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【 CASE REPORT 】

Giant Cell Arteritis Presenting with Ptosis and Diplopia

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Abstract:
Giant cell arteritis (GCA) is vasculitis of large-sized vessels that can lead to vision loss. We herein report a rare case of GCA accompanied by ptosis and diplopia as early symptoms, which were caused by third nerve palsy. A 78-year-old man presented with fever, right temporal headache, right eyelid ptosis, and diplopia. GCA was confirmed by a temporal artery biopsy. The symptoms disappeared after a slight delay following the administration of prednisolone. Unlike vision loss, ptosis and diplopia are considered to be reversible and responsive to treatment. GCA should not be ruled out if patients exhibit these ophthalmic symptoms.

Key words: giant cell arteritis, eye involvements, large-vessel vasculitis

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Introduction

Giant cell arteritis (GCA) is a common type of systemic vasculitis that usually affects older people (>50 years old). GCA is categorized as vasculitis of large-sized vessels (1). Common symptoms include new-onset headache, fever, fatigue, weight loss, and jaw claudication. Visual symptoms may develop because of ischemia of the visual pathways (12-70%) (2) and visual loss is a well-known visual symptom. In addition, there are other ophthalmic manifestations such as amaurosis fugax, diplopia, ptosis, and eye pain (2). Diplopia is reported by from 1-19% of patients with GCA (2). The most common cause of diplopia is sixth nerve palsy, whereas third and fourth nerve palsies are rare causes (2-5). Concomitant ptosis and diplopia in GCA are both rare (6-8). We herein report the rare case of a GCA patient who developed diplopia and ptosis as early symptoms.

Case Report

A 78-year-old Japanese man presented with fever, right temporal headache, ptosis of the upper right eyelid, and diplopia 2 weeks before admission. These symptoms developed within the same day. He did not have any vision loss. Prior to these symptoms, he was healthy and not taking any medications. The symptoms progressively worsened, and physical examination revealed ptosis of the right eyelid and impairment of right eye movement, including adduction palsy, supraduction palsy, and infraduction palsy (Fig. 1A, B, D). Abduction was intact (Fig. 1C). His pupils were round at 3 mm and reactive to light, and other physical examinations were normal. He did not have myalgia or arthralgia and there was no complication of polymyalgia rheumatica. The C-reactive protein (CRP) concentration was 69.5 mg per liter (normal range, 0 to 2) and the erythrocyte sedimentation rate (ESR) was 56.1 mm per hour (normal range, 0 to 10). There were no other significant abnormalities on blood testing. Contrast-enhanced computerized tomography (CT) revealed enhancement of the walls of the bilateral temporal and vertebral arteries, and thickening of walls of the left subclavian artery and aortic arch (Fig. 2). Fluorodeoxyglucose positron emission tomography (FDG-PET-CT) demonstrated a FDG uptake in the right temporal artery, bilateral vertebral arteries, and aorta, especially in the aortic arch (Fig. 3). No abnormalities were observed on brain MRI. The diagnosis was confirmed by a biopsy of the right temporal artery, which revealed a narrowed lumen and granulomas with giant cells (Fig. 4). He was treated from 2 weeks after the development of ptosis and diplopia by 1-g intravenous methylprednisolone for three days followed by high-dose prednisolone (1 mg/kg of weight). At 2 days after administration, the headache and fever resolved promptly, and CRP normalized. Ptosis and diplopia improved 3 weeks

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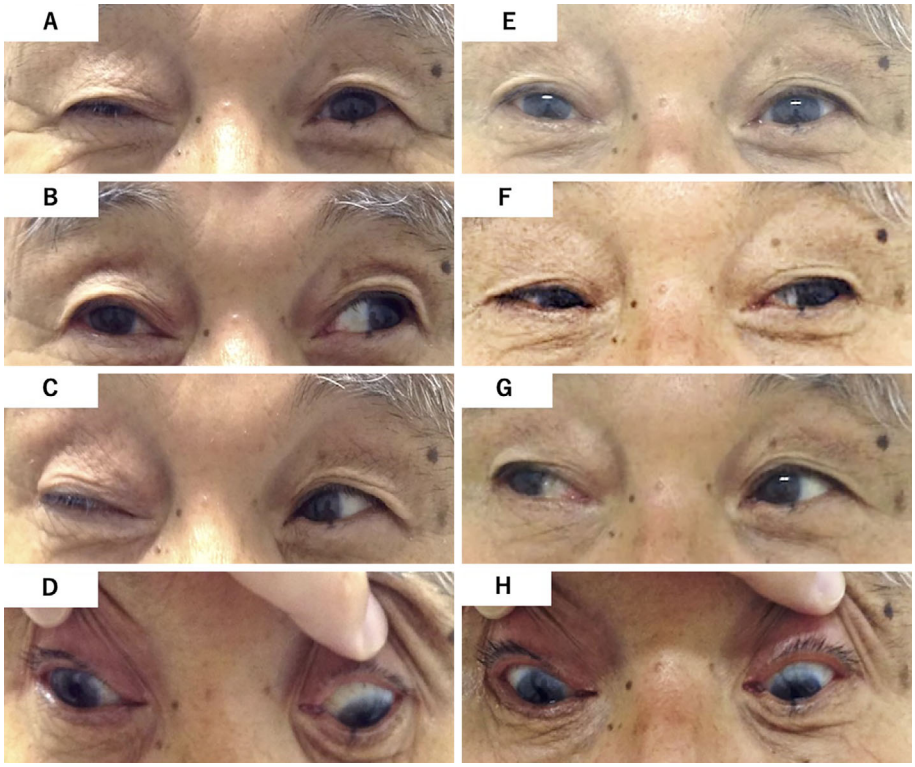

Figure 1. **Oculomotor findings before and after treatment. A-D: Ptosis of the right eyelid (A) and impairment of right eye movement; adduction palsy (B) and infraduction palsy (D). Abduction was intact (C). E-H: Ptosis and diplopia improved 3 weeks after treatment. Photos E, F, G, and H correspond to A, B, C, and D, respectively.**

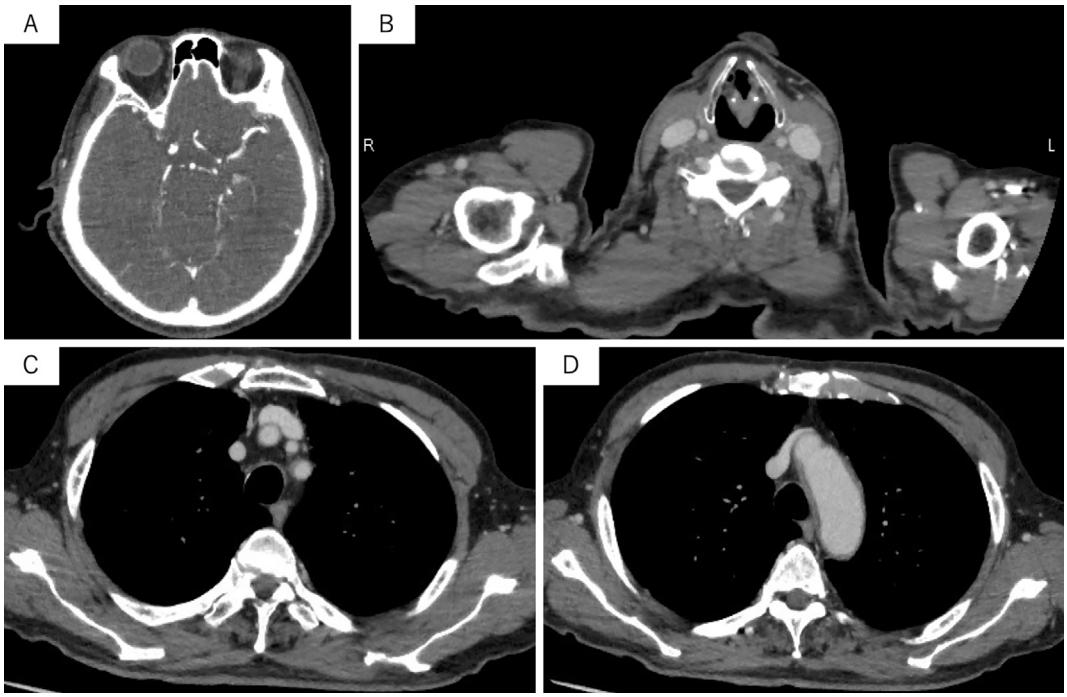

Figure 2. **Contrast-enhanced CT scan. Enhancement of the walls of the bilateral temporal (A) and vertebral arteries (B), and thickening of walls of the left subclavian artery (C) and aortic arch (D) were observed.**

after the initiation of therapy (5 weeks after onset) (Fig. 1E-H).

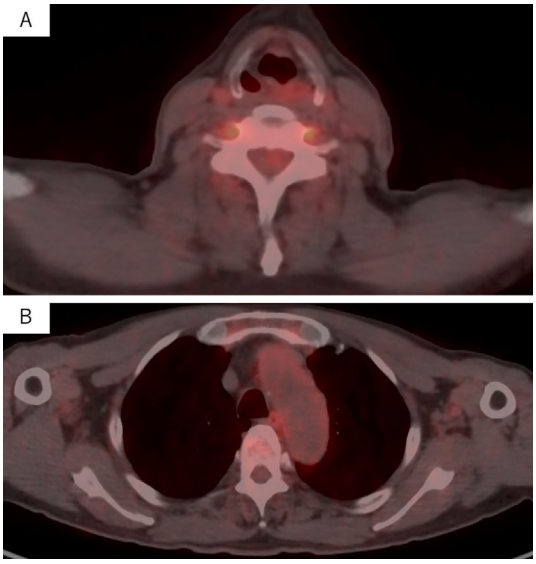

Figure 3. **FDG-PET-CT scan. A FDG uptake in the bilateral vertebral arteries (A) and aorta (B) was observed.**

Discussion

In this case, no evidence of brain infarctions, especially brainstem infarctions, was found on MRI of the head. Therefore, ischemia of the vasa nervorum around the third nerve was considered to be the cause. Thalamoperforating arteries (TPAs), which are branches of posterior cerebral arteries (PCAs), supply blood to some parts of the third nerve. PCAs are also branches of vertebral arteries, which were damaged according to CT and PET-CT. As TPA injury and occlusion cause oculomotor nerve paresis (9), TPAs may have been the cause in this case.

This patient did not have mydriasis even though ptosis and diplopia concomitantly developed. This is because pupillary fibers are peripherally located and receive more collateral blood supply than the main central nerve trunk (10). As a result, in third nerve palsy caused by compressive lesions, such as aneurism or mass, patients develop papillary involvement early. On the other hand, in third nerve palsy caused by microvascular ischemia, such as arteritis or diabetes mellitus, the pupils are spared. Thus, pupil-sparing is helpful for diagnosing the cause of third nerve palsy.

Matthew et al. reviewed four GCA patients with third

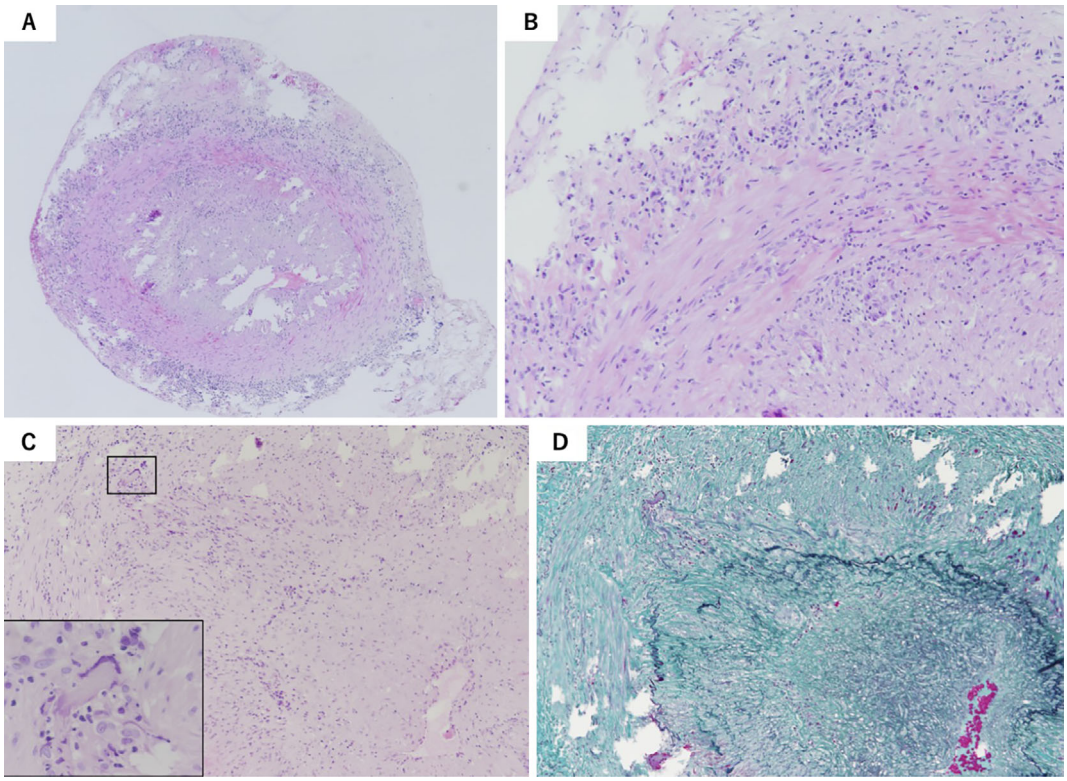

Figure 4. **Histopathological findings. Transverse sections of the right temporal artery specimen. A, B: Adventitial to medial inflammation with lymphocytes, plasma cells, and macrophages in a granulomatous pattern. Vascular destruction was observed in the inner half of the media, consistent with giant cell arteritis. Intimal fibrosis and luminal stenosis were also present. Hematoxylin and Eosin (H&E) staining; magnification, ×40 (A) and ×400 (B). C, D: Fragmentation of the internal elastic lamina was seen on Elastica Masson staining and multinucleated giant cells were seen on H&E staining of the same area. 200× H&E staining and Elastica Masson staining, inset: ×400.**

nerve palsy, and reported that all of them exhibited rapid improvement in symptoms and signs after starting high-dose oral prednisone, with complete recovery within 3 weeks even though three of them had a longer than one-week history of ophthalmic symptoms (8). We started therapy 2 weeks after the development of ptosis and diplopia, and these symptoms disappeared. Based on the above, ptosis and diplopia in GCA are responsive to treatment with a slight delay after starting treatment, unlike vision loss. When GCA patients develop vision loss, they may develop retinal ischemia in addition to arteritic ischemic neuropathy, which differs from ptosis and diplopia (2). This may be related to the irreversibility of vision loss because the retina is highly susceptible to ischemia and permanent vision loss may develop if ischemia is prolonged for more than approximately 100 minutes (11).

In this report, we describe a rare case of GCA with concomitant ptosis and diplopia caused mainly by third nerve palsy, and its therapeutic response. Although there are many reports of diplopia caused by sixth nerve palsy, ptosis and diplopia should be recognized as ophthalmic manifestations of GCA, and GCA should not be ruled out if patients exhibit these ophthalmic symptoms.

The authors state that they have no Conflict of Interest (COI).

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