# Temporal Arteritis: A Review and Case History

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Temporal cell arteritis is a systemic vasculitis occurring mainly in the elderly. Classic symptoms such as headache, jaw claudication, visual changes, and polymyalgia rheumatica make the diagnosis relatively easy. Occult or asymptomatic presentations, however, are often missed or attributed to another cause. It is important for clinicians to be aware of the diverse presentations of this disease to avoid unnecessary investigations and prevent complications such as visual loss, myocardial ischemia, cerebrovascular accident, and death. This report presents the case of a patient who was referred with anemia and an elevated erythrocyte sedimentation rate, and who developed a headache during the hospital admission.

**KEY WORDS.** Temporal arteritis; anemia; asymptomatic; polymyalgia rheumatica. *(J Fam Pract 1996; 43:294-300)*

Temporal arteritis, also known as giant cell arteritis, is a systemic inflammatory process of small- and medium-sized vessels first described in 1890 by Hutchinson.[1] Extracranial branches of the arch of the aorta and ophthalmic vessels are most commonly affected, but the process may involve any artery in the body.

It is generally a disease of people over 50 years of age. The mean age of onset is 71 years, and the incidence increases with age. Annual incidence rates have been reported to vary from 0.49 to 23.3 per 100,000 persons.[2] It has a striking predilection for whites, and the reported female-to-male ratio ranges from 2:1 to 5:1.[3]

The origin of temporal arteritis is unknown, but it is believed that both humoral and cellular immune mechanisms are involved. The patchy inflammatory changes are centered around the internal elastic lamina, which is fragmented. The infiltrate is granulomatous, with lymphocytes and monocytes being the predominant cell types. The multinucleated giant cell is characteristic but may not always be present. Lumen narrowing from intimal edema and vessel thrombosis at the sites of inflammation may occur.[4]

There is great diversity in the presentation of temporal arteritis. The Table lists the common initial manifestations and the signs and symptoms encountered. Headache, the most common initial manifestation, may be continuous or intermittent and has been described as sharp, aching, or throbbing.[5] It is usually localized to the temporal or occipital region. The artery may be nonpulsatile and feel cordlike on palpation. Scalp tenderness may also be present.

Polymyalgia rheumatica (PMR) is another common presentation of temporal arteritis. One study[6] reported that when PMR and temporal arteritis occurred concomitantly, PMR was the initial clinical presentation more than one half (53%) of the time. Temporal arteritis presented first in 21% of patients, whereas PMR and temporal arteritis began concurrently in 18%.

Bilateral jaw claudication attributable to arterial occlusion of the temporalis or masseter muscles is also a classic symptom.[5] Patients with temporal arteritis often present with occult symptomatology, however, or may even be asymptomatic and the condition discovered postmortem. A case of temporal arteritis presenting with anemia and other occult presentations is discussed, and the diagnosis, treatment, and prognosis are reviewed.

## Case History

A 72-year-old woman was referred for medical assessment after investigations for fatigue revealed a hemoglobin (Hgb) of 94 g/L and an erythrocyte sedimentation rate (ESR) of 100 mm/h. In addition to fatigue over the past few months, she

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reported a decreased appetite with a weight loss of 10 lb over the past year.

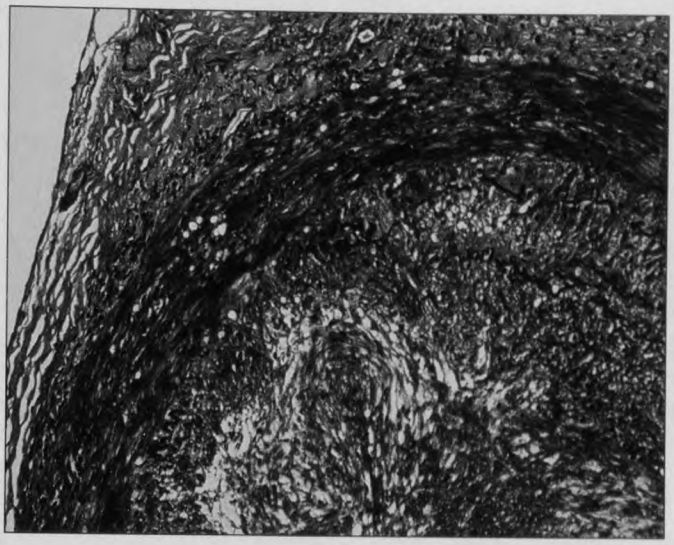
Her past medical history was significant for hypothyroidism, aortic valve replacement, and atrial fibrillation. Her medications included thyroid hormone, sotalol, warfarin sodium, digoxin, furosemide, potassium, and iron supplements. Findings from a physical examination were unremarkable except for a systolic ejection murmur and a rectal examination positive for occult blood.

Investigations showed a Hgb of 86 g/L with a mean corpuscular volume of 91 fL, leukocyte count of $8.6 \times 10^9$/L, and a platelet count of $339 \times 10^9$/L. Blood smear showed anisocytosis and ovalocytosis. The reticulocyte count was within normal limits. Electrolytes, creatinine, and urea values were also within normal limites, as were the urinalysis results. Iron studies revealed a serum iron of 6 μmol/L (normal 12 to 26), a ferritin level of 261 μg/L (normal 9 to 190), and total transferrin and total iron binding capacity (TIBC) at the lower limits of normal. Liver enzymes and function tests were within normal limits except for an alkaline phosphatase of 145 U/L (normal 40 to 125). Thyroid-stimulating hormone (TSH) was slightly elevated but thyroid hormone levels were normal. Esophogastroduodenoscopy showed small angiodysplastic lesions in the stomach, but these were considered an unlikely source of hemorrhage. Findings from a colonoscopy were entirely normal. Abdominal ultrasound revealed a 1-cm mass in the liver that was thought to be a hemangioma.

On the third day following admission, the patient developed a right-sided temporal headache that was mild and persistent, and radiated to the ear. It was alleviated with pressure and plain acetaminophen. She stated that headaches were very unusual for her. The temporal artery pulsation was easily palpable without nodules or associated tenderness. The headache gradually increased in intensity, and she required codeine for relief. An ophthalmologic examination did not reveal evidence of temporal arteritis. At that time it was decided to start treatment with 30 mg of prednisone (a lower-than-usual dosage was selected because of the patient's low weight), and vitamin D, calcium, and misoprostol. The headache remitted 2 days after the initiation of prednisone therapy. Temporal artery biopsy revealed classic temporal arteritis including giant cells and fragmentation of the elastic lamina (Figure).

**FIGURE**

**Temporal artery cross-sections from a 72-year-old woman presenting with anemia and elevated erythrocyte sedimentation rate. Verhoeff's stain for elastic tissue on high-power microscopy demonstrates the fragmented internal elastic lamina. Lumen narrowing with intimal edema is marked.**



The ESR started to decrease and was 23 mm/h on discharge, 8 days after the initiation of prednisone therapy. The ESR has subsequently dropped to 10 mm/h. The patient remains well on 2.5 mg of prednisone daily 1 year after presentation.

## Discussion

Temporal headache, polymyalgia rheumatica, and jaw claudication are classic symptoms that are easily recognizable. Up to 40% of patients, however, have atypical presentations.[7] This does not include the number of cases that remain undiagnosed, as found in autopsy studies.[8]

Although this patient eventually developed a headache, the initial presentation was that of anemia with fatigue and weight loss. She was otherwise asymptomatic. The diagnosis of temporal arteritis was seriously considered when the anemia and an elevated ESR were seen in light of a headache. Elevated alkaline phosphotase (ALP) is a common finding in temporal arterits. Iron studies

**TABLE**

**Initial Manifestations and Clinical Findings in 100 Patients with Temporal Arteritis**

| Symptom/Complaint | Number of Patients: Presenting Symptom | Number of Patients: Finding at Diagnosis |
|---|---|---|
| Headache | 32 | 68 |
| Polymyalgia rheumatica | 25 | 39 |
| Fever | 15 | 42 |
| Visual symptoms without loss | 7 | 30 |
| Weakness/malaise/fatigue | 5 | 40 |
| Tenderness over arteries | 5 | 27 |
| Myalgias | 4 | 30 |
| Weight loss/anorexia | 2 | 50 |
| Jaw claudication | 2 | 45 |
| Permanent loss of vision | 1 | 14 |
| Tongue claudication | 1 | 6 |
| Sore throat | 1 | 9 |
| Vasculitis on angiogram | 1 | — |
| Stiffness of hands and wrists | 1 | — |
| Decreased temporal artery pulsations | 0 | 46 |
| Erythematous, nodular, swollen arteries | 0 | 23 |
| Central nervous system abnormalities | 0 | 15 |
| Synovitis | 0 | 15 |
| Dysphagia | 0 | 8 |
| Limb claudication | 0 | 4 |
| Mean duration of symptoms before diagnosis, mo | — | 7* |

*Range 1 to 48 months.

Adapted from Hunder GG. Temporal arteritis and olymyalgia rheumatica. In: Kelly WN, Harris ED, Ruddy S, Sledge CB. *Textbook of Rheumatology.* 4th ed. Philadelphia, Pa; WB Saunders, 1993:1103-12.

were compatible with the usual pattern paralleling anemia of chronic disease.

It has been reported that 15% to 30% of patients with temporal arteritis have anemia.[9] Given that the cause of anemia is not found in 30% of anemic elderly patients,[7] a question arises of how many of these patients may have asymptomatic temporal arteritis.

Gallagher and colleagues[7] reported a case of an elderly patient with temporal arteritis who presented with anemia and an elevated ESR. They believed that it was the first report of asymptomatic temporal arteritis. They wondered if this patient would have developed classic symptoms, such as headache, had they not initiated steroid therapy, and whether all asymptomatic patients should be treated. This case demonstrates that asymptomatic temporal arteritis can progress to develop typical manifestations, presumably including the danger of visual complications. It is thus reasonable to conclude that biopsy-proven asymptomatic temporal arteritis should be treated in the same manner as a typical case of temporal arteritis.

There are other atypical or occult presentations of temporal arteritis. Fever is a common and often-missed manifestation of temporal arteritis. When confronted with a "fever of unknown origin" in an elderly patient, physicians should consider the diagnosis of temporal arteritis. Fevers can reach 39°C (102.2°F). Constitutional symptoms, such as weight loss, anorexia, and malaise, may accompany the fever or may be the sole initial manifestations. Weakness, fatigue, myalgia, and arthralgia also may be the only presenting symptoms.

Visual loss is a feared complication since it is usually irreversible. While it is not a common form of presentation, it eventually becomes apparent in 36% (reported range, 7% to 60%) of untreated cases.[10] Anterior ischemic optic neuropathy secondary to occlusion of the posterior ciliary arteries is the most common mechanism. Occlusion of the central retinal artery is less common. Fundoscopy may reveal disc pallor and edema (ischemia), cotton-wool spots, and retinal hemorrhages, followed by optic atrophy. Involvement is initially unilateral, with second eye involvement usually occurring within 1 to 10 days. Amaurosis fugax also can be seen in temporal arteritis. It is a transient symptom that should warn the physician of the risk of blindness and is an indication to measure the ESR.[4] Diplopia, another warning of possible visual complications,

is due to ischemia of the extraocular muscles. Upward gaze is most frequently affected, but ptosis and myosis may also be present.[10]

Although medium-sized branches of the carotid artery are most commonly affected, the aorta and its large branches may also be involved. Patients can present with intermittent claudication of an extremity, paresthesias, or Raynaud's phenomenon. Decreased or absent pulses and bruits may be found on physical examination. Coronary artery involvement may lead to myocardial ischemia, but it is difficult to determine the extent of contribution by atherosclerotic heart disease in patients of this age group.[11]

Neurological complications may occur in approximately 31% of the patients, as was found in a study of 166 biopsy-proven cases.[12] Neuropathies occurred in 14% of the patients. Mononeuropathies as well as peripheral polyneuropathies were observed and are presumed to be caused by ischemia of the nutrient arteries. Transient ischemic attacks or strokes (7%) were the next most frequent, followed by neuro-otologic findings in 7% (ie, vertigo, hearing loss, tinnitus). Tremors, psychiatric findings, and tongue numbness were also documented.[12]

Respiratory symptoms, such as sore throat, cough with or without sputum, and hoarseness, occur in approximately 10% of patients with temporal arteritis.[2] Patients may also present with tongue claudication or dysphagia due to ischemia of the muscles of deglutination. Temporal arteritis has also been reported in association with Churg-Strauss syndrome and periarteritis nodosa.[5]

In one study of 74 biopsy-proven cases of temporal arteritis,[13] 34 patients presented with "occult" symptoms. Fever of unknown origin (n=12) was most commonly encountered. Anorexia, weight loss, and elevated ALP suggesting malignancy (n=7) were the next most common. Three patients had anemia, and four had neurologic complaints (two with diplopia and two with acute weakness of one arm). In four patients, claudication involving the jaw (n=2), the leg (n=1), and the arm (n=1) was noted.

In patients who present with complaints or symptoms atypical of temporal arteritis, the clinician's index of suspicion is usually low. As a result, these patients are investigated extensively with invasive tests, and often the cause of their problem is either not found or attributed to another cause. A classification scheme has been proposed that divides temporal arteritis into classic and masked forms. The masked category was further subdivided into the following types: malignant (cachectic), anemic, febrile, aneurysmal (eg, aortic regurgitation and ruptured aortic aneurysm), and occlusive (eg, stroke, intermittent claudication, and myocardial ischemia).[14] This classification system may help increase clinicians' awareness of occult symptoms associated with temporal arteritis.

## Laboratory Findings

An ESR of approximately 100 mm/h is a characteristic laboratory finding. Using the Westergren method,[10] normal values in individuals over 50 years of age are less than 20 mm/h in men and up to 30 mm/h in women. There have been many cases of biopsy-proven temporal arteritis with normal ESR, however. One study concluded that most of these cases involved patients with a history of PMR or prior steroid therapy that lowered their ESR.[15] Temporal arteritis with an unexplained normal ESR is uncommon, but it does not rule out the diagnosis definitively.

Mild to moderate hypoproliferative normochromic normocytic anemia is a common finding in patients with temporal arteritis. It parallels the anemia of chronic inflammatory disease. Iron studies usually reveal a low serum iron, high serum ferritin, and reduced transferritin and total iron binding capacity (TIBC). The reticulocyte count remains lower than the expected level. Platelets are often increased but the leukocyte and differential counts remain normal.[16]

Nonspecific changes in acute-phase reactant proteins, including elevated fibrinogen, haptoglobin, and an elevated $\alpha_2$-globulin fraction, frequently occur. In patients with temporal arteritis, the fibrinogen level is typically elevated in proportion to the increase in ESR; this does not occur with the other types of vasculitis. Elevated factor VIII (von Willebrand's factor) has been reported and appears to be related to vessel damage. Also, the concentration of albumin is occasionally decreased. Serology for rheumatoid factor and antinuclear antibodies is characteristically negative.[2]

Abnormalities in liver function tests most commonly manifest as prolonged prothrombin time. An elevated ALP occurs in about 50% of patients, and

elevated aspartate aminotransferase is found occasionally.[10] Granulomatous changes in the liver have been found on biopsy.[2] Serum enzymes reflecting muscle damage, renal function tests, and urinalysis are usually normal.[10]

## Diagnosis

Temporal arteritis should be considered in the differential diagnosis in anyone over 50 years of age who presents with an elevated ESR, a new form of headache, scalp tenderness, jaw claudication, visual disturbance, polymyalgia rheumatica, anemia with anorexia, or unexplained prolonged fever. Careful questioning and physical examination is essential to strengthen the clinical suspicion.

Artery biopsy is the "gold standard" in making the diagnosis and is warranted in any patient in whom temporal arteritis is suspected. The morbidity of the biopsy is very low, and the predictive value with respect to the final diagnosis is approximately 90%.[5] Histologic evidence justifies exposing the patient to the many adverse effects of prolonged steroid treatment.

Temporal arteritis is a patchy disease and, hence, a biopsy may yield false-negative results. It is suggested that samples be taken from inflamed, tender, or palpable arteries. If there is no obvious sign of abnormality in any artery, a long segment (4 to 6 cm) of the temporal artery should be taken.[5] Serial transverse sections should be examined and, if negative, the artery on the opposite side should be biopsied. One study showed that 18% of the 234 positive biopsies were found only because of a biopsy on the second side.[17] A normal biopsy finding predicted that there was no need for corticosteroid therapy in 91% of the patients.

If the biopsies of both arteries are negative and the clinical suspicion is high, the physician must make a clinical judgment about corticosteroid treatment. Recently, Hunder and associates[18] developed criteria for the classification of temporal arteritis. According to their classification system, a patient can be classified as having temporal arteritis if three of the following five criteria are met: (1) age of onset ≥50 years; (2) localized headache of new onset or quality; (3) temporal artery abnormality such as tenderness or decreased pulsations unrelated to arteriosclerosis of cervical arteries; (4) ESR ≥50 mm/h by the Westergren method; and (5) abnormal artery biopsy with predominance of mononuclear cells or granulomatous inflammation, usually with multinucleated giant cells. The presence of three or more of these criteria yields a sensitivity of 93.5% and a specificity of 91.2%. When these criteria are met, long-term corticosteroid therapy can be initiated. A criterion proposed by another researcher suggested that prolonged corticosteroid can be started if the patient has an ESR ≥50 mm/h and four of the following symptoms: tender scalp, jaw claudication, recent visual symptoms, polymyalgia rheumatica, and good response to a trial of corticosteroids for 48 hours. Specificity with respect to histologic diagnosis was 100% but the sensitivity was only 24%.

Arteriography is recommended in patients with evidence of large artery involvement, such as limb claudication. Treatment of temporal arteritis is warranted if classic symptoms are absent but angiography reveals a smooth tapering stenosis characteristic of vasculitis and the patient is older than 50 years of age and has an elevated ESR.[11]

In the diagnosis of temporal arteritis, noninvasive techniques such as Doppler studies, gallium scans, and ocular pneumoplethysmography (which would show decreased ocular blood flow) are promising but require further studies.

While a differential diagnosis of temporal arteritis should include Takayasu's disease, polyarteritis nodosa, systemic lupus, and hypersensitivity vasculitis, it is often easy to make the distinction between temporal arteritis and other arteritides based on the patient's age, distribution of the lesions, organ involvement, and presence of polymyalgia rheumatica. Malignancies or other degenerative diseases should be considered in patients with systemic complaints who do not respond to a trial of corticosteroids.[10]

## Treatment

Temporal arteritis is a medical emergency. Once the diagnosis is established, treatment must be initiated immediately.

Glucocorticoids are the mainstay of therapy for patients with temporal arteritis. When there is a high level of clinical suspicion, especially in conjunction with visual symptoms, treatment can be initiated before a temporal artery biopsy. It has

been shown that a biopsy will remain positive after a week of corticosteroid therapy, and sometimes changes may persist for weeks or months after the institution of therapy.[19]

Recommended doses of prednisone vary from 30 mg to 100 mg, but most authors recommend 40 mg to 60 mg in divided daily doses as initially adequate in uncomplicated cases of temporal arteritis. With this regimen, a response is usually seen within 72 hours; however, the dose should be increased if the patient does not respond within a few days. There have been reports of visual symptoms worsening despite initiating treatment.[20] Based on a review of the literature, one report proposed that temporal arteritis with ocular involvement should initially be treated with high-dose intravenous corticosteroids (250 mg of methylprednisolone every 6 hours for 3 to 5 days) and then be switched to 80 to 100 mg per day of oral prednisone.[20] Therapy for osteoporosis should be initiated in these elderly patients because of the negative effect of prednisone on bone mass.

Steroid tapering should begin when all reversible symptoms have disappeared and laboratory values have returned to normal, which usually takes 2 to 4 weeks. Prednisone dosage should be tapered slowly over 6 to 12 months, with a written schedule of reduction given to the patient. Patients should be instructed to report any recurrence of signs and symptoms or any new pertinent symptoms. Although other acute-phase reactants have been shown to be helpful in following the course of the arteritis, ESR levels remain the most useful for this purpose. Tapering should be temporarily discontinued if symptoms recur or if the ESR increases. When the ESR returns to normal, tapering may be resumed at a more gradual pace. Every few months, an attempt can be made to taper the dose to zero.

Various modalities have been studied in an attempt to decrease the doses of corticosteroids required. Azathioprine and cyclophosphamide have been reported to be useful, but dapsone has shown the best results. One study reported that patients who were given prednisone with dapsone at the onset of disease had a lower total duration of therapy and a lower total dose of prednisone. Dapsone 75 to 100 mg daily has been recommended for this purpose.[21] More trials are needed with these corticosteroid-sparing agents.

### Course and Prognosis

Treatment can be expected to continue for 1 to 2 years and up to 5 years. Recurrences have been reported to occur as late as several years after the original diagnosis, but usually occur within 18 months following diagnosis or within 12 months of the discontinuation of therapy.[22] Although visual loss is considered irreversible, improvement in visual acuity has been reported, especially in patients who initially receive high-dose intravenous therapy.[20] It has been suggested the 15% of patients with profound visual loss will have some degree of improvement.[23]

If temporal arteritis is not properly treated, death may result from aortic rupture, stroke, or myocardial infarction. Survival of patients with temporal arteritis who receive treatment, however, seems to be the same as that of the age- and sex-matched population.

## SUMMARY

Problems associated with the diagnosis of temporal arteritis in patients with atypical presentations are significant. Physicians need to be aware of the occult manifestations of this disease and to have a high index of suspicion. Elderly patients with fever, anemia, or vague symptoms and an indolent illness should be investigated for temporal arteritis.

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