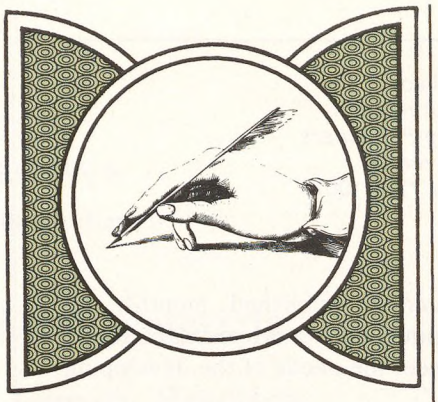
Letters to the Editor



The Journal welcomes Letters to the Editor; if found suitable, they will be published as space allows. Letters should be typed double-spaced, should not exceed 400 words, and are subject to abridgment and other editorial changes in accordance with journal style.

Letter from a Patient

The following letter from a former patient was received recently by a family physician now engaged in full-time teaching who left practice several years ago. It is presented in an anonymous but otherwise unchanged form as an example of the qualities reasonably expected by patients of their physicians.

Dear ———,

When are you coming back to town for good? Back to a small building where I can be treated with gentle dialogue, respected for who and what I am, and even be called by my first name at the beginning of a sentence now and then. Even scolded when I needed that, and even called on the phone by you first—not a return call for me.

Do you remember the Thanksgiving morning when you were puzzled by a lab report with my name on it, and you called me. My first words were "You got the wrong lady—I'm not sick, and I didn't call you." You said, "Can't I do what a good physican is supposed to do, even if it's Thanksgiving Day?" and we both laughed. I miss your laugh.

When are you coming back to a room with a blackboard in the office, so you can draw pictures for me to understand things better, since I didn't go to medical school. I still remember all the kids pictures you used to tape on the blackboard if they brought you a picture the day you were checking them. My kids never remembered you needed a picture.

I need all those things real bad right now, so if you're not coming back to our town in a small building real soon, could you, or would you tell if you know anyone who does at least some—or most of those things. I was in tears on the phone last Sunday night, talking to a doctor I've never met, all because I was trying to get the name of someone like you, to call in the morning. He was trying to be understanding, and it was my fault for not waiting until Monday morning. But my God, do you have any idea what it's like to get a doctor to talk to you on Monday morning. It took four phone calls for me to get an appointment with a specialist recently, and it wasn't even on a Monday. I miss you, and love you, and wish you'd come back to a small building in our town.

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Temporal Arteritis and Polymyalgia Rheumatica

To the Editor:

Realizing that controversial aspects of temporal arteritis (TA) and polymyalgia rheumatica (PMR) exist, Dr. Atkins must be prepared for the questions inevitably raised by his review (*Atkins SD: Giant Cell Arteritis: A systemic spectrum including temporal arteritis and polymyalgia rheumatica. J Fam Pract 7:1109, 1978*). My areas of concern are three: use of arterial biopsy, identifying symptoms which precede blindness, and use of erythrocyte sedimentation rate (ESR) during long-term treatment follow-ups.

Dr. Atkins concludes that because certain risks may be involved in arterial biopsy and skip lesions exist, which make unilateral biopsy inconclusive if normal, that biopsy itself becomes a nonessential investigation. The other line of reasoning is ignored. Here one accepts these facts, but adds further analysis and concludes that an aggressive biopsy protocol is a more logical assessment. My argument for aggressive biopsy policy accepts procedural risks, but declares the common problems from temporal artery litigation (during segmented biopsy) to be local hemostasis, wound infection, or postoperative cutaneous dysesthesiae.[1] The risk of focal cerebral ischemia is given undue emphasis by Atkins's remarks. Fisher has reported the only cerebrovascular accident related to interruption of internal carotid blood flood by temporal artery litigation; ipsilateral proximal internal carotid artery occlusion existed and small collateral anastomoses from the temporal artery fed more peripheral tributaries.[2] Taken in context, this lone report among the thousands of biopsies done stands as testimony to the relative safety of the procedure.

Unilateral biopsy is inconclusive if normal—one may have removed a skip lesion or be dealing with a patient with unilateral arteritic involvement.[3] However, there are important reasons for moving to a contralateral biopsy if the first is normal. PMR and TA are closely but, at this point, unclearly related. Some with PMR obtain relief from their symptoms with nonsteroidal anti-inflammatory agents or low (10 to 20 mg daily) corticosteroid therapy. Another 20 to 40 percent with PMR, although not distinguishable from the former group by symptoms, harbor occult TA with the same high (35 to 40 percent) risk of blindness; nonsteroidal anti-inflammatory agents or low dose steroids would be disastrous in this group since neither regimen protects from visual loss. The natural history of TA is now thought to be two years, but can be as long as 14 years.[4] Histologic confirmation should be required before such an aged population is subjected to the inherent risks of long-term corticosteroid therapy.

As a first counter to the problem presented by skip lesions, most authors recommend a 4 to 6 cm arterial segment be removed rather than the 1 cm mentioned by Atkins.[5] If negative, then a contralateral biopsy of temporal, occipital, or facial artery should be done as a second procedure—detailed sequential analysis of frozen section specimen is usually not feasible.[6,7] Emphasis should be added to the fact that palpatory abnormalities need not be present for the biopsy

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to be worthwhile. Sorenson and Lorenzen demonstrated a lack of local temporal artery induration, erythema, or tenderness on more than 50 percent of the 25 patients in their series with arteritis on biopsy; they also had a high rate of positive findings on contralateral biopsies—54 percent (7/13).[8]

A second area of concern is the implication by Atkins that visual symptoms usually occur before the onset of blindness in TA. This, quite simply, is misinterpretation of available data. It would be invaluable to have clearly identifiable indicators, but as yet no one has found any symptom, sign, or laboratory abnormality that consistently labels patients at risk for blindness. Hamilton indicates that 13 of his 14 patients with blindness as a result of TA had premonitory symptoms of the disease before visual loss—but these were not uniformly visual.[9] Simmons and Cogan described a TA variant called "occult temporal arteritis" in 1962.[10] These patients present with blindness unaccompanied by classic symptoms or local temporal artery signs despite biopsy-proven arteritis. Although the frequency with which this perplexing forme fruste occurs is unknown, there seem to be no warning signals for the clinician. The ominous importance of oculomotor ophthalmoplegia, evidenced by diplopia or ptosis, is an important sign. In Hollenhorst's review of 175 patients, 50 percent of the 34 with diplopia progressed to blindness.[11] Fisher noted that although valuable, this sign was not uniformly a harbinger of blindness.[12]

Finally, although invaluable as a device for monitoring effectiveness of corticosteroid therapy in TA, the ESR is not infallible. Exacerbations of arteritic activity, carrying risk of vascular sequelae, may occur without altering a normal ESR.[13] Reappearance of earlier symptoms of TA or PMR can be considered just as reliable an indicator of disease activity as any laboratory abnormality.

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The preceding letter was referred to Dr. Atkins who responds as follows:

In response to Dr. Goodman's thoughtful comments on "Giant

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Cell Arteritis'' (*Atkins SD: J Fam Pract 7:1109, 1978*), I concur that the erythrocyte sedimentation rate (ESR) cannot be relied upon fully as a measure of response to treatment, and that exacerbations of arteritic processes can occur before any elevation of the sedimentation rate. This is precisely why I recommend close follow-up with the patient to review not only serial sedimentation rates but evidence as well of focal and systemic exacerbations of disease.

Regarding the temporal relationship of visual symptoms to complete monocular blindness, there does not seem to be any study that specifically focuses on this problem that would suffice to prove or disprove my impression that visual symptoms usually precede complete blindness. Certainly, however, one cannot assume, in a patient with proven giant cell arteritis or even in one in whom it is highly suspected, that if there exist no visual symptoms that blindness is unlikely. There being no study that focuses on this specific problem, we must rely more or less on other indirect evidence to substantiate our impressions.

Anderson and Bayles[1] note that "Of importance is the fact that blindness rarely is the initial symptom of giant cell arteritis, although it may be. . . .Most patients have premonitory visual symptoms such as blurring of vision, amaurosis fugax (transient visual loss), iritis, conjunctivitis, scintillating scotomata, photophobia, glaucoma, or transient diplopia.''

Of 42 patients reviewed by Huston, Hunder, et al[2] with temporal arteritis, 17 displayed visual symptoms. Among these patients there were 22 episodes of visual symptoms of various sorts including blurred vision, diplopia, transient loss of vision, and permanent but partial loss, and only four episodes of permanent and complete loss of vision in one or both eyes. In this and other studies, it seems that the frequency of "visual events'' short of blindness was significantly more common than those of complete monocular blindness. It is, of course, unknown exactly what percentage of those patients with "visual events'' proceed into complete monocular blindness.

Certainly many authors do comment on the "sudden appearance'' of blindness in patients with giant cell arteritis. It is not uncommon that these episodes of "sudden blindness'' have followed a period of various visual symptoms, although the interval between the premonitory visual symptoms and the blindness that follows is frequently within hours or days of the initial visual symptom.[3-5]

The most common cause of blindness in giant cell arteritis is ischemic optic neuropathy. Given that this is the result of involvement of the short posterior ciliary vessels to the optic nerve head, and given that there are usually 5 to 6 posterior ciliary arteries supplying the optic nerve head, it is not surprising there would be a variety of sector deficits that could occur prior to total loss of vision of the eye.

Regarding temporal artery biopsy, I recommended that a 1 cm biopsy specimen would be the minimal acceptable length and that more was highly desirable. Usually the size of the biopsy segment, when explicitly discussed within the literature varied between 1 and $2^1/_2$ cm.

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Recently, Healey and Wilske[6] in their text, *The Systemic Manifestations of Temporal Arteritis*, cite a study by Kattwinkel and Fernandez-Herlihy[7] indicating that when patients have evidence of cranial arteritis (eg, visual symptoms, jaw claudication, tongue pain, or bruit), then biopsies may be positive up to 75 percent of the time; however, they noted that the percentage of positive biopsies decreased to 6 to 40 percent in patients without evidence of clinical cranial involvement. Given these relatively high incidences of negative biopsies, I question that the decision to treat with corticosteroids should rest solely on the existence of positive biopsy, at least in those especially high-risk patients with cranial symptoms. Certainly, as a practical matter, many patients and their families will not accept multiple diagnostic biopsies.

In many cases I feel that GCA will remain a clinical diagnosis. In less than obvious cases, I feel that a patient with a high suspicion of GCA, in whom it is assumed that a careful history and physical examination has been performed and the appropriate exclusionary considerations have been made, that, if the initial temporal artery biopsy is negative, it is at least as reasonable to request a consultation from a colleague familiar with this disorder as it is to employ second and possibly third diagnostic biopsies and then to withhold treatment if the biopsies are not diagnostic.

I agree that morbidity from temporal artery biopsy seems to be rather remote. Nonetheless, physicians must continue to ask themselves specifically for what reason they are considering an invasive diagnostic test. Further, one must keep in mind that the anastomosis of the superficial temporal artery to the middle cerebral artery is graduating from the experimental to the clinically applicable realm among our neurosurgical repertoire and that in certain cases the destruction of this artery may not be advisable.

Basically, what this controversy indicates more than anything else is that temporal artery biopsy is an imperfect means of documentation. It is hoped that immunofluorescent staining techniques[8] and perhaps other immunological studies[9] can some day help to assist in the diagnosis of GCA.

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