

GIANT CELL ARTERITIS: REPORT OF TWO SAUDI PATIENTS AND REVIEW OF THE LITERATURE

Abdulrahman Al Tahan, MRCP; Molham Al Rayess, MRCPath;
Mohammad Abduljabbar, FRCP; Mansour Al Moallem, ABIM

Giant cell, or temporal, arteritis (GCA) is a necrotizing arteritis of unknown etiology, described as a distinct clinicopathological entity in 1932 by Horton et al.¹ It afflicts elderly people, presenting with severe headache, tenderness of the temporal arteries, highly elevated erythrocyte sedimentation rate (ESR) and abnormal temporal artery biopsy.¹⁻⁴ Although GCA has been recognized worldwide, it has not been previously reported in the Saudi population, even in large population-based epidemiological studies.⁵⁻¹⁰ We describe the clinical, laboratory and pathological findings of two Saudi patients with a review of relevant literature.

Case 1

A 78-year-old Saudi male presented with a six-month history of severe, persistent headache and general malaise associated with a burning sensation in both temples, worse on the left. He had been blind in the left eye from childhood and had bilateral cataracts. Both temples were tender, but temporal arteries were pulsating. Apart from a mild pyrexia, the rest of his physical examination was normal.

Investigations included hemoglobin of 11.3 g/L, ESR (on two occasions) of 114 and 120 mm/1st hour. Head CT scan and cerebrospinal fluid examination were normal. Biopsy from the left temporal artery confirmed the diagnosis of GCA (Figure 1). He was started on 80 mg of prednisolone with rapid clinical improvement. The ESR returned to normal and he was stable on follow-up for the past year.

Case 2

A 74-year-old Saudi female presented with a six-week history of severe bilateral, mainly left-sided headache

associated with generalized weakness and weight loss. She has long-standing arterial hypertension and bronchiectasis. She was afebrile with normal visual acuity. Both temporal arteries were tender and nonpulsating, and her proximal muscles were weak and painful. Investigations revealed normochromic, normocytic anemia with hemoglobin of 10.5 g/L. ESR values were 62 and 85 mm/1st hour on two occasions. The transaminases were moderately elevated, with negative serology for hepatitis viruses and normal creatine kinase. Head CT scan was normal. Treatment with 60 mg of prednisolone resulted in rapid relief of the headache within a few days. Temporal artery biopsy done after eight days revealed features consistent with healed GCA of irregular fibrosis and scarring of the media with long breaks of internal elastic lamella and foci of mononuclear inflammatory cells. However, she stopped her medication after a short time and suffered a fatal stroke three months later. Her ESR then was 90 mm/1st hour.

Discussion

These cases illustrate the typical features of GCA, which form the criteria selected by the American College of Rheumatology for the format classification of GCA and include age ≥ 50 years, recent localized headache, temporal artery tenderness, ESR ≥ 50 mm/hour, and a positive temporal artery biopsy.⁴ All criteria are noted in the first patient, while only four occurred in the second. The presence of three or more of these criteria is associated with more than 90% sensitivity and specificity.⁴ The age criterion is of special importance, as this disease is almost exclusively seen in patients over 50 years old.²⁻⁴ The incidence increases dramatically with age, rising by almost tenfold in the ninth decade.⁶ It is also worth noting that the symptom of visual loss was not included in this criteria because, contrary to long-held views, it was shown to be less common than previously considered.^{4,11,15} It is also of low specificity compared with other symptoms, such as claudication of the jaw and the tongue on deglutition which is considered highly specific and was included in another classification.⁴

From the Department of Medicine, Divisions of Neurology (Drs. Al Tahan, Abduljabbar, Al Moallem) and Pathology (Dr. Al Rayess), King Khalid University Hospital, Riyadh.

Address reprint requests and correspondence to Dr. Al Tahan: Associate Professor, King Khalid University Hospital, P.O. Box 7805 (38), Riyadh 11472, Saudi Arabia.

Accepted for publication 16 September 1996. Received 24 July 1996.



FIGURE 1. Low (A) and high (B) magnification of the temporal artery biopsy in the first patient showing features of active giant cell arteritis: narrowing of the lumen, intimal edema, transmural inflammatory infiltrates including giant cells (arrowhead) and disruption of the internal elastic lamella (H & E, 100x).

Apart from these typical features, GCA may present with a wide variety of clinical manifestations, which probably contribute to its recognized underdiagnosis.¹¹ Almost any large or medium-sized artery in the body may be involved, including those of the limbs, liver, intestine, lungs, uterus, breast and skin.^{2-4,12-14} Involvement of the aorta and coronary arteries may lead to a dissecting aortic aneurysm or myocardial infarction, which, together with cerebral infarction, are considered the most common causes of death in GCA.¹³ The latter was probably the cause of death in our second patient. Neurological involvement, however, is the most common, occurring in 30% to 40% of patients.^{14,15} Complications include cerebrovascular accidents, neuro-ophthalmologic and neuro-otologic syndromes, neuropsychiatric disorders, tremor, aseptic meningitis and different types of peripheral neuropathy.¹⁴⁻¹⁶

In view of these variable clinical presentations of GCA, the value of pathological confirmation becomes more important, especially in atypical cases. The characteristic pathological features of active GCA are illustrated in the biopsy findings of our first patient, and include the presence of giant cells, in association with the different features of necrotizing arteritis (Figure 1). However, it is worth stressing that these features hold the same value in the diagnosis of GCA, even in the absence of giant cells.^{4,17} Other recognized pathological findings are those of healed arteritis, which were seen in our second patient, and are distinguished from changes seen in arteriosclerosis by the presence of focal mononuclear aggregates, and the long breaks in elastic lamella.¹⁷ A normal biopsy, on the other hand, is not uncommon, reaching 40% and even 80% in patients treated with steroids for more than one week.¹⁷⁻¹⁸ Negative biopsies may be minimized by earlier biopsy and by taking a long segment of the artery from areas showing palpable tender lesions.¹⁹

Another important investigation in the diagnosis of GCA is the elevated ESR, which is highly sensitive but of low specific (47.7%).⁴ A normal ESR, however, does not exclude the diagnosis of GCA.^{4,20}

Other abnormal tests include normocytic normochromic anemia, thrombocytopenia, low serum albumin, and elevated levels of hepatic enzymes, all of which are of little help in the diagnosis.²⁻⁴

Once the diagnosis of GCA is seriously considered, treatment with steroids should be instituted at once in order to prevent serious complications.^{3,21} The optimum dose of prednisolone is still controversial and extends from 20 to 80 mg per day, with a maintenance dose of 20 mg for about two years.^{11,22} The use of intravenous methylprednisolone was reported to offer better results for recovery of vision and protection of the other eye.²³ However, no controlled studies are available to confirm this claim.

Polymyalgia rheumatica is a self-limiting disorder afflicting mainly elderly persons and characterized by severe aching and stiffness of proximal limb muscles in association with constitutional symptoms. It is accepted as part of the clinical spectrum of GCA, occurring concomitantly with temporal arteritis in up to 50% of patients.²⁻⁴ Our second patient probably suffered from both conditions simultaneously.

The etiology of GCA is still unknown, and different hypotheses blame both environmental and genetic factors, with increasing evidence suggesting an autoimmune mechanism directed against aging elastin.²⁴

The incidence of GCA varies widely in the West, and ranges from 6:100,000 in Spain to 27:100,000 in Iceland.^{8,9} Racial factors apparently play an important role in these variations. The incidence is higher in populations of Nordic origin,^{5,6,9} while it is significantly lower in blacks.⁷ GCA is very rare in Orientals and the first two

cases were only recently reported from China.²⁵

GCA has not been reported before from Saudi Arabia, either as individual cases or in epidemiological studies. Although underdiagnosis and/or underreporting probably contribute to this state, we may conclude that Saudi Arabia is probably one of the low incidence countries. This may have a racial or environmental cause or both. However, the Saudi population is relatively young at the moment, with individuals above the age of 50 years accounting for only 4% of the population.¹⁰ As the ratio of elderly people is expected to rise with improvement of living standards, the incidence of GCA will be expected to rise as well. Physicians involved in the care of the elderly should be alert, keeping in mind the wide range of serious but preventable complications associated with GCA.

References

- Horton BT, Magath TB, Brown GE. An undescribed form of arteritis of temporal vessels. *Mayo Clin Proc* 1932;7:700-1.
- Calamia KT, Hunder GG. Clinical manifestations of giant cell (temporal) arteritis. *Clin Rheum Dis* 1980;6:389-403.
- Hunder GG. Giant cell (temporal) arteritis. *Rheum Dis Clin North Am* 1990;16:399-409.
- Hunder GG, Bloch DA, Michel BA, et al. The American College of Rheumatology 1990 Criteria for the Classification of Giant Cell Arteritis. *Arthritis Rheum* 1990;33:1122-8.
- Sorensen PS, Lorenzen I. Giant-cell arteritis, temporal arteritis and polymyalgia rheumatica. A retrospective study of 63 patients. *Acta Med Scand* 1977;201:207-13.
- Bengtsson BA, Malmrall BE. The epidemiology of giant cell arteritis including temporal arteritis and polymyalgia rheumatica: incidences of different clinical presentations and eye complications. *Arthritis Rheum* 1981;24:899-904.
- Smiser CA, Fidler WJ, Pinals RS. The epidemiology of giant cell arteritis, report of a ten-year study in Shelby County, Tennessee. *Arthritis Rheum* 1983;10:1214-9.
- Gonzalez Gay MA, Alonso MD, Aguero JJ, Bal M, Fernandez Cambor B, Sanchez Andrade A. Temporal arteritis in a northwestern area of Spain: study of 57 biopsy-proven patients. *J Rheumatol* 1992;19:277-280.
- Baldursson O, Steinsson K, Bjornsson J, Liet JT. Giant cell arteritis in Iceland: an epidemiologic and histopathologic analysis. *Arthritis Rheum* 1994;37:1007-12.
- Al Rajeh S, Bademosi O, Awada A, Al Freihi H, Barollosi M, Al Shammari S, et al. A community survey of neurological disorders in Saudi Arabia: the Thugbah study. *Neuroepidemiology* 1993;12:164-78.
- Mason JC, Walport MJ. Giant cell arteritis: probably underdiagnosed and overtreated. *Br Med J* 1992;305:67-8.
- Wilkinson IMS, Russell RWR. Arteries of the head and neck in giant cell arteritis. *Arch Neurol* 1972;27:378-91.
- Nordborg E, Bengtsson BA. Death rates and causes of death in 284 consecutive patients with giant cell arteritis confirmed by biopsy. *Br Med J* 1989;299:549-550. *Arch Neurol* 1972;27:378-91.
- Huston KA, Hunder GG, Liet JT, Kennedy RH, Elveback LR. Temporal arteritis: a 25-year epidemiologic, clinical and pathologic study. *Ann Intern Med* 1978;88:162-7.
- Casselli RJ, Hunder GG, Whisnant JP. Neurologic disease in biopsy-proven giant cell (temporal) arteritis. *Neurology* 1988;38:352-9.
- Casselli RJ, Daube JR, Hunder GG, Whisnant JP. Peripheral neuropathic syndromes in giant cell (temporal) arteritis. *Neurology* 1985;38:685-9.
- Allsop CJ, Gallagher PJ. Temporal artery biopsy in giant-cell arteritis: a reappraisal. *Am J Sur Pathol* 1981;5:317-323.
- Allison MC, Gallagher PJ. Temporal artery biopsy and corticosteroid treatment. *Ann Rheum Dis* 1984;43:416-7.
- Albert DM, Ruchman MC, Keltner JL. Skip areas in temporal arteritis. *Arch Ophthalmol* 1976;94:2072-7.
- Biller J, Asconapé J, Weinblatt ME, Toole JF. Temporal arteritis associated with normal sedimentation rate. *JAMA* 1982;247:486-7.
- Aiello PD, Trautman JC, McPhee TJ, Kunselman AR, Hunder GG. Visual prognosis in giant cell arteritis. *Ophthalmology* 1993;100:550-5.
- Kyle V, Hazleman BL. Stopping steroids in polymyalgia rheumatica and giant cell arteritis. *Br Med J* 1991;300:344-5.
- Rosenfield SI, Kosmorsky GS, Klingele TG, Burde RM, Cohn EM. Treatment of temporal arteritis with ocular involvement. *Am J Med* 1986;80:143-5.
- Hunder GG, Lie JT, Goronzy JJ, Weyand CM. Pathogenesis of giant cell arteritis. *Arthritis Rheum* 1993;36:757-61.
- Wing YK, Kay RL, Lai FM. Giant cell arteritis in two Chinese patients. *Aust N Z J Med* 1991;21:751-2.