Chapter 33

The Headache That Changed

Marek Gawel, MD, FRCP

Case History

A 58-year-old lady of Hungarian decent had a 20-year history of headache. The headaches were typically bitemporal, squeezing-type pains associated with light sensitivity. They were not made worse by bending, stooping, sneezing, or coughing. They occurred on an almost daily basis and were often present when she woke up in the morning.

Previous examination had revealed no abnormality neurologically. A computed tomography (CT) scan had been normal, and full blood work had also been performed and was normal. Specifically, her complete blood count and sedimentation rate were normal.

Three years after these investigations had been performed, she presented with an exacerbation of the headache. The headache was identical in nature to her previous headache, being a squeezing pain around the temples associated with some light sensitivity. Elucidation of the history showed that she had been undergoing some stress at work. She was working as a cook in a nursing home, and there had been some layoffs so she was concerned about her job. She also found that the work was getting more and more difficult as the nursing home increased in size and she had to work under a greater pressure.

On examination, she had a blood pressure of 130/80. Her fundi, cranial nerves, power, tone, and reflexes were all normal. She had normal neck movements. There was some bilateral temporal tenderness, but this had been present on her previous examination. A CT scan was performed, which was normal. A diagnosis of chronic tension-type headache was made and she was started on amitriptyline 25 mg at night.

In follow-up, she was feeling somewhat better so she was referred back to her family doctor. Six months later, she presented at the emergency room (ER) with transient loss of vision in the left eye, which had lasted 10 minutes and cleared, but she said that her vision was still somewhat hazy. In the ER, she had another CT scan performed, and blood was drawn for hematology and electrolytes. This time, her sedimentation rate was elevated at 68 mm/min. She was referred for follow-up, and examination showed increasing tenderness in the temples to palpation.

Review of her history showed that in the last 3 months she had developed some aches in her muscles, which were stiff in the morning, and she also had some problems when chewing, in that her jaw would become painful if she had to eat something chewy such as a steak. She was immediately started on prednisone 40 mg. A temporal artery biopsy was ordered and found to be positive for temporal arteritis, with typical giant cells and inflammatory infiltrate. She was continued on prednisone, the dose being adjusted to keep her sedimentation low and herself free of headache.

As soon as she started the prednisone, she began to feel a lot better. Her headache disappeared and she found she had far less stiffness or achiness in the muscles. She was followed for 3 years; the prednisone was reduced gradually, and by the end of that time, she was able to completely discontinue it with no recurrence of the headache or the muscle stiffness.

Questions on the Case

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

- Does the location and nature of the pain give you any clues concerning the etiology?
- What is the International Headache Society definition of chronic tension-type headache?
- How often does one find transient visual obscurrations in temporal arteritis?

- What is the significance of the elevated sedimentation rate? What other tests could be done to complement this?
- What is the significane of pain in the jaw on chewing?
- What is the natural history of temporal arteritis?

Management Strategies

The primary treatment for temporal arteritis and polymyalgia rheumatica is the use of corticosteriods. Treatment should start immediately if clinical suspicion appears. Patients can go blind very quickly without treatment. Also, in some cases, positive temporal artery biopsies have been noted up to 3 to 6 months after starting treatment, and therefore treatment should not prevent a surgical diagnosis. Generally, however, it is better to do the biopsy within a week. Starting doses of 40 mg prednisolone for giant cell arteritis and 15 mg prednisolone for polymyalgia rheumatica would appear to be appropriate initial treatments. Some ophthalmologists recommend 80 mg for giant cell arteritis because of the high risk of arteritic complications. Intravenous steroid treatment has been recommended for patients who have visual signs or symptoms such as amaurosis fugax. Between a third and a half of patients can stop steroids after 2 years. Other immunosuppressants such as azathioprine or methotrexate can be used in this disorder.

A recent study by Hayreh and Zimmerman recommended high doses of oral prednisone daily (80 to 120 mg) or intravenous megadose systemic corticosteroids (usually 150 mg dexamethasone every 8 hours for 1 to 3 days) followed by oral prednisone. Deterioration subsequent to starting the steroids occurred in 6 of 48 patients on intravenous steroid therapy and in 9 of 97 patients on oral therapy. These differences are not statistically significant and they were left with no evidence to suggest that one approach was better than another.

Case Summary

- In our patient, the pain evolved from her previous headache, and she did not think at the beginning that it was any different from her previous presumed tension-type headache.
- Our patient was lucky that the visual disturbance was amaurosis fugax and not complete blindness, since the former has a better prognosis in terms of vision.
- The case points out the importance of getting a complete history and carefully examining patients with headaches persisting into old age.

Overview of Temporal Arteritis

Horton, Magath, and Brown first described temporal arteritis in 1932 as a disease mainly involving the vessels of the carotid arterial system. The pathologic features are a subacute granulomatous inflammation with typical giant cells noted. In a review by Bayard Horton in 1962, he notes that its clinical features consist of headache, painful tender nodules over the scalp, difficulty chewing, musculoskeletal aches and pains, fever, night sweats, anorexia, weakness, general malaise and lassitude, and other vague symptoms of mild sepsis.

Horton describes his discovery of the condition by first observing typically swollen nodular and reddened temporal arteritis in two patients he had seen in 1931. The description of the two cases in 1932 pointed out that it was probably a focal localization of some systemic disease. Further reports by Magath and Horton in 1937 were followed by further cases appearing in the literature, so that by 1962, 300 cases had been reported. It was noted that facial arteries became thrombosed during the course of the illness, but this was not mentioned in the original case study.

In 1935, a report from the Mayo Clinic described a women with a tender raised nodule over the radial artery at the wrist, which turned out to have the same pathology as that of temporal arteritis. She was in fact being observed for temporal arteritis herself.

In Horton's report, it is noted that the mean age for the first 105 patients studied from 1931 to 1954 was 69.1 years. He notes that there is an elevated sedimentation rate, often over 100 mm in 1 hour as determined by the Westergren method, and states that recent onset of severe headache in an older patient should alert the physician to the possibility of temporal arteritis.

In 1937, the first case of a man aged 70 years with bilateral loss of vision was described; by the time he was examined, his arteries were not tender and pulsations were not present. Apparently he went into remission, but remained completely blind, eventually dying at the age of 81 years. The loss of vision is characterized ophthalmoscopically by ischemic optic neuritis, ischemic retrobulbar neuritis, or central retinal artery thrombosis. The onset of his vision loss was described as being sudden and usually unilateral to begin with, the other eye being involved within a few days. Horton states that 31% of 105 patients whom he described developed blindness of one or both eyes with a complication. Twenty percent of patients in the world literature had bilateral blindness.

Horton points out that the headache of temporal arteritis is not diagnostic, but varies from patient to patient. In his cases, the patients felt that the headache differed from any other headaches they might have experienced previously. It was apparently more severe and persistent throughout the waking hours, and tended to grow worse at night. In his review, Horton describes further cases and notes that there may be an acute phase of the disease, which progresses quite rapidly, followed by a more chronic phase.

The pathology has been described and is essentially classical with "bilateral acute and subacute temporal arteritis and periarteritis with marked thickening and infiltration of arterial wall obliterative endarteritis and periarteritis. The cellular infiltration consists of plasma cells, lymphocytes, eosinophils, and foreign-body giant cells." At that time, Horton was of the opinion that extensive resection of the temporal arteries was the treatment of choice, with the pain disappearing immediately after the resection.

He proceeds to describe a case presenting in 1957. A patient had been given 0.5 mg of ergotamine tartrate intramuscularly, which relieved the headaches for a short time. However, the headaches returned and further injections were not effective. Following the injection of ergotamine tartrate, the patient developed bilateral intermittent claudication of the thighs. This patient at a later date had cortisone therapy, first intramuscularly and then orally, with total relief of his headaches.

A review by Swannell describes the incidence, clinical presentation, and features of temporal arteritis and polymyalgia rheumatica. He notes that temporal arteritis and polymyalgia are clinical syndromes affecting elderly people, which may occur in the same patients. These conditions produce constitutional symptoms with increased acute-phase reactants. He points out that the disease is almost always found in white people, with the highest incidence being in Scandinavia and Northern Europe (between 17 and 18 cases per 100,000 population aged over 50 years), then in France, Spain, and Israel.

Polymyalgia rheumatica alone has an incidence of 12.7 cases per 100,000 in the population aged over 50 years in Italy, whereas studies in Sweden show figures of between 20.4 and 68.3 per 100,000. Swannell quotes Jones' suggestion that the presenting symptoms may be classified as described below.

1. Systemic: malaise, anorexia, fever, night sweats, weight loss, and depression
2. Myalgic: proximal, symmetrical muscle pain, and stiffness of polymyalgia rheumatica
3. Arteritic: involvement of the artery, which may produce
   - pain, swelling, erythema, and tenderness over the affected artery
   - partial occlusion resulting in "claudication-like" symptoms
   - total occlusions resulting in ischemia and necrosis of structures supplied by the affected vessel

Involvement of any of the cranial arteries may result in necrosis or gangrene of the area of supply, thus toothache has been described, as well as gangrene of the scalp. Involvement of the vertebrobasilar and carotid circulation may lead to stroke or coronary involvement and can result in myocardial infarction. The arteritis may also involve other areas, leading to aortic incompetence and aortic artery syndrome.

Swannell points out that a temporal artery biopsy is the only diagnostic investigation, but samples may be only positive in 60 to 80% of patients with temporal arteritis and in 15 to 20% of patients with polymyalgia rheumatica. Elevation of the erythrocyte sedimentation rate is important in helping make the diagnosis, but C-reactive protein may also be important, whereas other abnormalities such as increased alkaline phosphatase of liver origin occurs in a third to a half of the patients. This may be in normochromic anemia, as well as thrombocytosis and raised immunoglobulin G levels. He states that close attention should be paid to the history, while all the peripheral pulses should be examined, as well as palpation of the temporal, posterior, auricular, facial, and occipital arteries to elicit tenderness.

An analysis of surgical efficiency of temporal artery biopsies was performed by Galloway and colleagues. In this study, it was found that ophthalmology does most of the biopsies and has the better pick-up rate, since they removed longer pieces of the temporal artery.

In contrast to Horton's descriptions of the acute onset of temporal arteritis, Liozon and colleagues described what they call silent or masked giant cell arteritis. These patients generally presented with a fever of unknown origin, and had a higher mean sedimentation rate and C-reactive protein, with low levels of albumin and hemoglobin. They felt that this type may represent a distinct subset of giant cell arteritis marked by a protracted inflammatory response and a relatively benign short-term outcome, with excellent response to corticosteroids and no visual ischemic events, despite being more difficult to diagnose.

A case report by Cockerham and colleagues described a 75-year-old women who developed acute loss of vision in the right eye, ipsilateral periocular pain, an afferent pupillary defect, sectoral optic disc edema, and later, ipsilateral proptosis and intraconal mass. In this case, the sedimentation rate was normal, while biopsy of the orbit showed a granulomatous inflammation, and a temporal artery biopsy showed changes consistent with temporal arteritis. The patient appears to have had treatment with external beam radiation with complete efficacy.

An intriguing study by Murgatroyd and colleagues suggests that ultrasound of the temporal artery may be a useful adjunct to the temporal artery biopsy. Affected arteries have a hypoechoic or "halo" effect.

A study done in Sweden by Nordborg and colleagues suggests that the incidence of temporal arteritis is actually increasing, especially in younger women aged 50 years or older. It was thus stated that it was unlikely to be due to an increased age of the general population but to other factors, however the authors do not specify which factors. It was also suggested that giant cell arteritis was not truly infectious vasculitis, but that infection could be a triggering factor.

Treatment in the elderly patient over the age of 75 years is described by a French group, who recommend that antiaggregant or anticoagulant treatment ought to be given at the beginning of steroid treatment to prevent ischemic complications.

A study by Soloman and Cappa on the demographics of headache in older people examined patients aged 65 years or older. Fifteen percent of their patients presenting with headaches over the age of 65 years had temporal arteritis. In their study, both temporal arteritis and migraine were predominantly seen in women, and the average age of onset of temporal arteritis was 73.7 years, whereas benign headache patients had an average age of onset of 73.5 years.

To point out that the headache of temporal arteritis can be nonspecific, they reviewed the records of 24 patients with biopsy-proven temporal arteritis to determine the site of pain and headache. The temporal area was the sole site of the headache in only 6 of 24 patients; the temple was included in pain that affected the frontal vertex and occipital areas in another 7 patients; 2 patients had generalized headaches; 7 patients did not have temporal pain; and 2 patients had no headache at all.

Selected Readings

Chevalet P, Masseau-Imbert A, Durand-Fix MH, et al. Giant cell arteritis after the age of 75. Ann Med Interne (Paris) 2002;153:373–7.

Cockerham KP, Cockerham GC, Brown HG, Hidayat AA. Radiosensitive orbital inflammation associated with temporal arteritis. J Neuroopthalmol 2003;23:117–21.

Galloway GD, Klebe B, Riordan-Eva P. Surgical performance for specialities undertaking temporal artery biopsies: who should perform them? Br J Ophthalmol 2002;86:250.

Hayreh SS, Zimmerman B. Visual deterioration in giant cell arteritis patients while on high doses of corticosteroid therapy. Opthalmology 2003;110:1204–15.

Horth BT, Magath TB. Arteritis of the temporal vessels. Report of seven cases. Proc Staff Meet Mayo Clinic 1937;12:548–53.

Horton BT. Headache and intermittent claudication of the jaw in temporal arteritis. Headache 1962;11:30–9.

Horton BT, Magath TB, Brown GE. An undescribed form of arteritis of the temporal vessels. Proc Staff Meet Mayo Clinic 1932;7:700–1.

Horton BT, Magath TB, Brown GE. Arteritis of the temporal vessels. A previously undescribed form. Arch Intern Med 1939;53:400–9.

Jones JG. Clinical features of giant cell arteritis. In: Hazleman BL, Bengtsson B-A, editors. Baillière's clinical rheumatology. London: Baillière Tindall; 1991. p. 414.

Liozon E, Boutros-Toni R, Ly K, et al. Silent, or masked, giant cell arteritis is associated with a strong inflammatory response and benign short term course. J Rheumatol 2003;30:1272–6.

Murgatroyd H, Nimmo M, Evans A, MacEwen C. The use of ultrasound as an aid in the diagnosis of giant cell arteritis: a pilot study comparing histological features with ultrasound findings. Eye 2003;17:415–9.

Nordborg C, Johansson H, Petrusdottir V, Nordberg E. The epidemiology of biopsy-positive giant cell arteritis: a special reference to changes in the age of the population. Rheumatology (Oxford) 2003;42:549–52.

Soloman S, Cappa KG. The headache of temporal arteritis. J Am Geriatr Soc 1987;35:163–5

Swannell AJ. Polymyalgia rheumatica and temporal arteritis: diagnosis and management. BMJ 1997;314:1329–32.

Editorial Comments

Few causes of headache have traditionally been of such great concern to clinicians and in particular neurologists as "temporal arteritis." By definition, the name of the disorder is misleading, as the "arteritis" can involve other intracranial and extracranial sites. Also, most clinicians have spent a lifetime in search of the elusive elevated erythrocyte sedimentation rate in older patients with headache who have other "systemic symptoms," along with subjecting many patients to temporal artery biopsies, most of which were negative or nondiagnostic, to say the least. This case by Dr. Gawel is important for many reasons, not the least was what appears to be a long gestation of symptoms until the diagnosis was clear, and of course, the nonspecific nature of symptoms in this disorder. Another important reason is to remind us that when the headache significantly worsens or changes, we have to start the search again. Even when two magnetic resonance imaging scans are negative, or blood work is unrevealing, the changing headache deserves our undivided attention. The historic review is welcome in this case, and this case complements another case in this volume (see Chapter 55, "The Elderly Woman with Sharp, Shooting Orbitotemporal Pains and Visual Loss").

Final diagnosis:

Temporal arteritis