Chapter 55

The Elderly Woman with Sharp, Shooting Orbitotemporal Pains and Visual Loss

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Case History

A 69-year-old woman, recently widowed, reported vague neck and shoulder pain accompanied by global headaches that fluctuated from mild to moderate for the past 4 months. For 2 weeks, she experienced sharp, shooting pain, lasting from a few seconds up to 1 minute in the right orbit and temporal region. One morning, she awoke with loss of vision in her left eye. Four hours later, while awaiting her sister to pick her up to see the doctor, she lost vision in the right eye. Her family physician was seen immediately.

Questions on the Case

Please read the questions, try to answer them, and reflect on your answers before reading the author's discussion.

- Upon hearing this history, what differential diagnosis comes to mind?
- What physical signs should be carefully looked for?
- What laboratory tests would you draw or do?
- How would you formulate your evaluation and management plans?

Diagnosis

The examining physician reported mild scalp tenderness in both temples, but there were no nodules or thickening of the temporal arteries. The patient was normotensive and afebrile. Her physician did not record specific ophthalmologic changes other than that she was blind. Neck and shoulder muscles were mildly tender to touch.

Erythrocyte sedimentation rate (ESR) was 79 mm per hour. She was not anemic. Dexamethasone was administered at 10 mg intramuscularly.

When I first examined the patient 3 days later, she had some gradual improvement and could visualize hand movement only in the right eye, but there was no light perception in the left eye. The right optic disc was edematous and there was a marked right temporal visual field loss of the right eye. The left eye was blind and there was no abnormality by funduscopic examination. Pupils were 4 mm without light reaction on either side.

Right temporal artery biopsy proved normal. She was treated with continuous steroids, beginning with prednisone 20 mg every 6 hours. ESR fell to 32 mm per hour within 1 week. One month later, there was slight improvement of vision in the right eye, but not the left. She took prednisone 60 mg every other day. She reported gastric upset only on the days taking prednisone. Three months later, ESR was 44 mm per hour. She had gained 10 pounds. Blood pressure had risen from 125/78 to 180/95. She was treated with hydrochlorothiazide 50 mg plus potassium supplement. Blood pressure fell to 140–150/85–90. Over the next 6 months, there was mild improvement of right eye vision such that she could distinguish colors and read

some large signs, but not newsprint. No vision returned in the left eye. She was headache free. ESR remained stable at 44 mm per hour, and prednisone 40 mg every other day was continued.

Case Discussion

The theme of this book is that case-based study is valuable. This patient has temporal arteritis (TA). This case underscores several important principles regarding TA. First, TA must be near the top of our differential diagnosis in every elderly patient who presents with any kind of headache. The headache may be continuous, throbbing, burning, aching, shooting, stabbing, dull, mild, or severe—in short, there is no reliable typical headache characteristic. The scalp is often tender to touch, and the patient may complain of unilateral scalp tenderness or discomfort by simply lying on a pillow or by gently brushing the hair. Longstanding benign headaches are common, but we must remain alert to elderly patients who present with a new type of headache. The headache of TA is not always localized to the temporal region. TA may present as vague shoulder girdle myalgia and mild neck pain, which is all too often attributed to cervical arthritis. The neck pain may radiate to the face. Fatigue or pain with chewing—jaw claudication or claudication of the tongue—is a characteristic symptom and highly specific for TA (Table 55-1). The temporal arteries are described as thickened, engorged, beaded, and tender in most reports. However, I have 25 years of specific interest in TA and always examined the temporal arteries carefully; fewer than 50% of my patients had convincing temporal artery physical findings. The absence of physical signs must not dissuade the physician from the correct diagnosis.

The American College of Rheumatology has established widely accepted criteria for diagnosis of TA (Table 55-2).

The second important principle is that TA is a medical emergency. Correct diagnosis and urgent treatment may prevent irreversible blindness, as happened in this case. TA may cause typical amaurosis fugax, which heralds the blindness. Fifty percent of patients with TA who become blind in one eye will lose vision in the remaining eye soon after the first eye becomes symptomatic. This case demonstrated the classic presentation of visual symptoms: sudden painless unilateral visual loss upon awakening in the morning. As soon as the diagnosis is suspected, treatment with steroids should be initiated immediately. ESR is obtained at the same time, but treatment should not be delayed for the result of the ESR. A single dose of steroid is safer than waiting. Discussion of specific treatment issues will follow.

The third principle to consider is what to do if the ESR is low. Large series indicate that 86% of cases have ESR over 50 mm per hour. I have seen several cases of TA with ESR less than 23 mm per hour rising to 80 mm per hour within 24 hours. When the diagnosis is suspected despite a low ESR, the patient should be treated and the ESR repeated the next day. If the ESR remains low, it is especially important to perform a temporal artery biopsy to establish the diagnosis, because we are committing the patient to long-term steroids with potential for serious side effects. If the scalp tenderness is maximal in the occipital region, it may be prudent to biopsy the occipital artery rather than the temporal artery. The occipital artery is one of the largest branches of the external carotid artery, and there are several reported cases with a positive occipital artery biopsy after normal bilateral temporal artery biopsy.

The fourth principle is to recognize that TA is a general systemic condition. The term TA focuses on the frequent clinical involvement of the temporal artery. When this same condition is called giant cell arteritis (GCA), the emphasis is on the pathology of the disease, and the term cranial arteritis points to the most common anatomic region affected. If generalized muscle aching dominates the presentation, the term polymyalgia rheumatica has been applied to this clinical syndrome, which represents a continuum of the same process of GCA. Anorexia, weight loss, and sleep disturbance are common, and fever is pre-

Table 55-1. Specificity and Sensitivity of Signs and Symptoms in Patients with Suspected Giant Cell Arteritis

Sign or Symptoms	Sensitivity (95% CI)*	Positive Likelihood Ratio (95% CI)†
Jaw claudication	0.34 (0.29–0.41)	4.2 (2.8–6.2)
Diplopia	0.09 (0.07–0.13)	3.4 (1.3–8.6)
Prominent or enlarged temporal artery	0.47 (0.40–0.54)	4.3 (2.1–8.9)
Synovitis	–	0.4 (0.2–0.7)
Headaches	0.76 (0.72–0.79)	1.2 (1.1–1.4)
Erythrocyte sedimentation rate	0.96 (0.93–0.97)	1.1 (1.0–1.2)

Adapted from Weyand C and Goronzy J, 2003.

*Defined as the frequency in patients with biopsy-proven giant cell arteritis.

†Defined as the frequency of the sign or symptom in patients with giant cell arteritis compared with the frequency in patients who had a negative result on temporal artery biopsy.

Table 55-2. 1990 Criteria for the Classification of Giant Cell (Temporal) Arteritis (Traditional Format)*

Criterion	Definition
1. Age at disease onset ≥ 50 years	Development of symptoms or findings beginning at age 50 years or older
2. New headache	New onset or new type of localized pain in the head
3. Temporal artery abnormality	Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
4. Elevated erythrocyte sedimentation rate	Erythrocyte sedimentation rate ≥ 50 mm/h by the Westergren method
5. Abnormal artery biopsy	Biopsy specimen with artery showing vasculitis characterized by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

Adapted from Hunder GG et al, 1990.

* For purposes of classification, a patient shall be said to have giant cell (temporal) arteritis if at least three of these five criteria are present. The presence of any three or more criteria yields a sensitivity of 93.5% and a specificity of 91.2%.

sent in over 50% of cases. When these general systemic symptoms dominate the picture, diagnosis is frequently delayed. One series of 50 patients documented that successful early diagnosis was made in only 20% of patients who presented with systemic symptoms, whereas 75% of patients presenting with headache were correctly diagnosed at the first visit. Disseminated GCA may cause death from aortic dissection, myocardial infarction, intestinal gangrene, and stroke. Of patients diagnosed with TA who died, pathologic abnormalities with GCA involving the carotid and basilar arteries were documented in 70 to 100% of cases at autopsy. And yet, in large clinical case studies of TA, stroke is reported in only 3%. This discrepancy is surprising and tells us that the pathologic involvement of the carotid and vertebral arteries is high, but the clinical expression of the disease is low; there is little emphasis on this discrepancy in the literature.

Clinical problem solving becomes even more difficult when we consider the age group of patients with TA. These elderly patients are, of course, susceptible to stroke from atherosclerotic occlusive, hypertensive, and thromboembolic diseases unassociated with TA.

Management Strategies

One of the most controversial elements is that of treatment and long-term management of TA. The literature provides little data on specific management guidelines regarding dose and duration of necessary steroid treatment, or how often and how fast to reduce the dose of steroids.

Behn and colleagues examined this question 20 years ago and concluded that low doses that suppress the symptoms are sufficient; the dose can be increased if symptoms reoccur. Patients and family must be educated about TA, what symptoms to watch for, and the importance of immediately reporting to the physician any suspicious symptoms.

Some investigators have stated that moderate fluctuations of ESR unaccompanied by symptoms do not require increased dose of steroids. Others recommend tapering and stopping steroids when ESR falls to normal levels. As our case report demonstrates, the ESR may smolder along at 40 to 50 mm per hour while the patient's symptoms have subsided. In this situation, do we continue to treat with relatively high-dose steroids?

Published recommendations for reduction of steroids are really based on expert opinion without firm evidenced-based data. Several influential publications recommend an initial dose of 60 mg of prednisone. Time-to-peak plasma concentration of an oral dose of prednisone is 1.3 hours; therefore, the patient can begin treatment in the office faster than a trip to the hospital, where treatment may be delayed in the admission office, waiting to place an intravenous line, etc. Almost all documented cases of TA responded favorably within hours or days of treatment. Fixed visual defects seldom improved. Relapse of symptoms will occur in 50% of patients as the dose of steroids is reduced, and so all patients must return frequently for follow-up assessments. One study reported that of 87 patients who ultimately had a permanent remission and were able to discontinue treatment, 39 had one or more relapses as the steroid dose was reduced. Median time to permanent remission in these 87 patients was 22 months, but cases requiring treatment for up to 9 years are common.

The mortality rate of patients diagnosed with TA is double that of age- and sex-matched populations without TA. Some of these deaths are disease related, but death resulting from treatment-related complications is less appreciated. Two studies that focused on steroid-related complications found that 48 to 58% of patients with TA had severe side effects (Table 55-3). Of nine TA patients with severe infections, none experienced infections before starting steroids (pneumonia, lung abscesses, urosepsis, and candida sepsis). Six of these nine patients died. Hypertension, diabetes, psychotic reactions, gastrointestinal bleeding, and avascular necrosis were other serious steroid-related complications. Painful spinal compression fracture was the most common side effect; this may render the patient immobile and thereby susceptible to pulmonary infections. Although death resulting from the disease itself was recognized, of 43 TA patients, seven deaths were attributed to steroid treatment.

Table 55-3. Prevalence of Major Steroid (CS)-related Complications in 43 Patients with Temporal Arteritis

CS-related Complication	Patients (*N*)	Duration of Therapy until Complication (mean ± SD)
Fractures	15	12 ± 11 months
Infections	9	6 ± 5 months
DM, CHF, hypertension	8	3 ± 2 weeks
Depression/psychosis	3	4 ± 2 weeks
Bleeding peptic ulcers	2	4 ± 3 months
AVN of hip joint	2	19 ± 6 months

Adapted from Nesher G et al, 1994.

AVN = avascular necrosis; CHF = congestive heart failure; DM = diabetes mellitus.

Because side effects of prednisone are closely related to total dose, some investigators suggest that suppression of disease can be achieved with lower doses of prednisone, beginning with 40 mg daily dose, and tapering down according to the patient's symptoms and ESR responses. However, for patients who present with symptoms of impending visual loss, many experts begin treatment with higher doses such as 80 mg per day.

Some studies have demonstrated that steroid dose can be tapered more rapidly when combined with other immune suppressants such as methotrexate and azathioprine. These drugs afford steroid-sparing protective treatment. However, treatment cohorts were small and other studies have not confirmed this benefit. Methotrexate is not without potential harm, and some of these patients were withdrawn from the study when oral mucus membrane ulcers and bone marrow suppression developed.

Case Summary

- Temporal arteritis is one of the most common causes of secondary headaches in the elderly.
- TA is a medical emergency; our case report underscores the need for prompt diagnosis and urgent treatment.
- Corticosteroids are the specific treatment, but experienced judgment and close follow-up are required to capture the balance between treatment efficacy and serious and even life-threatening side effects.

Selected Readings

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Editorial Comments

Dr. Blumenthal has captured the clinical essence and seriousness of this disease in his clearly written chapter, and his summary should be echoed. TA is a medical emergency, and treatment with steroids should be initiated immediately. His solution of using intramuscular dexamethasone while awaiting laboratory and biopsy results is particularly clever and could prevent blindness. His clinical experience and review of the literature also yields a very important adage: "The pathological manifestations of giant cell arteritis are far in excess of the clinical signs and symptoms of patients." It is well worth remembering the scope of the pathologic changes, and the potential for severe morbidity and mortality of the disease—it is not an illness to be underestimated.

TA tends to affect older women, who often present with headache, visual loss, fever, and abnormal laboratory findings. Physicians should think about this diagnosis in a woman over 55 years who has malaise, fever, aches, and pains, even if there is no visual problem, temporal pain, scalp tenderness, and a history of jaw or tongue claudication. Because of the seriousness of the complications of the disease, treatment should be started in the office if the diagnosis is strongly suspected. Careful monitoring of the treatment may help to avoid the serious treatment emergent adverse events that may occur. Read this chapter carefully, as it teaches us a lot.

Final diagnosis:

Temporal arteritis