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Case Report

# Giant Cell (Temporal) Arteritis With Anterior Ischemic Optic Neuropathy: A Biopsy-proven Case in Taiwan

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Giant cell arteritis with arteritic anterior ischemic optic neuropathy has rarely been diagnosed in Taiwan. Recently, we encountered a 76-year-old Taiwanese patient who presented with right visual impairment and marked pale swelling of his right disc. He also suffered body weight loss, general malaise and many typical manifestations of giant cell arteritis, such as jaw claudication, a tender, non-pulsating engorgement of his temporal arteries, and a highly elevated erythrocyte sedimentation rate and C-reactive protein level. Biopsy of his right superficial temporal artery revealed a granulomatous inflammation with multinucleated giant cell infiltration. This was a biopsy-proven case of giant cell arteritis with arteritic anterior ischemic optic neuropathy and indicated that although rare, this disease could occur in patients in Taiwan.

**Key Words:** anterior ischemic optic neuropathy, biopsy, Chinese, giant cell arteritis, temporal arteritis, Taiwan

Giant cell arteritis (GCA), also referred to as temporal arteritis, is an inflammatory vasculitis that mainly involves large and medium vessels, particularly the cranial branches of the aorta. Superficial temporal, occipital, vertebral, ophthalmic and posterior ciliary arteries are the most frequently affected vessels. It has a predilection for elderly patients, with a mean presenting age of 70 years.[1] Ocular involvement might occur in up to 50–70% of patients with GCA[1,2] and represents a true ophthalmic emergency because the possibility of visual loss is very high if it is not recognized and treated promptly.[1]

The incidence of GCA varies widely in different areas throughout the world. In Caucasian people, the disease is not uncommon.[3] The highest worldwide incidence of GCA occurs in Scandinavia, with an annual incidence between 20.4 and 32.8 per 100,000 for the population aged > 50 years. In contrast, the incidence is generally very low in African, Hispanic and Asian countries.[3] Although the incidence among the Chinese is not known, it is believed to be very low. As far as we are aware, there have only been a few anecdotal case reports in the literature.[4–8] In Taiwan, biopsy-proven cases of GCA are even rarer.[9] Recently, we examined and treated an elderly Taiwanese patient with anterior ischemic optic neuropathy (AION) and many typical manifestations of GCA, such as jaw claudication, a tender, non-pulsating engorgement of the



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temporal artery, and highly elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level. Biopsy of his right superficial temporal artery revealed a granulomatous inflammation with multinucleated giant cell infiltration. Because of the paucity of studies of GCA in Taiwan, we believe that this report could be beneficial to our colleagues.

## Case Report

A 76-year-old Taiwanese man of Chinese descent with sudden onset of painless visual field loss and visual decrease in his right eye for 5 days was referred to our ophthalmological department on September 27, 2007. Visual field testing in the local clinic showed severe arcuate scotoma of his right eye and mild arcuate scotoma of his left eye (Figure 1). He had been suffering scalp tenderness that radiated to the bilateral temporal area, and jaw claudication for 6 weeks. Anorexia, general malaise and body weight loss were also noted during this period. He had a 3-year history of diabetes mellitus under good medical supervision. No chest pain or breathing difficulty was noted. The right vision deteriorated very quickly from 6/7.5 to 6/20 in 2 days. He was then admitted for further treatment. On examination, his best corrected visual acuity was 6/20 in the right eye and 6/6 in the left. The intraocular pressure of his right and left eye was 9 mmHg and 7 mmHg, respectively. Both eyes were freely movable and non-tender upon eye movement. A relative afferent pupillary defect of light reflex was noted in his right eye. Ophthalmoscopic examination revealed a pale swelling of the right optic disc (Figure 2). A mild color disturbance was noted on an Ishihara color plate test (5/15 errors, normal ≤ 4/15) and a Farnsworth D-15 color discrimination test (one error line in

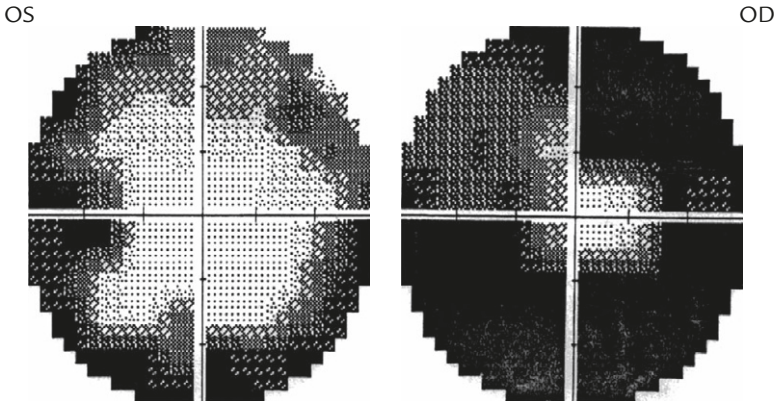


**Figure 1.** An automatic visual field test showed more marked arcuate scotomata of both upper and lower field of right eye (OD) than left eye (OS).

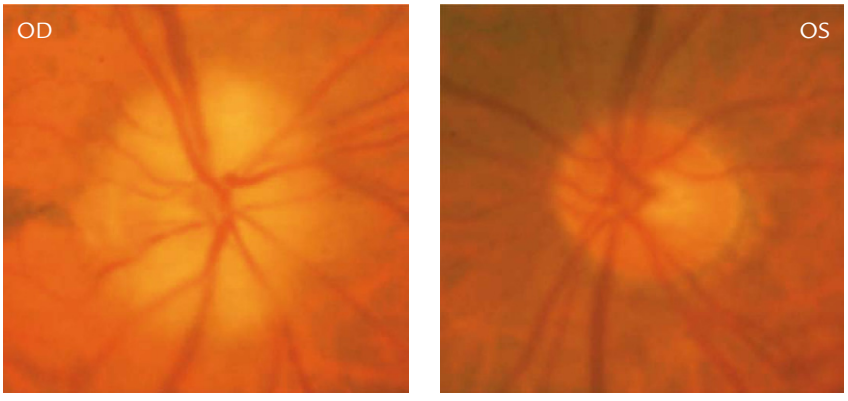


**Figure 2.** Fundus pictures showed marked pale swelling of the right optic disc (OD) as compared to left optic disc (OS).

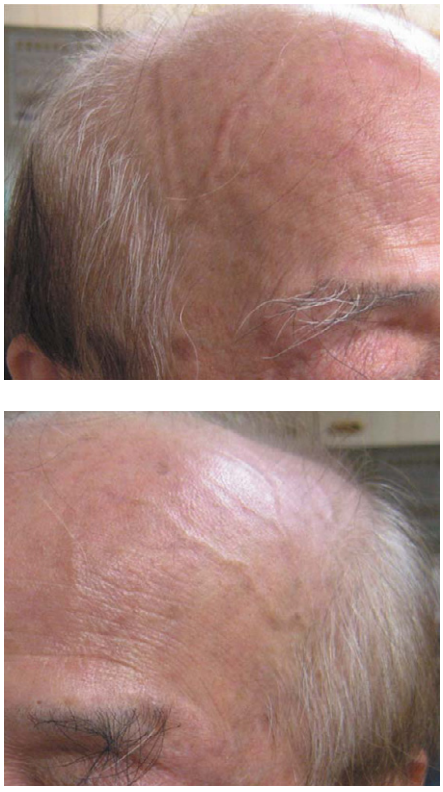

**Figure 3.** Bilateral superficial temporal arteries showed engorgement and torturosity (upper: right side, lower: left side).

the yellow-green confusion axis) of his right eye. Both superficial temporal arteries were engorged, hardened and non-pulsating (Figure 3). Investigation revealed an ESR of 114 mm/hr and a CRP level of 2.11 mg/dL (normal, < 0.5 mg/dL). His hemoglobin was 13.0 g/dL and the blood cell profile was within normal limits. The serum anticardiolipin antibody was 13.8 GPLU (normal, < 11 GPLU). A chest X-ray, renal and liver function tests, other immunological factors such as antinuclear antibody, and brain magnetic resonance imaging were all normal.

A biopsy of the right superficial temporal artery of about 10 cm in length was performed. The histopathological examination revealed duplication and fragmentation of the internal elastic lamina of the artery. The vascular wall was thickened and was infiltrated with lymphocytic and multinucleated giant cells (Figure 4).

The patient was treated with 1 mg/kg/day oral prednisolone. The patient's symptoms improved rapidly, with subsidence of jaw claudication, general malaise and temporal headache. The ESR

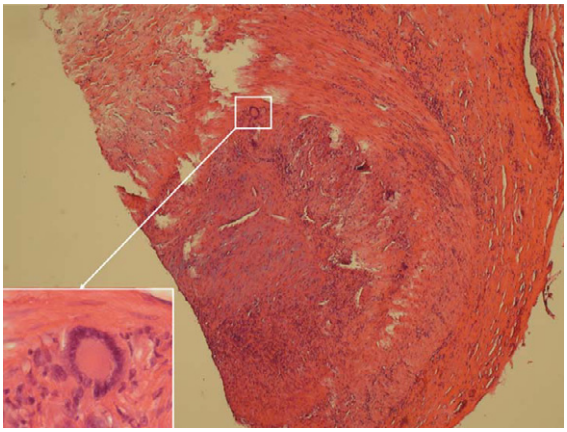

**Figure 4.** The histopathological examination of right superficial temporal artery biopsy revealed thickened vascular wall with infiltration of lymphocytes and multinucleated giant cells. (Inlet) A typical multinuclear giant cell.

decreased to 19 mm/hr and CRP level decreased to < 0.1 mg/dL within 10 days of treatment. The visual acuity improved after treatment and recovered 2 months later, going from 6/20 to 6/8.6. At the time of the preparation of this article, the patient was still being treated with oral prednisolone on a careful tapering schedule, with monthly monitoring of ESR and CRP level for over 6 months.

## Discussion

This article presents a case of arteritic anterior ischemic optic neuropathy (A-AION), with typical pathological findings of multinucleated giant cell infiltration of the temporal artery in a Taiwanese patient of Chinese descent. To the best of our knowledge, there has been no report of a biopsy-proven case of A-AION in Taiwan. In 2005, Chen et al[9] reported on two Taiwanese GCA patients with headache and general malaise. Biopsies of the temporal arteries of these patients showed lymphocytic and mononuclear cell infiltration. In one patient, there were occasional multinucleated giant cells found on microscopic examination. However, unlike our patient, neither of these patients had ocular symptoms and signs of ischemic optic neuropathy.

A-AION is the most frequent ocular complication of GCA. It can cause acute visual loss in

one or both eyes. Pale disc edema with occasional splinter hemorrhages at the disc margin can be seen on ophthalmoscopic examination. Visual loss varies from a minimal disturbance to profound impairment with no light perception. Visual field defects most often include an altitudinal hemivisual field defect, a sector defect, or a central scotoma. Sometimes, other types of visual field defects can be found.[1] Our case presented with a visual disturbance in the right eye and pale swelling of the optic disc. The visual field defect in our case showed arcuate scotomata of the upper and lower visual fields. Although the patient did not complain of visual disturbance in his left eye, the left visual field test also revealed a mild peripheral arcuate visual field defect, which suggested early involvement of his left eye. It is fortunate that the visual disturbance had not yet progressed to the catastrophic status, and could be recovered to a mild subnormal level after prompt treatment. Ischemic optic neuropathy usually progresses very rapidly; therefore, prompt treatment with an adequate dosage of steroids is essential to save visual function. Involvement of the fellow eye is also a frequent complication, even during steroid therapy.[1] Close follow-up of the fellow eye while treating these patients is important.

In our case, the combination of typical systemic signs and symptoms and an AION led us to suspect GCA. The elevated ESR and CRP level indicated the need for prompt steroid treatment and biopsy of the superficial temporal artery. ESR and CRP level are considered the most highly predictive laboratory factors of CGA, with a combined sensitivity of up to 99.2%.[10,11] They are also very useful for monitoring GCA activity and regulating steroid therapy. Our case had an ESR of 114 mm/hr and a CRP level of 2.11 mg/dL, which were far above normal levels. In the treatment course of this patient, ESR and CRP level were regularly checked in order to monitor the disease and the effectiveness of steroid treatment.

In our case, anticardiolipin antibody was elevated. Anticardiolipin antibodies can be present in up to 50% of patients with GCA.[12] In long-term follow-up of 58 patients with biopsy-proven GCA, Liozon et al[13] found that increased serum levels of anticardiolipin antibodies correlate well with clinical recurrence during or after treatment, and might help exclude infection as a cause of recurrent symptoms. However, their significance and the association with ischemic complications or ocular complications of GCA are still not fully understood.[12]

Temporal artery biopsy (TAB) is the gold standard for diagnosis of GCA. To establish the diagnosis and prevent treatment delay of this potentially blinding disease, TAB should be performed as soon as possible in every patient suspected of having GCA or A-AION.[1] The American College of Rheumatologists have proposed criteria for the histological diagnosis of GCA, which include histological evidence of necrotizing arteritis in the temporal artery, with predominantly mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells.[14] Although the presence of giant cells in the biopsy is not essential for pathological diagnosis, some authors have advocated that the presence of giant cells might be associated with a higher rate of visual impairment.[15] False negative results have been reported in 5–13% of temporal artery biopsies in GCA cases. The rate of false negatives might be associated with the length of the specimen obtained, the presence of skip lesions, pathological sectioning techniques, and duration of treatment before biopsy. To reduce the chance of false negative results caused by skip lesions, it is recommended that, in TAB, at least a 2.5-cm section is obtained, together with more closely spaced sections (0.25 mm or 0.5 mm) for pathological examination.[1] A long TAB (about 10 cm) was performed in our case, which revealed typical pathological features of granulomatous inflammation of arterial walls, with infiltration of multinucleated giant cells and mononuclear cells. It provided unequivocal evidence for the diagnosis of GCA and support for continued treatment with corticosteroid.

In conclusion, this report presented a biopsy-proven case of CGA with A-AION and indicated that, although very rare, it can occur in patients in Taiwan. A high sensitivity to the signs and

symptoms of GCA in every AION case, a rapid diagnostic work-up such as ESR, CRP and TAB, and prompt treatment with corticosteroid could be necessary to save the vision of patients with complications of GCA.

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