Occipital Artery Vasculitis Not Identified as a Mechanism of Occipital Neuralgia–Related Chronic Migraine Headaches

Ivica Ducic, M.D., Ph.D.
John M. Felder III, M.D.
Jeffrey E. Janis, M.D.

Washington, D.C.; and Dallas, Texas

Background: Recent evidence has shown that some cases of occipital neuralgia are attributable to musculofascial compression of the greater occipital nerve and improve with neurolysis. A mechanical interaction at the intersection of the nerve and the occipital artery may also be capable of producing neuralgia, although that mechanism remains one theoretical possibility among several. The authors evaluated the possibility of unrecognized vasculitis of the occipital artery as a potential mechanism of occipital neuralgia arising from the occipital artery/greater occipital nerve junction.

Methods: Twenty-five patients with preoperatively documented bilateral occipital neuralgia–related chronic headaches underwent peripheral nerve surgery with decompression of the greater occipital nerve bilaterally, including the area of its intersection with the occipital artery. In 15 patients, a 2-cm segment of the occipital artery was excised and submitted for pathologic evaluation. All patients were evaluated intraoperatively for evidence of arterially mediated greater occipital nerve compression, and the configuration of the nerve-vessel intersection was noted.

Results: None of the 15 specimens submitted for pathologic evaluation showed vasculitis. Intraoperatively, all 50 sites examined showed an intimate physical association between the occipital artery and greater occipital nerve.

Conclusions: Surgical specimens from this first in vivo study provided no histologic evidence of vasculitis as a cause of greater occipital nerve irritation at the occipital artery/greater occipital nerve junction in patients with chronic headaches caused by occipital neuralgia. Based on these findings, mechanical (and not primary inflammatory) irritation of the nerve by the occipital artery remains an important theoretical cause for otherwise idiopathic cases. The authors have adopted an operative technique that includes physical separation of the nerve-artery intersection (in addition to musculofascial neurolysis) for a more thorough surgical treatment of occipital neuralgia. (*Plast. Reconstr. Surg.* 128: 908, 2011.)

CLINICAL QUESTION/LEVEL OF EVIDENCE: Therapeutic, IV.

Occipital neuralgia is a headache syndrome characterized by paroxysmal or continuous lancinating pain in the posterior scalp (in the dermatomes of the greater, lesser, and/or dorsal occipital nerves), which may or may not include an element of frontal spread. Although not as common as other headache types, it has been historically underrecognized as a cause of frequent, often severe headaches. However, recent years have seen an improvement in the treatment for this condition because of a greater understanding of the anatomical basis for this nerve-related pain. Many occipital "migraines" and other headache types previously thought to be central in origin are now beginning to be understood as neuralgic pain caused by a demonstrable irritation of the occipital nerves (as defined by localized tenderness and response to anesthetic blocks or botulinum toxin) at defined anatomical locations. Causes that have

From the Department of Plastic Surgery, Peripheral Nerve Surgery Institute, Georgetown University Hospital, and the Department of Plastic Surgery, University of Texas Southwestern Medical Center.
Received for publication November 14, 2010; accepted January 21, 2011.

DOI: 10.1097/PRS.0b013e3182174229

Disclosure: *The authors have no financial interest to declare in relation to the content of this article.*

been so far reported in the literature include proximal irritation of the C1/C2 nerve roots by bony arthrosis, intracranial arteriovenous malformations or aberrant vasculature, perineural tumors, occipital involvement of temporal arteritis, and others.[1–5] However, the majority of cases of medically refractory occipital neuralgia appear to be attributable to a compression mechanism involving the more distal greater occipital nerve proper as it courses through a variety of tight musculofascial "tunnels" in its course from its deep origin toward the superficial scalp,[6] and surgical decompression of these tunnels has been shown to relieve the pain of occipital neuralgia.[7,8] Recent cadaveric studies by Shimizu and colleagues have postulated that the point of intersection between the greater occipital nerve and the occipital artery (Fig. 1) may be an additional generator of occipital neuralgia in otherwise idiopathic cases,[9] by mechanical compression of the artery on the nerve; however, to date there has been no study in patients with diagnosed occipital neuralgia to provide evidence for this hypothesis or to rule out other possible mechanisms.

Separately, it is well known that the prevalence of headaches is significantly increased among patients with vasculitic disorders such as lupus and Behçet syndrome[10–16] and with disorders such as Raynaud phenomenon,[17–23] which are themselves often associated with the presence of vasculitis. There have, in fact, been documented cases of temporal arteritis producing occipital neuralgia by means of occipital artery involvement.[1,24,25] However, to this point, there has been no discussion in the literature of whether otherwise idiopathic cases of occipital neuralgia may actually be explained by vasculitis of the occipital artery rather than, or in addition to, a mechanical nerve-artery compression mechanism. Given the intimate anatomical association of the occipital artery and greater occipital nerve and the high prevalence of headaches among patients with vasculitides, the present study was undertaken in an attempt to seek evidence either for or against the possibility that occipital artery vasculitis is an unrecognized cause of occipital neuralgia arising from the occipital artery/greater occipital nerve intersection.

PATIENTS AND METHODS

Twenty-five surgical candidates were diagnosed preoperatively with bilateral occipital neuralgia and selected for surgical treatment according to criteria and algorithms published previously.[8] Greater occipital nerve decompression surgery was performed by the senior author (I.D.) according to a previously published technique[8] that routinely includes inspection of the occipital artery/greater occipital nerve intersection and separation of the nerve and artery, with segmental excision and ligation of the occipital vessels in instances where they are observed to be compressing the greater occipital nerve. In all 25 cases, bilateral greater occipital nerve decompression included the site of the occipital artery/greater occipital nerve intersection (Fig. 1), and the anatomy of the junction was noted for reference. In the first 15 of these 25 patients, a 2-cm segment of the occipital artery at its junction with the greater occipital nerve (1 cm on either side of the junction) was submitted for pathologic evaluation rather than being discarded

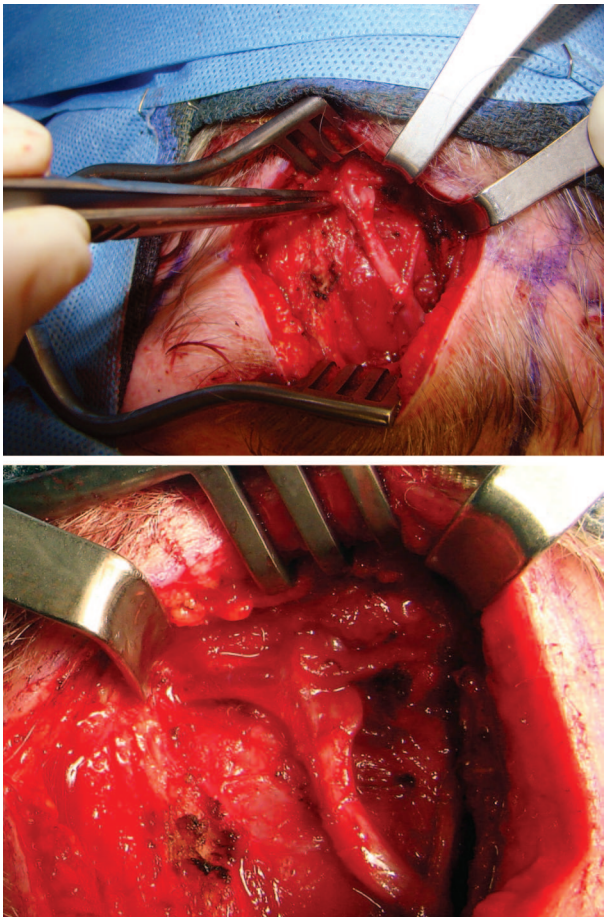

Fig. 1. (*Above*) Intraoperative view of the intersection of the occipital artery and greater occipital nerve. A forceps holds the artery; one branch goes behind the nerve. The other branch splits the nerve and continues in front of it. (*Below*) Detail of the occipital artery/greater occipital nerve intersection displaying an intimate anatomical relationship. The artery travels transversely between the two small retractors, piercing the nerve. The nerve is oriented superoinferiorly.

in the usual fashion. Specimens were fixed in formalin, serially sectioned, chemically stained, and reviewed by a senior staff pathologist specifically for the presence of vasculitis. After results for 15 cases were received, data were felt to be conclusive and the submission of specimens was terminated to avoid unnecessary expenditure.

RESULTS

None of the 15 specimens submitted for pathologic evaluation showed evidence of vasculitis (Fig. 2). As ancillary data, the nerve-artery anatomy of the biopsy site was noted in each case. All 25 patients showed an intersection point between the occipital artery and the greater occipital nerve. Of 50 sites examined, 47 had the nerve anterior and three posterior to the occipital artery. The path of the occipital vein matched that of the artery in all but five sites, where one branch coursed anterior and the other branch posterior to the nerve. Seven sites were noted to have small arterial branches, at the intersection or immediately distal to it, piercing the greater occipital nerve.

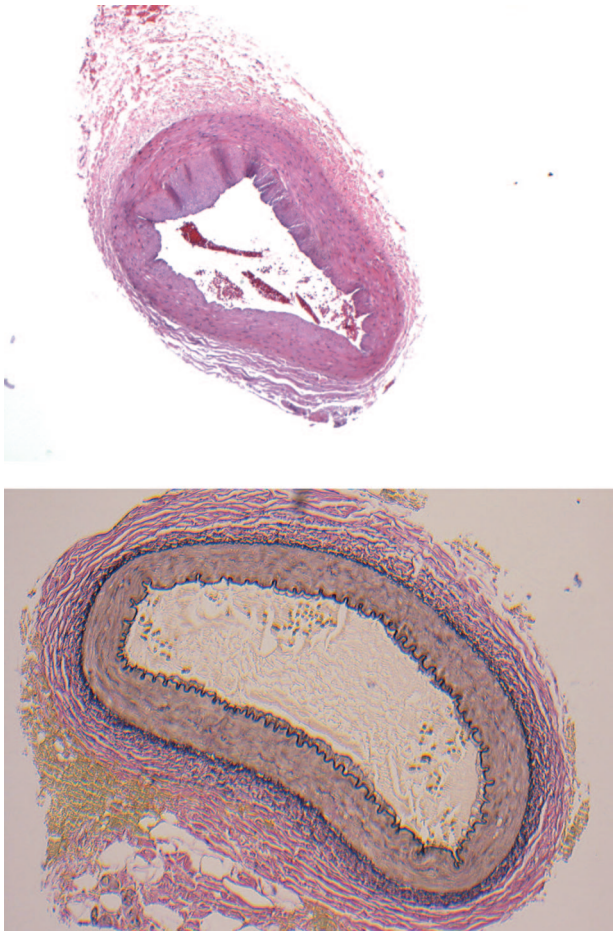

Fig. 2. Representative pathologic slides of the occipital artery, excised at its junction with the greater occipital nerve. The artery demonstrates normal appearance and architecture. The basement membrane is intact and there is an absence of leukocytic infiltration of the vessel wall. There is no histologic evidence of vasculitis. (*Above*) Hematoxylin and eosin stain. (*Below*) Elastin stain; original magnification, ×40.

DISCUSSION

It has been made clear through anatomical,[6,26–28] dynamic,[29] and clinical studies[7,30–32] that musculofascial structures can produce compression of the occipital nerve, leading to the clinical syndrome of occipital neuralgia, and that decompression of these structures may reverse the syndrome. What has been previously proposed on an anatomical basis[9,27,33] but remains somewhat less clinically clear is whether occipital artery mechanical compression of the greater occipital nerve (e.g., by vasodilation or pulsation) may be the cause of occipital neuralgia in otherwise idiopathic cases. Although pure mechanical compression of the greater occipital nerve by the occipital artery is an intuitive theory, the possibility must also be considered that the intimately associated nerve and artery may interact pathologically by means of different mechanisms.

Of particular interest along this train of thought would be unrecognized occipital artery vasculitis contributing to greater occipital nerve irritation. Vasculitis makes sense as a proposed pathophysiologic mechanism not only because of the close proximity of the occipital artery to the greater occipital nerve, but because of the known potential for inflammatory vasculitides such as temporal arteritis to spread on occasion to involve the occipital artery, producing occipital neuralgia.[1,24,25] Furthermore, the frequency of chronic headaches in populations with inflammatory vasculitides such as systemic lupus erythematosus and Behçet syndrome is empirically noted to be increased significantly above that of the normal population.[10–16,34] The association between migraine headaches and Raynaud phenomenon, itself a frequent corollary of vasculitic disease, is also well known.[19–23,35]

The aim of this study was to search for evidence either in support of or against the possibility that vasculitis might be an unrecognized cause of occipital neuralgia emanating from the occipital artery/greater occipital nerve junction. Importantly, this is the first study to examine tissue from living patients with documented occipital neuralgia for evidence of vasculitis. Although limited by sample size and the necessity (imposed by the involvement of living patients) to minimize the size of our biopsy specimen, this preliminary result

is a negative one in that no surgical specimens of the occipital artery from our series showed evidence of vasculitis. The significance of this negative result is appreciated when viewed in context of the efforts of Shimizu and colleagues,[9] who also questioned whether interaction of the nerve and artery at their intersection contributes to the clinical picture of occipital neuralgia and in cadaveric studies and were able to demonstrate a consistent crossover point between nerve and artery with an indentation of the nerve in the region of contact. Their findings lead to the logical proposal that mechanical impingement of nerve by artery is the likely mechanism of a pathologic interaction between the two, but do not account for the possible alternative mechanism of vasculitis at the junction. Our new data encourage further investigation into the premise of a mechanically produced and surgically treatable mechanism by effectively ruling out the possibility of a vasculitic mechanism (which presumably would be best treated pharmacologically) in the majority of cases. This mechanism for occipital neuralgia would be consistent with the already clinically accepted paradigm of vascular compression leading to neuralgic pain in other similar disorders, such as trigeminal neuralgia, where microvascular decompression is a proven modality for relieving pain and reversing disease features.[36]

This small study was designed specifically to evaluate the hypothesis that vasculitis may be a generator of neuralgia at the occipital artery/greater occipital nerve junction, where a mechanical interaction (proposed by other investigators) was the null hypothesis. It is not an outcomes study meant to evaluate the effectiveness of mechanical decompression of the junction. As such, our conclusions from this work about the actual nature of any pathologic interaction between the occipital artery and greater occipital nerve are a reversion to the null hypothesis, made in light of our negative results. The proposed mechanical compression mechanism at the occipital artery/greater occipital nerve junction therefore remains theoretical but gains theoretical weight by the elimination of a competing hypothesis. The possibility of a mechanical compression mechanism at the occipital artery/greater occipital nerve junction makes understanding and appreciation of anatomical variations of the occipital artery and possible compression points of the greater occipital nerve[27] important for peripheral nerve surgeons and suggests that comprehensive surgical decompression of the occipital nerve should include intraoperative evaluation and separation of the nerve-artery intersection site within the trapezial tunnel.

CONCLUSIONS

Pathologic analysis of occipital artery specimens from our patients with preoperatively diagnosed occipital neuralgia provided no histologic evidence for vasculitis as the direct cause of headaches. Anatomical observations of the occipital artery/greater occipital nerve intersection in our series show an intimate physical association in all cases and several variations in the structure of the junction, including instances where the nerve is pierced by an artery. Along with other reports,[8,9,33] our results suggest that mechanical (and not primary inflammatory) irritation of the greater occipital nerve by the occipital artery remains an important theoretical cause of otherwise idiopathic occipital neuralgia. Therefore, in patients intraoperatively found to have an intimate connection between nerve and artery, it is our practice[8] to use an operative technique that includes routine separation of the artery from the nerve (in addition to standard musculofascial neurolysis) to achieve the most thorough mechanical decompression possible in the surgical treatment of occipital neuralgia.

Ivica Ducic, M.D., Ph.D.
Department of Plastic Surgery
Peripheral Nerve Surgery Institute
Georgetown University Hospital
3800 Reservoir Road, NW
Washington, D.C. 20007
ducici@gunet.georgetown.edu

REFERENCES

1. Jundt JW, Mock D. Temporal arteritis with normal erythrocyte sedimentation rates presenting as occipital neuralgia. *Arthritis Rheum.* 1991;34:217–219.
2. Hildebrandt J, Jansen J. Vascular compression of the C2 and C3 roots: Yet another cause of chronic intermittent hemicrania? *Cephalalgia* 1984;4:167–170.
3. Ballesteros-Del Rio B, Ares-Luque A, Tejada-Garcia J, Muela-Molinero A. Occipital (Arnold) neuralgia secondary to greater occipital nerve schwannoma. *Headache* 2003;43:804–807.
4. Martin BC, Fagan PJ. The surgical therapy of certain occipital headaches. *Plast Reconstr Surg.* 1964;33:266–268.
5. Vanelderen P, Lataster A, Levy R, Mekhail N, van Kleef M, Van Zundert J. 8. Occipital neuralgia. *Pain Pract.* 2010;10:137–144.
6. Mosser SW, Guyuron B, Janis JE, Rohrich RJ. The anatomy of the greater occipital nerve: Implications for the etiology of migraine headaches. *Plast Reconstr Surg.* 2004;113:693–697; discussion 698–700.
7. Guyuron B, Reed D, Kriegler JS, Davis J, Pashmini N, Amini S. A placebo-controlled surgical trial of the treatment of migraine headaches. *Plast Reconstr Surg.* 2009;124:461–468.
8. Ducic I, Hartmann EC, Larson EE. Indications and outcomes for surgical treatment of patients with chronic migraine headaches caused by occipital neuralgia. *Plast Reconstr Surg.* 2009;123:1453–1461.
9. Shimizu S, Oka H, Osawa S, et al. Can proximity of the occipital artery to the greater occipital nerve act as a cause

of idiopathic greater occipital neuralgia? An anatomical and histological evaluation of the artery-nerve relationship. *Plast Reconstr Surg.* 2007;119:2029–2034; discussion 2035–2036.
10. Lessa B, Santana A, Lima I, Almeida JM, Santiago M. Prevalence and classification of headache in patients with systemic lupus erythematosus. *Clin Rheumatol.* 2006;25:850–853.
11. Saip S, Siva A, Altintas A, et al. Headache in Behçet's syndrome. *Headache* 2005;45:911–919.
12. Whitelaw DA, Hugo F, Spangenberg JJ, Rickman R. Headaches in patients with systemic lupus erythematosus: A comparative study. *Lupus* 2004;13:501–505.
13. Appenzeller S, Costallat LT. Clinical implications of migraine in systemic lupus erythematosus: Relation to cumulative organ damage. *Cephalalgia* 2004;24:1024–1030.
14. Sfikakis PP, Mitsikostas DD, Manoussakis MN, Foukaneli D, Moutsopoulos HM. Headache in systemic lupus erythematosus: A controlled study. *Br J Rheumatol.* 1998;37:300–303.
15. Pal B, Gibson C, Passmore J, Griffiths ID, Dick WC. A study of headaches and migraine in Sjögren's syndrome and other rheumatic disorders. *Ann Rheum Dis.* 1989;48:312–316.
16. Isenberg DA, Meyrick-Thomas D, Snaith ML, McKeran RO, Royston JP. A study of migraine in systemic lupus erythematosus. *Ann Rheum Dis.* 1982;41:30–32.
17. Annese V, Tomietto P, Venturini P, D'Agostini S, Ferraccioli G. Migraine in SLE: Role of antiphospholipid antibodies and Raynaud's phenomenon (in Italian). *Reumatismo* 2006;58: 50–58.
18. Voulgari PV, Alamanos Y, Papazisi D, Christou K, Papanikolaou C, Drosos AA. Prevalence of Raynaud's phenomenon in a healthy Greek population. *Ann Rheum Dis.* 2000;59:206–210.
19. Terwindt GM, Haan J, Ophoff RA, et al. Clinical and genetic analysis of a large Dutch family with autosomal dominant vascular retinopathy, migraine and Raynaud's phenomenon. *Brain* 1998;121:303–316.
20. O'Keeffe ST, Tsapatsaris NP, Beetham WP Jr. Association between Raynaud's phenomenon and migraine in a random population of hospital employees. *J Rheumatol.* 1993;20:1187–1188.
21. de Trafford JC, Lafferty K, Potter CE, Roberts VC, Cotton LT. An epidemiological survey of Raynaud's phenomenon. *Eur J Vasc Surg.* 1988;2:167–170.
22. Leppert J, Aberg H, Ringqvist I, Sörensson S. Raynaud's phenomenon in a female population: Prevalence and association with other conditions. *Angiology* 1987;38:871–877.
23. Zahavi I, Chagnac A, Hering R, Davidovich S, Kuritzky A. Prevalence of Raynaud's phenomenon in patients with migraine. *Arch Intern Med.* 1984;144:742–744.
24. González-Gay MA, García-Porrúa C, Brañas F, Alba-Losada J. Giant cell arteritis presenting as occipital neuralgia. *Clin Exp Rheumatol.* 2001;19:479.
25. Albornoz MA, Sharit S, Myers AR. Multi-vessel biopsy-proven giant cell arteritis involving the occipital and temporal arteries. *J Clin Rheumatol.* 1997;3:227–229.
26. Austad ED. The anatomy of the greater occipital nerve: Implications for the etiology of migraine headaches (Discussion). *Plast Reconstr Surg.* 2004;113:698–700.
27. Janis JE, Hatef DA, Ducic I, et al. The anatomy of the greater occipital nerve: Part II. Compression point topography. *Plast Reconstr Surg.* 2010;126:1563–1572.
28. Ducic I, Moriarty M, Al-Attar A. Anatomical variations of the occipital nerves: Implications for the treatment of chronic headaches. *Plast Reconstr Surg.* 2009;123:859–863; discussion 864.
29. Vital JM, Grenier F, Dautheribes M, Baspeyre H, Lavignolle B, Sénégas J. An anatomic and dynamic study of the greater occipital nerve (n. of Arnold): Applications to the treatment of Arnold's neuralgia. *Surg Radiol Anat.* 1989; 11:205–210.
30. Guyuron B, Kriegler JS, Davis J, Amini SB. Comprehensive surgical treatment of migraine headaches. *Plast Reconstr Surg.* 2005;115:1–9.
31. Gille O, Lavignolle B, Vital JM. Surgical treatment of greater occipital neuralgia by neurolysis of the greater occipital nerve and sectioning of the inferior oblique muscle. *Spine (Phila Pa 1976)* 2004;29:828–832.
32. Ducic I, Hartmann EC, Larson EE. Indications and outcomes for surgical treatment of patients with chronic migraine headaches caused by occipital neuralgia. *Plast Reconstr Surg.* 2009;123:1453–1461.
33. Janis JE, Hatef DA, Reece EM, McCluskey PD, Schaub TA, Guyuron B. Neurovascular compression of the greater occipital nerve: Implications for migraine headaches. *Plast Reconstr Surg.* 2010;126:1996–2001.
34. Kidd D. The prevalence of headache in Behçet's syndrome. *Rheumatology (Oxford)* 2006;45:621–623.
35. Geraud G, Fabre N, Soulages X, Bes A. Migraine and the idiopathic Raynaud phenomenon (in French). *Rev Neurol (Paris)* 1986;142:638–640.
36. McLaughlin MR, Jannetta PJ, Clyde BL, Subach BR, Comey CH, Resnick DK. Microvascular decompression of cranial nerves: Lessons learned after 4400 operations. *J Neurosurg.* 1999;90:1–8.