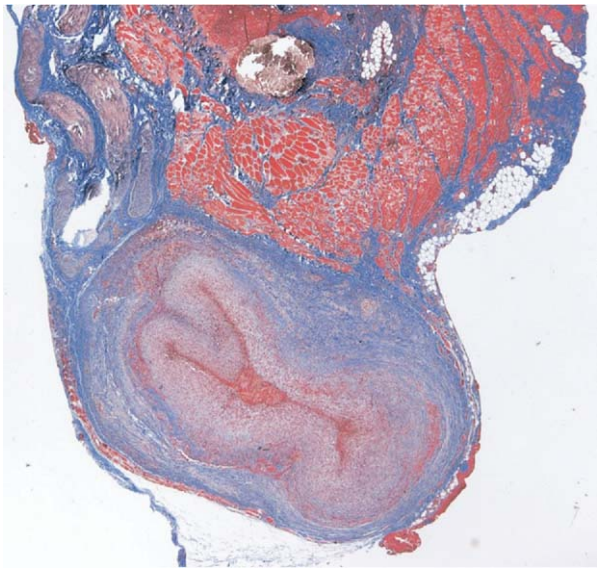

Fig 2. Photomicrograph of temporal artery with lymphoplasmacytic infiltrate associated with internal elastic lamina and medial destruction with marked intimal hyperplasia with resultant luminal obstruction (Masson trichrome ×20).

encased nearby nerves. The fibrosis appeared to be older than the active arteritis and presumably represented healing of a previous inflammatory process.

Postoperatively, the patient was treated with prednisone (0.5mg/kg daily) in addition to his baseline medications. The wound healed well, and he was weaned to an alternate-day dosage of prednisone without any complications. Prednisone was tapered over 3 months. Three years after surgery, he has remained well and has reported no recurrence of temporal artery disease.

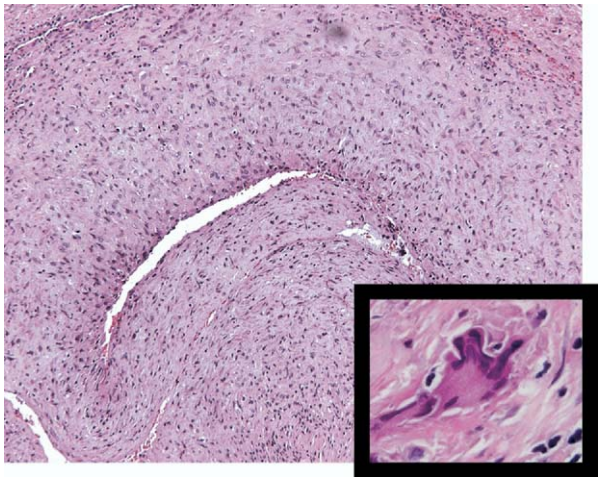

Fig 3. Higher power (Masson trichrome ×100) photomicrograph demonstrates intimal hyperplasia composed of many smooth muscle cells and scattered lymphocytes with rare giant cells (*insert* ×400).

DISCUSSION

Temporal arteritis is a disease that very rarely affects persons <50 years of age. In the largest series reported, all patients were ≥50 years, and 95% of them were >60 years.[12] Age ≥50 years at disease onset is the first of five criteria formulated by the American College of Rheumatology for the diagnosis of temporal arteritis. The other four criteria are (1) new onset of localized headache, (2) temporal artery tenderness or decreased temporal artery pulse, (3) elevated erythrocyte sedimentation rate (Westergren) ≥50 mm/h, and (4) a biopsy sample that shows necrotizing arteritis and is characterized by a predominance of mononuclear cells or granulomas with multinucleated giant cells.

In the last three decades, 13 patients <40 years old (range, 7 to 35 years) have been reported with vasculitis of their temporal arteries. This small group of patients can be divided into two subgroups by the microscopic features of their biopsy tissues.

The first form of the disease is characterized by panarteritis, occasionally with a prominent eosinophilic infiltrate[4,5] and no granulomas or giant cells. This type of arteritis was first described in detail by Lie et al[1] in 1975 and was termed *juvenile temporal arteritis.*[2-3] Its original description may have been as early as 1948, however.[13] The disease has a predilection for young males and presents as a tender pulsatile mass in the temporal region without any other accompanying signs or symptoms. On laboratory evaluation, the erythrocyte sedimentation rate is normal, and the peripheral blood often shows hypereosinophilia. A

recent report of a 31-year-old with juvenile temporal arteritis found an associated activated protein C resistance.[6] The disease has a benign course and requires no corticosteroid treatment.[14]

The classic microscopic features of the adult form of giant cell (temporal) arteritis have only been rarely reported in younger subjects.[6-9] In these few cases, there was a necrotizing arteritis characterized by a preponderance of mononuclear cells or granulomas with multinucleated giant cells. To our knowledge, only five cases, including the present one, have been reported.[6-9] The patients are usually male and generally describe symptoms lasting several weeks or months that include localized headache, temporal artery tenderness, and malaise. Laboratory findings are not consistent. The results of hematologic studies are entirely normal in some patients, whereas others have leukocytosis and an elevated erythrocyte sedimentation rate. Treatment with prednisone has been associated with resolution of symptoms and absence of recurrences.

The current patient had no systemic complaints, although his history was complicated by long-standing ANA-positive uveitis, a recent first-time presentation of juvenile arthritis, allergic rhinitis, and asthma. Nevertheless, none of these pre-existing clinical conditions were associated with temporal arteritis in our literature review. His significant allergic rhinitis and mild asthma would be expected to be associated with peripheral blood eosinophilia, although his peripheral eosinophil count was not elevated. A microscopic review of temporal artery biopsy tissue provided a definitive diagnosis and resulted in treatment with corticosteroids.

Among young adults or adolescents, the literature provides no distinct differences in the clinical presentation between temporal arteritis with giant cells and juvenile temporal arteritis. Our case represents the only reported patient with a positive ANA and another rheumatologic condition (uveitis and arthritis). The evaluation of a young patient with a temporal mass should include a complete blood count and a sedimentation rate. A total serum immunoglobulin E might be of some assistance in patients with peripheral eosinophilia. Ultimately, the diagnosis is usually established by microscopic evaluation of temporal artery biopsy tissue. The critical features the pathologist should describe are the presence or absence of giant cells and eosinophils. The diagnosis of classic giant cell arteritis should be considered in all patients (even in those <50 years) presenting with temporal artery related symptoms or signs.

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