When a temporal artery biopsy reveals a diagnosis other than temporal arteritis: eosinophilic granulomatosis with polyangiitis

Case Report

A 60-year-old male patient was seen in a community hospital for an episode of amaurosis fugax in the left eye in August 2010. His erythrocyte sedimentation rate was 100 mm/hour with mild leukocytosis of 12.9 $\times$ 10^9/L, a platelet count of 307 $\times$ 10^9/L, and a positive C-reactive protein as indicated in the lab report. A tentative diagnosis of giant cell arteritis (GCA) was suspected, and he was started on oral prednisone at 50 mg/day. Subsequently, the patient was referred to our institution for a temporal artery biopsy (TAB) to confirm the diagnosis. On further questioning, the patient had a history of a nonproductive cough and fever for several weeks; he also had lethargy, malaise, anorexia, and weight loss of 6 lbs over the previous 2 months. During the same period, he developed shooting pain and paresthesia of his left foot. His medical history was significant for asthma, recurrent sinusitis, and nasal polypectomy. A chest x-ray done at the community hospital showed a vague consolidation in the medial segment of the right middle lobe in the lung.

Histopathological examination of the left superficial temporal artery revealed a normal temporal artery, but focal necrotizing small vessel vasculitis with eosinophils were present in the soft tissue adjacent to the main artery (Figs. 1 and 2). This was not consistent with GCA but rather suggested the differential diagnosis of small vessel vasculitis such as granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis (EGPA, formerly known as Churg–Strauss syndrome), or microscopic polyangiitis. Serologic investigations revealed MPO-ANCA positivity (900 units; normal $<$99 units). An electromyogram revealed a mononeuritis monoplex in the left leg, and referral to the rheumatology service confirmed a diagnosis of EGPA based on the patient meeting 5 out of the 6 diagnostic criteria as established by the American College of Rheumatology (ACR). Cyclophosphamide was added, and he has responded well to therapy to date.

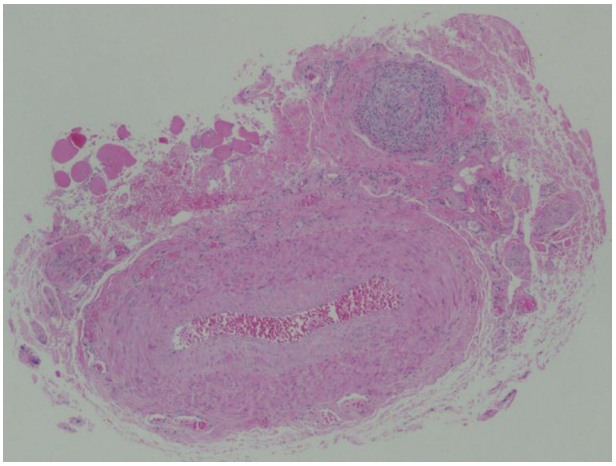

Fig 1—Temporal artery biopsy including large main artery and smaller branch vessel superiorly. Note normal appearance of main artery. (Hematoxylin and eosin 25X).

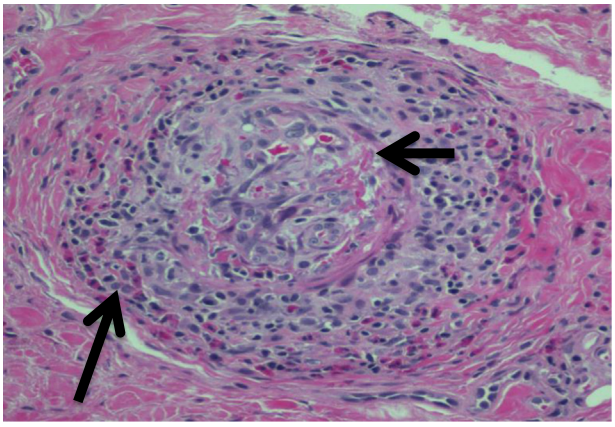

Fig 2—Small arterial branch vessel showing necrotizing vasculitis with Fibrinoid necrosis (short arrow) and infiltration with eosinophils and lymphocytes (long arrow). (Hematoxylin and eosin, 200X).

The 2012 Revised International Chapel Hill Consensus Conference on the nomenclature of vasculitides has defined EGPA as "eosinophil-rich and granulomatous inflammation often involving the respiratory tract and necrotizing vasculitis affecting primarily small- to medium-sized vessels, and is associated with asthma and eosinophilia. ANCA is more frequent when glomerulonephritis is present."[1,9] It is a rare disorder that was first described by the pathologists Churg and Strauss in 13 patients in 1951.

There are different classification criteria for diagnosing EGPA and the most commonly used is the 1990 ACR criteria. Our patient met 5 out of 6 criteria defined by the ACR for the diagnosis of EGPA: asthma, neuropathy (as mononeuritis multiplex involving the left foot), nonfixed pulmonary infiltrates (as right infrahilar infiltrates), paranasal sinus abnormality (as recurrent sinusitis), and extravascular eosinophilia (as was seen on TAB).[2] Ocular involvement in EGPA can vary from purely ocular to neuro-ophthalmologic manifestations. Our patient presented with neuro-ophthalmic symptoms (1 episode of amaurosis fugax); other neuro-ophthalmologic manifestations include cranial nerve palsies and ischemic optic neuropathy.[3,4,5] Ophthalmologic involvement includes keratitis, conjunctival nodules, dacryoadenitis, myositis, uveitis, and retinal vascular occlusion.[5]

Temporal artery involvement in vasculitides other than GCA is rare but is reported in polyarteritis nodosa, EGPA, GPA, microscopic polyangiitis, hepatitis B virus–related polyarteritis nodosa, hepatitis C virus–related cryoglobulinemic vasculitis, and rheumatoid vasculitis as either case reports or small case series.[3–8,10,12–14] The diagnosis of EGPA in our patient was suggested based on TAB findings.

A descriptive study was done by Genereau et al. on patients from the French Vasculitis Study to assess the frequency of temporal artery involvement in different types of systemic necrotizing vasculitis (SNV).[7] From their cohort of 141 consecutive patients undergoing TAB for suspected GCA, they found 6 patients with SNV; they then accumulated 21 other patients with SNV identified on TAB retrospectively from collaborating institutions for a total of 27 patients diagnosed with SNV by support of TAB. Only 2 were known to have SVN before the TAB. They found that cephalic symptoms such as jaw claudication, clinically abnormal temporal arteries, and neuro-ophthalmologic symptoms were present in 81% of patients. In 70% (19 patients), the diagnosis of SNV was based on findings of the TAB. In the majority of these cases, temporal artery localization of the SNV was the first sign of vasculitis. Esteban et al. reviewed 28 patients in whom small vessel vasculitis in the soft tissue surrounding a spared temporal artery was the first histologic finding that led to the diagnosis of vasculitis.[10] They identified 3 patients with SNV but did not further subclassify them. Finally, Hamidou et al. retrospectively identified 7 patients with SNV who underwent TAB for cephalic symptoms.[8] They concluded that TAB is a simple tool to diagnose systemic vasculitis, but the histopathological findings need to be correlated with the clinical findings as it does not always discriminate between SNV and classic GCA.

This case highlights the importance of including other SNV in the differential diagnosis of GCA. Non–giant cell temporal arteritis is rare but is a documented finding in SNV; therefore, TAB should be considered as a site for biopsy when cephalic symptoms are present in the context of systemic vasculitis. The key to the correct diagnosis of SNV in TAB is a good arterial specimen with surrounding soft tissue to include small vessels as well as proper communication with the pathologist.[11]

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Innovative model for telemedicine-based screening for diabetic retinopathy in the developing world

The magnitude of diabetic retinopathy (DR) in the developing world and the need for periodic screening are universally accepted.[1] In India the numbers to be screened —across geographies, poor infrastructure, and resource constraints—make it a daunting proposition. Successful public health programs such as the National Diabetic Retinopathy Screening Service in the UK need expensive hardware, excellent network connectivity, and dedicated manpower.[2] We have run a DR screening and awareness program in rural India for the last 2 years, conducting 41 camps. Our model has 3 components as recommended