

Temporal Arteritis: An Atypical Presentation

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Abstract: Temporal (giant cell) arteritis is a systemic granulomatous vasculitis primarily involving branches of the carotid arteries in patients aged 50 years and greater. Its classic symptoms and signs are headache and elevated erythrocyte sedimentation rate (ESR), but this is not the only presentation. This case is the first reported in a Korean, whose chief complaint was pain in the tongue and headaches and whose ESR was normal. The diagnosis was confirmed by biopsy. Her symptoms abated when treated with prednisone, but she developed diabetes mellitus, osteoporosis, and compression fracture of lumbar vertebrae while being treated. Primary physicians should become aware of the atypical features of this disease, as well as the potential complications of treatment. (J Am Board Fam Pract 1991; 4:115-8.)

Temporal (giant cell) arteritis is a systemic granulomatous vasculitis, which primarily affects branches of the carotid artery in patients aged > 50 years.¹ The clinical manifestations, epidemiologic features, and laboratory findings of this disease are well documented.¹⁻³ The patient described here, however, had atypical complaints, epidemiologic features, and laboratory results.

Case Report

A 70-year-old Korean woman came to the family medicine center complaining of severe tongue pain, which was gradual in onset and present for several weeks. During this time, she also described diffuse frontal headaches, mild to moderate in severity, and a general sense of malaise. She denied jaw or tongue claudication, visual symptoms, or weight loss. Her examination was remarkable for bilaterally tender temporal arteries, which were pulsatile but not enlarged. All other physical findings were normal including the tongue and oral mucosa. Her laboratory tests included a Westergren erythrocyte sedimentation rate (ESR) of 38 mm/hr (normal) and a hematocrit of 34 percent (normal red cell indices).

Despite the relatively low ESR, the clinical diagnosis was that the patient had temporal arteritis, and she was treated with prednisone (60

mg/d) and scheduled for temporal artery biopsy. Two days after starting prednisone, all of her symptoms resolved completely; however, she developed new symptoms of polyuria and polydipsia, and her random serum glucose was 25.03 mmol/L (451 mg/dL). She then was admitted to the hospital for treatment of her diabetes mellitus and for a temporal artery biopsy.

The biopsy specimen showed diffuse symmetric intimal hyperplasia with focal loss of the internal elastic lamina associated with intimal thickening and asymmetric atrophy—findings consistent with temporal arteritis. She continued taking prednisone (a slowly tapering dosage) and felt well for 4 months when she again came to the family medical center complaining of severe low back pain without a clear precipitating event, such as a fall. Her physical examination demonstrated focal pain along the upper lumbar vertebrae.

A lumbosacral spine radiograph showed slight anterior wedging of the L₁ and L₂ vertebral bodies and reduced bone density. A bone scan documented intense radiotracer uptake in horizontal bands within the L₁ and L₂ vertebral bodies consistent with an acute compression fracture.

For treatment of osteoporosis, she was given conjugated estrogens (0.625 mg/d), calcium carbonate (1500 mg/d), vitamin D (50,000 units once weekly), and eventually fluoride (60 mg/d). Her back pain disappeared, and her diabetes mellitus was controlled. Twenty months after the onset of her initial symptoms of temporal arteritis, prednisone therapy was gradually stopped, and the patient has remained symptom free.

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Discussion

Clinical Symptoms

The clinical manifestations of temporal arteritis cluster into varying combinations of headache, other symptoms referable to the cranial blood supply, the polymyalgia syndrome, and constitutional symptoms.¹ The most frequent complaints (more than 50 percent of cases) include headache and temporal artery tenderness. Occasional symptoms (10 to 50 percent of cases) include visual problems, weight loss, fever, proximal myalgias, jaw claudication, facial pain, and scalp tenderness. Rare symptoms (less than 10 percent of cases) include blindness, extremity claudication, tongue claudication, ear pain, synovitis, stroke, and angina pectoris.¹⁻³

The headache of temporal arteritis is often nonspecific, resembling that of a tension headache. In some cases, it is more localized, with pain in one or both temporal arteries. It is typically described as superficial or burning, with paroxysmal lancinating pain.¹

Temporal artery symptoms may include swelling, nodularity, point tenderness, and diminished or absent pulse.^{3,4} Symptoms referable to the cranial blood supply include jaw, pharyngeal muscle, or tongue claudication. Pain, fatigue, or both are brought on by such activities as chewing, swallowing, or talking and are relieved by rest. Visual symptoms include amaurosis fugax, sudden blindness, ptosis, and ocular pain.^{1,5}

The polymyalgia syndrome is characterized by aching and morning stiffness in the proximal muscles, associated with an elevated ESR, and lasts more than 4 weeks and is responsive to low-dose corticosteroid therapy.

Nonspecific problems also may occur in combination with the already described symptoms or may be the isolated chief complaint. Fever, anemia, weight loss, and malaise are the most frequent. Healey and Wilske⁶ reported that 30 of 74 patients presented with one of these "occult" symptoms. My patient's chief complaint (severe tongue pain at rest) was not only unusual, but she lacked associated nonspecific complaints.

Laboratory Findings

The cornerstone in the diagnosis of a patient suspected of having temporal arteritis is an elevated ESR. The normal range for an ESR is age

and sex dependent: the upper limit of normal for men aged > 50 years is 20 mm/hr; for women, it is 30 mm/hr.⁷ Some authors suggest that the upper level of normal for patients more than 50 years should be 40 mm/hr.^{8,9} Most patients with temporal arteritis have an elevated ESR (median, 96 mm/hr; range, 50 to 132 mm/hr).^{6,10,11} In reports from Fauchald, et al.,¹² Huston, et al.,³ and Eshaghian and Goeken,¹³ the mean ESR exceeded 90 mm/hr in all three studies and exceeded 30 mm/hr in 136 of 138 patients.

Because an elevated ESR is so characteristic of temporal arteritis, the diagnosis is less likely to be pursued when the ESR is normal. Wong and Korn¹⁴ reported 36 cases of biopsy-proven temporal arteritis, 16 of whom had an ESR of 20 mm/hr or less and 20 had an ESR from 21 to 40 mm/hr. The clinical symptoms of patients with low ESR temporal arteritis were slightly different from patients with elevated ESRs. Headache and jaw claudication were significantly less frequent in patients with a low ESR.

Although my patient never had an elevated ESR, it is of interest that during treatment with prednisone, the relative changes in her ESR reflected changes in her symptomatology (Figure 1). Exacerbations of her disease never were reflected in an ESR greater than 38 mm/hr.

Despite the limitations of the ESR, it remains the single most useful laboratory test for the diagnosis of temporal arteritis.¹⁵ When the clinical

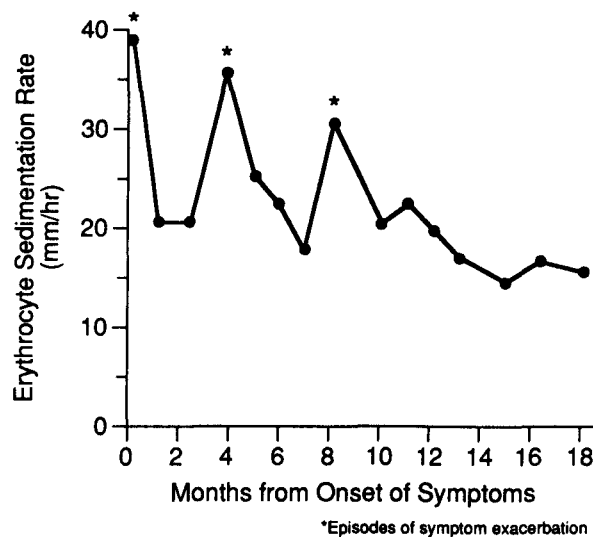


Figure 1. Erythrocyte sedimentation rate and exacerbation of temporal arteritis symptoms during the course of treatment.

evidence for temporal arteritis is weak, a normal ESR reduces the probability of the disease to less than 1 percent.¹⁵ Other laboratory findings that have been described are clearly less useful. A mild-to-moderate normochromic, normocytic anemia is present in more than 50 percent of cases.³ Other abnormalities include mild leukocytosis, thrombocytosis, increased alkaline phosphatase, glutamic oxalecetic transaminase levels and acute phase reactants.^{1,3} My patient had a normochromic, normocytic anemia (hematocrit = 34 percent), which after therapy for 6 months reverted to normal (hematocrit = 42 percent).

Epidemiology

Studies clearly indicate that the incidence of temporal arteritis increases with age. The rate is 6 to 30 times higher in the eighth decade than in the sixth decade. The disease is also more common in women, with female-to-male ratios ranging from 1.05 to 7.4.^{1,16,17} Until recently, temporal arteritis was diagnosed exclusively in white populations, but more recent reports include the diagnosis in blacks and hispanics.¹⁷ Biopsy-documented temporal arteritis in Asians has been rarely reported. In 1984 Wilske and Healey¹⁸ reported the first two cases in patients of Chinese extraction. The case presented here is the first report of a Korean with the disease.

Complications of Treatment

This report highlights the complications of steroid therapy in patients with temporal arteritis. This is primarily because most patients will require prolonged courses of treatment. The usual length of treatment varies from 6 months to 2 years,¹ and Huston, et al.³ showed that more than one-half require corticosteroid therapy for greater than 1 year. Six years of therapy was the mean reported by Fernandez-Herlihy.¹⁹ Complications of steroid therapy are well known and occur in approximately 50 percent of patients.³ Osteoporosis manifested by vertebral body compression fractures occurs in at least 50 percent of persons requiring long-term glucocorticoid therapy. Worsening of or new onset diabetes mellitus induced by glucocorticoid therapy is also well documented.²⁰ My patient had both complications, but both responded to specific therapy and became less of a problem as the steroid dosage was lowered.

Conclusion

The presenting features of temporal arteritis can be atypical with laboratory evidence and clinical features not supporting the diagnosis. In dealing with the diagnosis of this disease, it is important to consider the atypical features. Therapy, although curative, often is associated with adverse side effects.

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