Temporal Arteritis With Normal Erythrocyte Sedimentation Rate

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Abstract: **The erythrocyte sedimentation rate (ESR) is used to differentiate temporal arteritis from other forms of headache in the elderly. Though temporal arteritis can occur with a normal ESR, this is not generally appreciated in primary care. The case reported here is a 74-year-old woman with biopsy-proven temporal arteritis; her ESR was 22 mm/hr. Of note, her hematocrit was more than 40 percent, a condition previously reported to be associated with temporal arteritis and a normal ESR. This discussion focuses on the diagnosis of temporal arteritis and its associated symptoms. (J Am Board Fam Pract 1991; 4:119-22.)**

A rapid erythrocyte sedimentation rate (ESR) often is used to help distinguish headaches caused by temporal arteritis from less important causes, even though several reports have shown that the ESR may be normal in temporal arteritis.[1-4] This fact appears widely unrecognized in primary care; standard primary care texts claim that the elevated ESR is a key, if not essential, element in the diagnosis of temporal arteritis.[5-10] In the case presented here, the diagnosis of temporal arteritis was made despite a low ESR.

Case Report

A 74-year-old white woman, on her first visit to the family practice center, had a 2-day history of occipital headache that had a mild bitemporal component. She also described a general "achiness" of the shoulders but denied transient vision loss, chewing claudication, or fever. She had noticed some fatigue and a 7-pound weight loss in the last 4 months but attributed this to her distress about her brother's recently diagnosed breast cancer. She expressed concern that her symptoms indicated a diagnosis of temporal arteritis.

On physical examination, her temperature was not recorded, the other vital signs were normal, and her weight was 104 pounds. Her visual acuity was normal. Both temporal arteries were palpable and moderately tender, and there was a strong pulse in each. There was no neck bruit. Her lungs were clear, and the liver was not palpable. Laboratory results showed her ESR (Westergren) was 22 mm/hr (age-adjusted normal ≤ 30); hematocrit 42 percent; and normal liver enzymes. The rheumatology consultant believed that the diagnosis of temporal arteritis was highly unlikely given the normal ESR and recommended against a biopsy. However, because of her clinical picture and the potential gravity of temporal arteritis, prednisone therapy was prescribed (60 mg/d). Within 18 hours, the patient noted relief from the headache.

Thirty hours after presenting, a biopsy was performed that showed chronic inflammation, granulomatous inflammation, and giant cells consistent with the diagnosis of temporal arteritis. Because she had read that a postoperative patient should not take steroids, she discontinued the prednisone after surgery, only to have a reoccurance of the headache after 2 days. When she called the office, prednisone was reinstated, and the headache again disappeared.

On the 6th day of prednisone therapy the patient returned to the office saying that she had experienced a dramatic increase in both her energy level and her interests and a 4-pound weight gain. The prednisone was decreased to 50 mg/d with continued clinical effect. On the 6th day her ESR was 8 mm/hr, and by 2 weeks it dropped to 2 mm/hr. Two months after treatment began the patient fell and injured her back. Radiographs showed an L_{4-5} narrowing. Two months later she again fell, and this time radiographs showed an anterior wedge compression fracture of T_{12}. Her ESR was 17 mm/hr while she was taking 25 mg of prednisone each day. An attempt was made to taper her medication, but the ESR rose to 28 mm/hr when prednisone was dropped to 20

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mg/d. The dose was returned to 25 mg per day with a subsequent drop of the ESR. Seven months after the initial presentation, her prednisone dosage is 12 mg/d, and her ESR is 17 mm/hr. Throughout the entire course, the hematocrit remained stable, and after the initial recurrence, the headache has never returned.

Discussion

Temporal arteritis (giant cell arteritis, cranial arteritis) is a systemic vasculitis that affects primarily large- and medium-sized vessels. While signs and symptoms referable to the head (headaches, visual symptoms, including blindness, and chewing claudication) are the most commonly cited vasculitis elements of the disease, an extracranial vasculitis can affect the liver, kidney, and peripheral nervous system.[11,12] Local manifestations are dependent on which arteries are involved. For example, there may be a transient diplopia because of paralysis of the extraocular muscles. Chewing claudication is considered pathognomonic and indicates involvement of the facial artery. Nevertheless, not all patients have pain.[13]

In addition, approximately half of all patients with temporal arteritis have polymyalgia rheumatica, though polymyalgia rheumatica occurs more commonly than temporal arteritis.[14] Both conditions are usually found in persons more than 50 years old and are associated with an elevated ESR. Both respond to corticosteroid therapy, but temporal arteritis requires higher doses in order to control the symptoms and to prevent blindness.[14]

Temporal arteritis has an annual incidence of approximately 10 per 100,000 in populations aged more than 50 years and is predominately a disease of whites.[12] The systemic ramifications include anemia, fever, weight loss, malaise, abnormalities of liver enzymes, particularly alkaline phosphatase, polymyalgia rheumatica, and an elevated ESR. In some patients, these systemic symptoms may predominate and lead to such presenting complaints as anemia, fever of unknown origin, or, as in this case, with a nonspecific illness with anorexia, malaise, and weight loss. High-dose and long-term prednisone therapy, with a slow taper, is the treatment of choice though the symptoms may progress during therapy.[15]

This case is important for several reasons. First, there is the clinical question about how one should monitor and treat patients with temporal arteritis and a normal ESR. The literature is not helpful in this regard. The approach that was successful here relied on closely monitoring the headache and the ESR. Without the assistance of this symptom and sign, management would have been more difficult.

Second, because the ESR was normal, what are the sensitivity and specificity of temporal artery biopsy? The findings of chronic inflammation, granulomatous inflammation, and giant cells in a temporal artery biopsy are essentially pathognomonic for temporal arteritis, i.e., the specificity is 100 percent. In practice, such a complete picture will not be found universally. Still, Vilaseca and colleagues, in their review of 45 cases of temporal arteritis in 103 patients undergoing biopsy, show a sensitivity of 82 percent and specificity of 100 percent.[16] In patients with a recent onset headache, jaw claudication, or abnormal temporal arteries, the sensitivity of the biopsy rises to 100 percent.

Third, the patient manifested behaviors uncommon for a 74-year-old woman. For example, she came to the office with the correct diagnosis, i.e. claiming that her occipital headache and moderately tender temporal arteries were temporal arteritis. She was therefore quite amenable to prednisone therapy and temporal artery biopsy. In addition, this educated patient had read that prednisone was relatively contraindicated in patients undergoing surgery. For this reason, she stopped the prednisone after her own temporal artery biopsy. This example of a little information being harmful was not picked up by either the primary care physician or the surgeon until the symptoms had reoccurred.

The final important elements of the case report involve the ESR. It is known that temporal arteritis can occur in patients with a low ESR. One recent review[17] of three articles[18-20] found an average ESR greater than 90 mm/hr, but 2 of 138 patients (1.4 percent) had an ESR less than 30 mm/hr. Such studies may underestimate the frequency of a low ESR in temporal arteritis because patients with a low ESR are less likely to undergo biopsy. This case report emphasizes the importance of considering temporal arteritis when the clinical picture dictates, despite the results of the ESR.

That this patient had a hematocrit of 42 percent may explain the normal ESR at the time of

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presentation. As mentioned, anemia is a common finding in temporal arteritis, and in 1987, Jacobson and Slamovitz reported that the degree of ESR elevation in patients with temporal arteritis was correlated with the amount of anemia.[2] In their study, all 6 patients with a hematocrit > 40 percent had an ESR < 50 mm/hr (10–49 mm/hr), while all 18 patients with a hematocrit < 40 percent had an ESR > 50 mm/hr (59–150 mm/hr). The ESR did not correlate with duration or severity of symptoms, nor was it prognostic for complications of the disease.

Another feature of the ESR in this patient is the low rate achieved after prednisone therapy. The differential diagnosis of a low ESR includes sickle cell disease, anisocytosis, spherocytosis, polycythemia, leukocytosis, congestive heart failure, and microcytosis.[16,20] None of these was present in this patient. In one study of patients with a very low ESR, 38 percent had no evidence of disease, and only 6 percent had one of the diseases commonly associated with a low ESR.

The question arises about the utility of following the ESR in patients with temporal arteritis, especially when the ESR is low. In polymyalgia rheumatica, a related disorder, the ESR closely correlates with symptoms because they both decrease on treatment. In temporal arteritis, however, this relation is not as strong, and a patient may improve symptomatically without an associated decrease in the ESR. In the present case, a normal ESR dropped to below the normal range on treatment. This also was reported in a patient with an initial ESR of 12 mm/hr, which dropped to 1 mm/hr after treatment.[3] That patient also had a hematocrit > 40 percent. The ESR response to any disease is multifactorial. It is unclear why one patient will improve clinically while maintaining an elevated ESR, while, in another patient, the ESR falls as symptoms decrease. For these reasons, the routine measurement of ESR to follow patients treated for temporal arteritis remains controversial.

Conclusion

Temporal arteritis is an important, but relatively uncommon, cause of headache in the elderly. A majority of patients with temporal arteritis will have an elevated ESR, but the severity of symptoms and the prognosis do not correlate with the ESR. Any patient with a low ESR and clinical signs and symptoms consistent with temporal arteritis should be treated or have a temporal artery biopsy, especially if the hematocrit is normal.

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Editorial Comment

These two rather similar cases reporting temporal arteritis (page 115 and page 119) were received independently at about the same time; therefore, we chose to publish them both. They demonstrate very effectively that we must be suspicious of temporal arteritis even if the erythrocyte sedimentation rate is not elevated. Because temporal arteritis in the elderly is sufficiently common and potentially harmful, we need to be alert to the variations of its presentation. It is hoped that by presenting these two cases, our readers will become more conscious of this clinical phenomenon.

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