BILATERAL VERTEBRAL ARTERY OCCLUSION WITHOUT HEADACHE IN GIANT CELL ARTERITIS

Giant cell arteritis (GCA) is well-recognized as an important cause of headache in the elderly.1 It is also an important element of the differential diagnosis for patients with transient or permanent visual loss. Delay in diagnosis of GCA can lead to blindness. Therefore, clinicians should be alert to atypical presentations in which headache is either not prominent or absent. We present a patient with GCA who had a cerebellar stroke and severe posterior circulation occlusive disease without headache.

Case presentation. A 76-year-old man with history of hypertension and sleep apnea presented with a prolonged episode of gait instability. He had experienced an episode of double vision 1 year previously for which he did not seek medical attention. He also mentioned several episodes of gait instability lasting less than 5 minutes in the few months before admission. He denied headache, jaw claudication, facial droop, numbness, or speech problems. On physical examination, he had a broad-based gait, mild dysmetria in the right upper and lower extremities, and nystagmus. Other aspects of the neurologic examination were unremarkable. There was no temporal artery tenderness.

Brain MRI revealed an infarct in the right posterior inferior cerebellar artery distribution. Magnetic resonance angiography revealed poor visualization of both vertebral arteries and faint visualization of the basilar artery. The erythrocyte sedimentation rate (ESR) was 37 mm/h with C-reactive protein (CRP) of 49.9 mg/L. Cerebral angiography revealed extracranial bilateral vertebral artery occlusion 1–2 cm distal to the origin. Due to the vertebral artery occlusion and elevated CRP, a temporal biopsy was performed, which showed multinucleated giant cells, fragmentation of internal elastic lamina, and lymphocytic infiltrate consistent with diagnosis of GCA (figure, A and B).

The patient was placed on high-dose prednisone and was stroke-free during the ensuing 1 year of follow-up. There were no visual complaints during follow-up.

Discussion. The incidence of GCA is highest among whites in northern European populations (20 cases per 100,000) and it has a much lower incidence among populations of Asian or African descent.2 It usually involves large- and medium-sized arteries with predominance in the aorta and the supra-aortic vessels. In one study of 204 patients, the cumulative rate of any large vessel manifestation (stenosis, aneurysm) was 24.9% within 10 years of diagnosis.3 Although headache and visual disturbances are well-known as presenting symptoms of GCA, stroke can also be the initial clinical manifestation.⁴ Our patient did not have prominent systemic symptoms and his ESR was normal for his age. In a study of 177

Figure

Temporal artery biopsy specimen

(A) Temporal artery biopsy specimen shows fragmentation of internal elastic lamina, 100× magnification. (B) Biopsy specimen shows lymphocytic infiltrate. Hematoxylin & eosin stain, 400× magnification.
patients with confirmed GCA, the sensitivity of an elevated ESR was 84% and that of an elevated CRP was 86%. In this study, in 4% of patients with pathologically confirmed GCA, both laboratory tests were normal. Patients with less prominent systemic symptoms, as in our case, are more likely to have normal ESR or CRP values.5

Our patient would not have met the criteria for GCA espoused by the American Academy of Rheumatology due to the lack of headache, temporal artery tenderness, and ESR <50 mm/h.6 Therefore, it is important for clinicians to have a heightened index of suspicion for patients with an atypical GCA presentation due to the risk of disease progression without steroid treatment.

GCA can present in an atypical fashion (without headache, visual symptoms, or jaw claudication). Even in the presence of conventional risk factors, diagnosis of GCA should be strongly considered in patients presenting with posterior circulation stroke and extracranial vertebral artery stenosis or occlusion.

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