Giant cell arteritis presenting as occipital neuralgia

Sirs,

Headache and jaw claudication are among the most commonly reported manifestations of giant cell arteritis (GCA) (1,2). Other cranial manifestations such as carotidynia, and facial pain have previously been observed but are uncommon (3, 4). Jundt and Mock also described occipital pain as the presenting manifestation of GCA in 8 of their 46 patients (5). In their report, however, these authors did not clearly describe whether occipital pain was the only presenting symptom or just a remarkable manifestation, often associated with other typical cranial manifestations of the disease at presentation in all these cases. Indeed, although occipital pain has long time been reported as a cranial manifestation of GCA, neither its actual frequency in large series nor its specificity as the only presenting sign of GCA have been determined so far. For this reason, we retrospectively reviewed the frequency of occipital pain in a large series of more than 200 biopsy-proven GCA patients seen over the last 20 years at the single hospital for a well-defined population in Lugo, Northwest Spain (6).

Of note, occipital pain was a relatively common complain among patients from Lugo, as it was reported by almost 25% of the patients. This symptom was observed more commonly at the onset of the disease and it was also described by the patients at the time of admission. However, occipital neuralgia as an excruciating pain preceding for several days the onset of other cranial symptoms of the disease was only observed in a single case. We briefly describe this case here.

An 81-year-old woman presented at the out-patient clinic because of occipital pain. At the age of 72, she had been diagnosed with polymyalgia rheumatica (PMR) because of a 2-month history of aching, pain and stiffness involving the neck, shoulders, and hip girdles, morning stiffness lasting more than 1 hr and an erythrocyte sedimentation rate (ESR) of 45 mm/lh. In that time no cranial symptoms of GCA were detected and for this reason a temporal artery biopsy was not considered. In addition, a rapid response to prednisone (15 mg/day) was obtained. Steroid treatment was tapered up to complete discontinuation 12 months later. Since then she had been asymptomatic and no relapses of PMR had occurred.

Nine years later, due to her past history of PMR,she was again referred to the rheumatology out-patient clinic for evaluation of occipital pain. At that time, the patient recalled a 2-week history of continuous burning occipital pain. She also had paresthesia in the occipital area, but no other symptoms such as jaw claudication, temporal headache, amaurosis fugax or PMR were registered. Physical examination only disclosed tenderness limited to the occipital area. No temporal tenderness was found. Due to the excruciating pain a bilateral occipital nerve block was then performed. However, only partial relief was obtained on the left side. Three days later, because of the persistence of occipital pain a right temporal artery biopsy was performed. The biopsy showed disruption of the internal elastic laminae with infiltration of mononuclear cells and the presence of giant cells in the arterial wall. In addition, routine laboratory analysis yielded an ESR of 120 mm/hr. Following steroid therapy (40 mg/day prednisone) a rapid and dramatic improvement of symptoms was achieved. No new episodes of occipital neuralgia have occurred since then (3 months' follow-up).

This case may be a good example of clinical involvement of the occipital artery in GCA. Symptoms were more severe in the occipital region and no clear abnormality of the temporal arteries was noticed on examination. Occipital neuralgia is described as a continuous aching or burning occipital pain which may radiate to the frontal region. As in our case, localized tenderness of the nerve is felt in most cases (7). The pain is caused by injury to the occipital nerve along its course from the C2 dorsal root (8). The occipital artery lies in close proximity to the nerve and, due to this,in patients with GCA the occipital pain is likely to be related to inflammation of the occipital artery.

Biopsy of the occipital artery is technically more difficult than biopsy of the temporal artery. In our case, as suggested by Achkar and colleagues, who considered superficial temporal artery biopsy as the biopsy of choice in an elderly patient with high suspicion of GCA, a biopsy of the right superficial temporal artery was performed (9). Despite clinical findings in the temporal area being negligible, a pathologic diagnosis of GCA was confirmed.

With this case we would like to further emphasize the clinical importance of the occipital artery in GCA and reiterate the need for high physician awareness of GCA in elderly patients presenting with occipital neuralgia.

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