

# Temporal Arteritis

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Temporal arteritis, a chronic inflammatory vasculitis involving medium- and large-sized arteries, has rarely been reported in Asia. However, we report 2 cases, in which the patients initially presented with headache. Physical examination disclosed engorged, hard and palpable vessels in the temporal areas. Temporal-artery biopsy revealed 2 different types of arteritis: the multinucleated giant cell type and the panarteritis type without multinucleated giant cells. One patient was positive for immunoglobulin G anticardiolipin antibody. The pathologic findings of the different subsets of temporal arteritis, and the relationship between anticardiolipin antibody and the extent of vascular complications of temporal arteritis, are discussed. [*J Chin Med Assoc* 2005;68(7):333–335]

**Key Words:** anticardiolipin antibody, multinucleated giant cell, temporal arteritis

## Introduction

Temporal arteritis, also referred to as giant cell arteritis, is a disease with specific clinical manifestations (in the advanced stages) and histopathologic findings.<sup>1</sup> The incidence of temporal arteritis varies widely in different populations, regions, and races. The highest incidences are found in Scandinavians, and in Americans of Scandinavian descent. In North America, temporal arteritis is not uncommon: the incidence in individuals over 50 years of age is estimated at 0.017%.<sup>2</sup> The most common clinical manifestations of temporal arteritis are nonspecific, and include headache, low-grade fever, sweating, anorexia, weight loss, and general malaise. More specific manifestations, such as claudication of the jaw, or engorged tender vessels, always develop in advanced stages of the disorder. In Taiwan, the prevalence of temporal arteritis, including that of biopsy-proven cases, has not been reported.

Histologically, 2 distinct types of temporal arteritis have been proposed: a granulomatous multinucleated giant cell type and a panarteritis with mixed inflammatory cell infiltration.<sup>3</sup> The clinical significance of the different pathologic types of temporal arteritis

remains to be investigated. Anticardiolipin (aCL) antibody, an autoantibody with immunologic reactivity against phospholipids, plays a role in the thrombosis of some autoimmune diseases, such as antiphospholipid syndrome and systemic lupus erythematosus. In temporal arteritis, thrombosis in the affected vessel is found only occasionally; however, about 1-third of patients are positive for aCL antibodies.<sup>4</sup> Herein, we report 2 cases of temporal arteritis with different pathologic features, and in which 1 of the 2 patients was positive for aCL antibodies.

## Case Reports

### Case 1

A 76-year-old man was admitted to Taipei Veterans General Hospital on 23 June 2003 because of throbbing headache over the left temporal area that had lasted for 2 weeks, and intermittent claudication over the masseter area after meals. Three months before admission, general malaise had developed and the patient had lost 6 kg in body weight. No pain or stiffness over the neck, shoulder, or pelvic girdle was noted. On admission,

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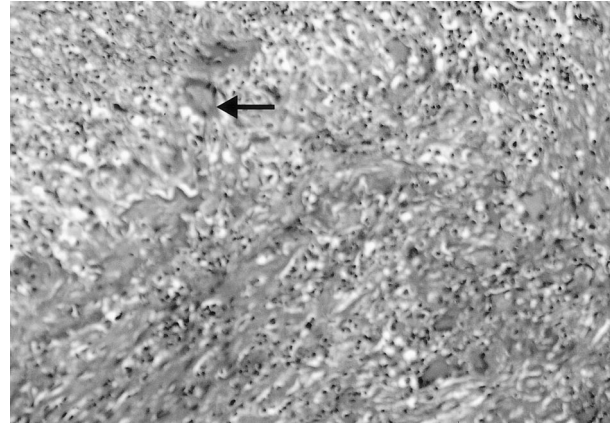
physical examination disclosed a hard, engorged vessel over the left frontotemporal area of the scalp. Sonographic studies showed complete obliteration of the left temporal artery. Biopsy of this vessel and pathologic studies revealed moderate destruction of the whole muscle layer, interruption of the internal elastic layer, and chronic infiltration of inflammatory cells, including granulomatous multinucleated giant cells (Figure 1). Further sonographic studies exploring the extent of vessel involvement in the bilateral common carotid arteries, internal carotid artery, vertebral and subclavian arteries, were unremarkable. Although the patient had no visual problems, a small delayed filling defect in the retinal vein was detected by fluorescent angiography.

Laboratory studies disclosed elevation of erythrocyte sedimentation rate (ESR; 119 mm/hr) and serum level of C-reactive protein (CRP; 7.21 mg/dL; normal < 0.50 mg/dL). The immunoglobulin G (IgG) aCL antibody was positive (28 GPL; normal < 15 GPL). The following tests were all within normal limits: complete blood count; urinalysis; routine stool cultures; hepatic and renal function tests; partial prothrombin time (PT); activated partial thromboplastin time (aPTT); and serum levels of IgG, IgA, IgM, anti-thrombin III, protein C, and protein S. Antinuclear antibody (ANA), anti-dsDNA, and antineutrophil cytoplasmic antibodies, were also negative.

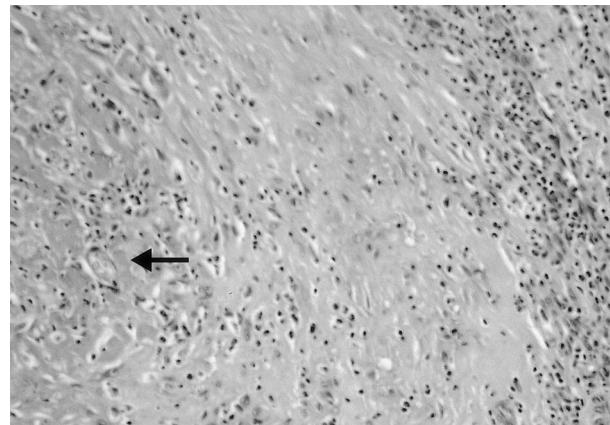
After diagnosis, the patient was treated with prednisolone 10 mg/day. The engorged temporal artery had become flat and less tender 2 weeks later; also, headache, mouth claudication and general malaise improved gradually but markedly. ESR declined from 119 mm/hour to 45 mm/hour 3 weeks after treatment.

### Case 2

A 74-year-old man visited our hospital on 4 September 2003 because of general malaise, a nodule-like lesion, and vague headache over both temporal areas of the head, and intermittent mouth claudication after meals for 1 month. No pain or stiffness was noted over the neck, shoulder, or pelvic girdle. Physical examination disclosed engorged and firm vessels over both temporal areas, and sonographic studies showed focal dilation and thickening of vascular walls in these vessels. Biopsy of the left temporal artery revealed fibrinoid necrosis, thickening of the intimal layer, interruption of the internal elastic lamina, and chronic inflammatory cell infiltration throughout the vascular layer with epithelioid histiocytes, plasma cells, lymphocytes, and eosinophils (Figure 2). The bilateral common carotid arteries, internal carotid artery, vertebral and subclavian arteries were unremarkable in sonographic studies.



**Figure 1.** Case 1: biopsy from the temporal artery showing destruction of the muscle layer with granulomatous giant cells (arrow). (Hematoxylin & eosin,  $\times 200$ .)



**Figure 2.** Case 2: biopsy from the temporal artery showing fibrinoid necrosis of the artery (arrow) and panarteritis with mixed-cell inflammation, including epithelioid histiocytes, plasma cells, lymphocytes, and eosinophils, but without multinucleated giant cells. (Hematoxylin & eosin,  $\times 200$ .)

Laboratory studies disclosed elevation of ESR (115 mm/hr) and serum CRP (10.6 mg/dL). The following tests were all within normal limits: complete blood count; urinalysis; routine stool cultures; hepatic and renal function tests; PT; aPTT; and serum levels of IgG, IgA, IgM, anti-thrombin III, protein C, and protein S. ANA, anti-dsDNA, and IgG aCL antibody were negative.

After diagnosis, because both temporal arteries were involved, the patient was treated with prednisolone 15 mg/day plus azathioprine 50 mg every other day. Three weeks later, the hard and engorged temporal arteries had become less tender, and headache and mouth claudication had improved; ESR had dropped from 115 mm/hour to 13 mm/hour, and CRP from 10.6 mg/dL to 1.29 mg/dL. Thus, prednisolone was tapered to 5 mg/day and azathioprine was discontinued.

## Discussion

In this report, both patients had temporal arteritis after 50 years of age. New-onset headache was the defining feature in both cases, and ESR values were high ( $\geq 50$  mm/hr). Temporal artery biopsies disclosed the characteristic findings of temporal arteritis, and these findings satisfied the 1990 American College of Rheumatology classification criteria for temporal arteritis.<sup>5</sup> Although temporal arteritis and polymyalgia rheumatica (PMR) can occur simultaneously,<sup>1</sup> our patients did not complain of pain or stiffness in the neck, shoulder, or pelvic areas. The coexistence of PMR with temporal arteritis was therefore unlikely in our cases.

Interestingly, aCL antibodies are found in many autoimmune diseases, including systemic lupus erythematosus, Sjögren's syndrome, rheumatoid arthritis, polymyositis, and dermatomyositis.<sup>6</sup> The incidence of aCL antibodies in temporal arteritis is not low: about 31–46% of patients with this disorder test positive for such antibodies.<sup>4,7</sup> However, the role of aCL antibodies in temporal arteritis remains to be elucidated. In the study by Manna et al,<sup>8</sup> aCL antibodies did not appear to be an important factor in the development of vascular complications such as ischemic stroke, transient ischemic attack, angina pectoris, myocardial infarction, occlusive ocular vascular disease, Raynaud's phenomenon, and jaw claudication. Conversely, in a prospective, longitudinal study conducted by Chakravarty et al,<sup>9</sup> 3 of 5 patients with temporal arteritis and high levels of aCL antibodies at presentation progressed to severe vascular complications, suggesting that elevated aCL levels may indicate a poor prognosis regarding such complications. One of our 2 patients was positive for aCL-IgG, but neither patient had involvement of large vessels other than the temporal artery. Overall, more studies are needed to elucidate the relationship between aCL antibodies and the severity of temporal arteritis and extent of vessel involvement.

Although multinucleated giant cells are characteristic pathologic findings in temporal arteritis, such cells are not essential to the diagnosis, because 2 distinct pathologic types of the disorder have been proposed: a granulomatous multinucleated giant cell type; and a panarteritis infiltrated with mononuclear

cells, neutrophils, and eosinophils, but without multinucleated giant cells. The patient in our first case had granulomatous giant cell temporal arteritis, whereas our second patient had panarteritis.

Temporal arteritis has rarely been reported in Taiwan. Since the common clinical manifestations of the disorder are usually nonspecific, careful physical examination of the temporal arteries is particularly important for making an early diagnosis. Further studies are needed to determine the relationships between the clinical manifestations of temporal arteritis, aCL antibodies, and the different pathologic subtypes of the condition.

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