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CASE REPORT

When Occipital Artery Biopsy is Preferred to Temporal Biopsy for Giant Cell Arteritis: A Step-By-Step Description of the Surgical Technique

May Dvir, MS; Ghaith Almhanni, MD; Huzaif Qaisar, MBBS; Andrew Calvin, MD; and Tiziano Tallarita, MD

Abstract

Giant cell arteritis is an autoimmune disease that affects large and medium blood vessels of the head and neck. Its prompt treatment is mandatory to avoid severe and permanent complications, such as blindness. Temporal artery biopsy is an important part of the diagnostic work-up, especially in those patients with cranial symptoms or in the elderly with a fever of unknown origin. Most patients have signs and symptoms matching the distribution of their arterial involvement. In the case scenario of occipital headache or nuchal pain, a biopsy of the occipital artery may be preferred to a temporal artery biopsy. This article provides important anatomical details of the course of the occipital artery and explains, in a stepwise fashion, how to perform an occipital artery biopsy.

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Giant cell arteritis (GCA) is an autoimmune disease that affects large and medium blood vessels of the head and neck. It is the most common systemic vasculitis in Western countries in patients over the age of 50 years.[1] The typical presentation of GCA is scalp tenderness, headache, jaw claudication, fever, systemic myalgias, and elevated inflammatory markers (eg, erythrocyte sedimentation rate [ESR] or C-creative protein [CRP]). If left untreated, it can lead to irreversible vision loss from ischemia of the optic nerve and retina.[2] Strong suspicion for GCA is based on symptoms, laboratory findings (elevated ESR or CRP), sonographic evidence of thickening of the temporal or occipital artery (OA), and mandates prompt initiation of steroid treatment, even before the diagnosis is confirmed. Temporal artery biopsy is an important part of the diagnostic work-up, especially in those patients with cranial symptoms or in elderly with fever of unknown origin (FUO).[3] However, it is becoming clearer that most patients have signs and symptoms matching the distribution of their arterial involvement.[4,5] In the presence of occipital headache or nuchal pain or previous negative temporal biopsy with atypical symptoms, an OA biopsy can confirm the diagnosis of GCA.[6,7]

Although harvest of a long segment of the OA through a very extensive scalp incision has been largely described in the neurosurgical literature for an extra-intracranial bypass, the technique of how to biopsy a short segment of the OA through a small incision has not been elucidated.

This article aims to illustrate the anatomical course of the OA and describe how to perform an OA biopsy in the setting of GCA.

From the Department of Cardiovascular Surgery, Mayo Clinic Health System, Eau Claire, WI (M.D., G.A., T.T.); University Cattolica del Sacro Cuore, Rome, Italy (M.D.); Department of Rheumatology, Mayo Clinic Health System, LaCrosse, WI (H.Q.); and Department of Cardiology, Mayo Clinic Health System, Eau Claire WI (A.C.).

Occipital Artery Anatomy

The OA originates from the posterior or lateral branch of the external carotid artery[7] and it is divided into 3 parts: digastric, intermuscular (or transitional), and subcutaneous (Figure 1A).[8]

The digastric segment runs medial to the posterior belly of the digastric muscle and is crossed posteriorly by the accessory cranial nerve (XI), internal jugular, and hypoglossal nerve (XII). Toward the end, it is located medial to the emergence of the facial nerve (VII) from the stylomastoid foramen. The intermuscular segment runs from the mastoid groove to the superior nuchal line (SNL), after piercing the sternocleidomastoid and the splenium capitis muscles. The subcutaneous segment runs from the SNL to the end, where it ramificates in numerous side branches feeding the skin and the local muscles. The side branches also connect with those from the contralateral OA.[7-9] For its entire course, the subcutaneous OA stays above the galea aponeurosis in a dense subcutaneous tissue (Figure 1B).[8,9] Most of the time, its main trunk is 5 cm long from the SNL.[7] The subcutaneous segment is the most superficial of the 3 segments, away from important structures, and the easiest to surgically access. For these reasons, it should be considered the ideal target for biopsy.

Occipital Artery Biopsy Technique

The patient is placed supine on the operating table. After general anesthesia is induced and the airways secured, the patient is turned to a prone position. Both arms are tucked along the body. The surgical field is prepped and draped in a standard sterile fashion. A sterile gel is applied to keep hairs away from the surgical field, circumventing the need for hair clip. Ultrasound guidance is used to locate the subcutaneous segment of the OA, which is marked on the skin, in preparation for the incision (Figure 2A-C). Of note, the subcutaneous segment of the OA is always located above the SNL. A 5 cm incision is made with a 15-blade scalpel. The incision is deepened down with sharp scissors dissection and bipolar electrocautery, as needed. The OA is in the connective tissue, just above the galea aponeurotica. The OA runs together with the occipital veins. The OA is identified and dissected free from the surrounding tissue for about 3 cm (Figure 3). When in doubt, a Doppler probe is used to distinguish the OA from the occipital vein. Side branches of the occipital nerve can be encountered and divided with impunity to improve vessel exposure. The artery is, then, ligated proximally and distally with 3-0 silk ties and divided. Because of the small caliber of the OA, it is sometimes difficult to insonate the vessel with the Doppler probe. In case the operator is still questioning the nature of the vessel, both artery and vein should be sent for

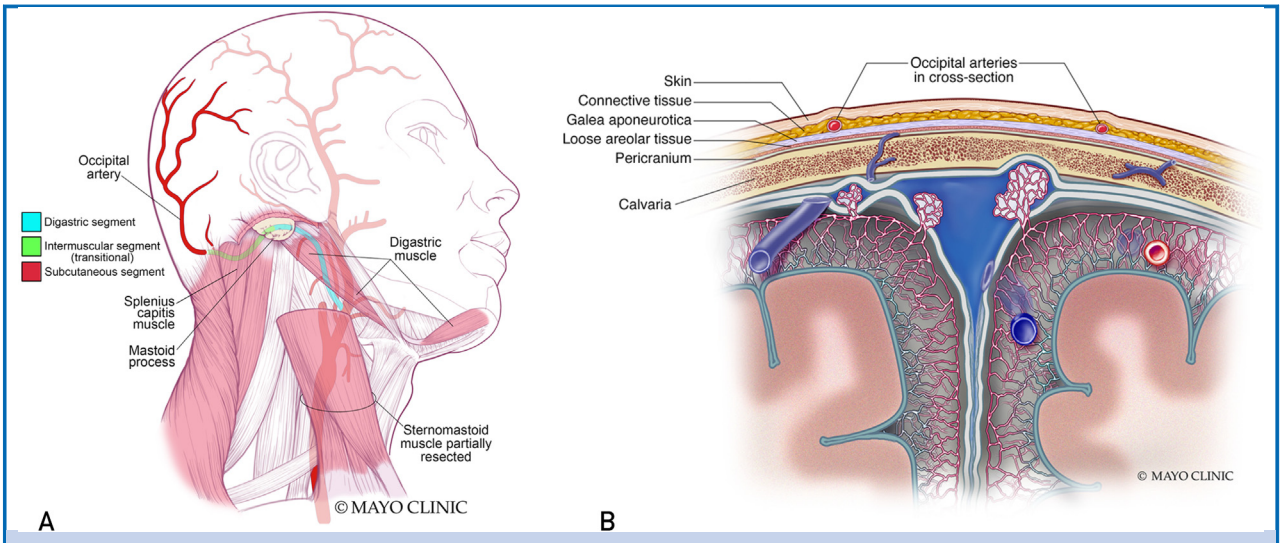

FIGURE 1. (A) The occipital artery is one of the 8 branches of the external carotid artery. Its course is divided into 3 main segments: digastric, intermuscular (or suboccipital), and subcutaneous, highlighting the nearby muscles. (B) Cross-sectional view of the occipital scalp layers.

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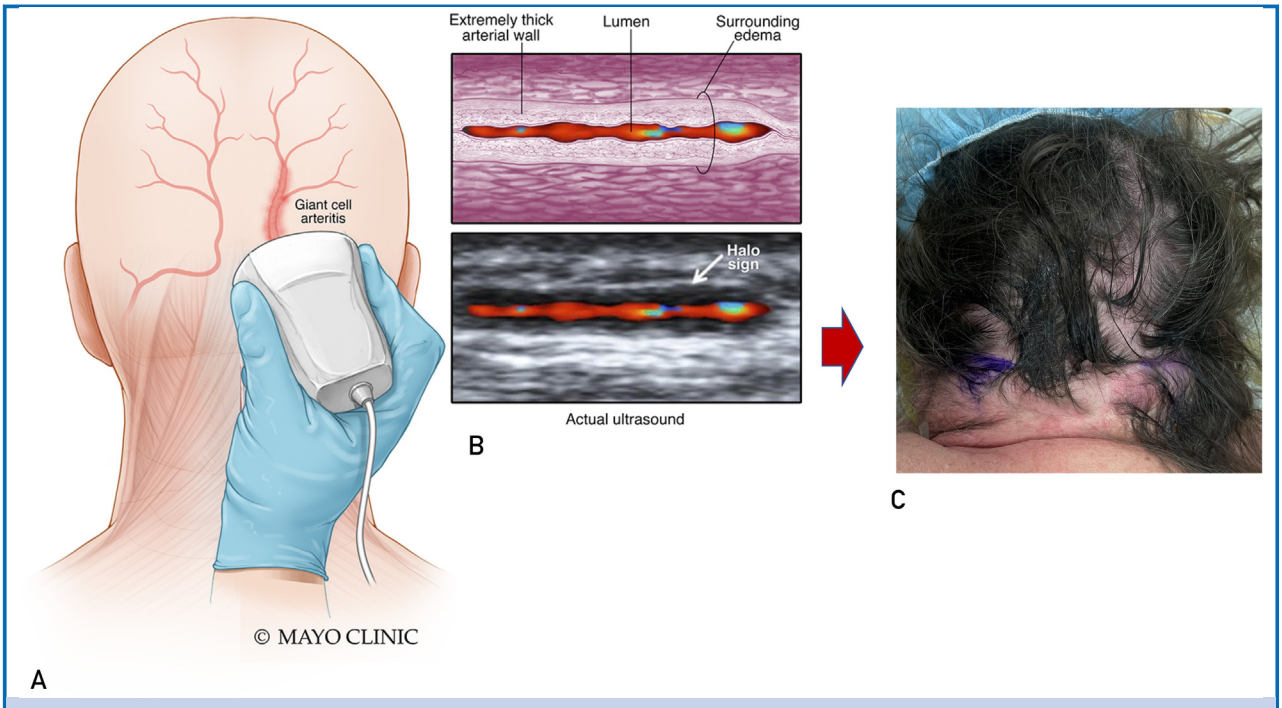

FIGURE 2. Identification the occipital artery by ultrasound (US) guidance (A), showing the characteristic halo sign, which is an echogenic ultrasonic signal of the thickened arterial wall and surrounding inflammatory edema (B). The surgical incision is marked on the patient's skin (C).

pathological analysis. Because GCA presents with typical inflammatory skip lesions, harvesting of at least 2 cm of arterial graft is recommended to decrease the chance of false negative pathological results.

The specimen is sent for both frozen and permanent analysis. If the frozen section comes back positive for GCA, the contralateral OA is not biopsied. Otherwise, the same procedure is performed on the contralateral OA. At the end of the procedure, hemostasis is checked and optimized. The wound is irrigated and closed with interrupted 3-0 Vicryl subdermal sutures, followed by 4-0 Monocryl subcuticular sutures. Ethicon Dermabond is applied on the incision.

DISCUSSION

Giant cell arteritis is not always straightforward to diagnose, and temporal artery biopsy has historically been a key part of the diagnostic work-up for GCA. However, its sensitivity ranges between 49% and 95%[10] and can be influenced by skip lesions,[11] glucocorticoid treatment (although this particular factor may be overstated)[12] or sparing of the temporal arteries by GCA. Atypical presentation, such as isolated occipital pain, FUO, and normal inflammatory markers, is present in 17% of patients with GCA,[13] and 66%-69% of these have OA involvement.[9] The presence of isolated FUO and high indices of inflammation in patients over the age of 50, can suggest GCA even in the absence of characteristic clinical signs and symptoms.[14] Positron emission tomography can be a valuable aid in the diagnosis of atypical cases, but only biopsy allows a definitive diagnosis.[14] In the case of FUO and negative temporal artery biopsy, OA biopsy could represent a novel option for possible definitive diagnosis.

More commonly in the elderly, polymyalgia rheumatica can be associated with GCA, which notoriously can cause neck stiffness or pain.[15] In fact, one-third of isolated patients with polymyalgia rheumatica have vascular uptake in the positron emission tomography scans, suggesting a clinically unrecognized, hidden GCA.[15] An OA biopsy could diagnose GCA in patients with occipital headache and negative temporal

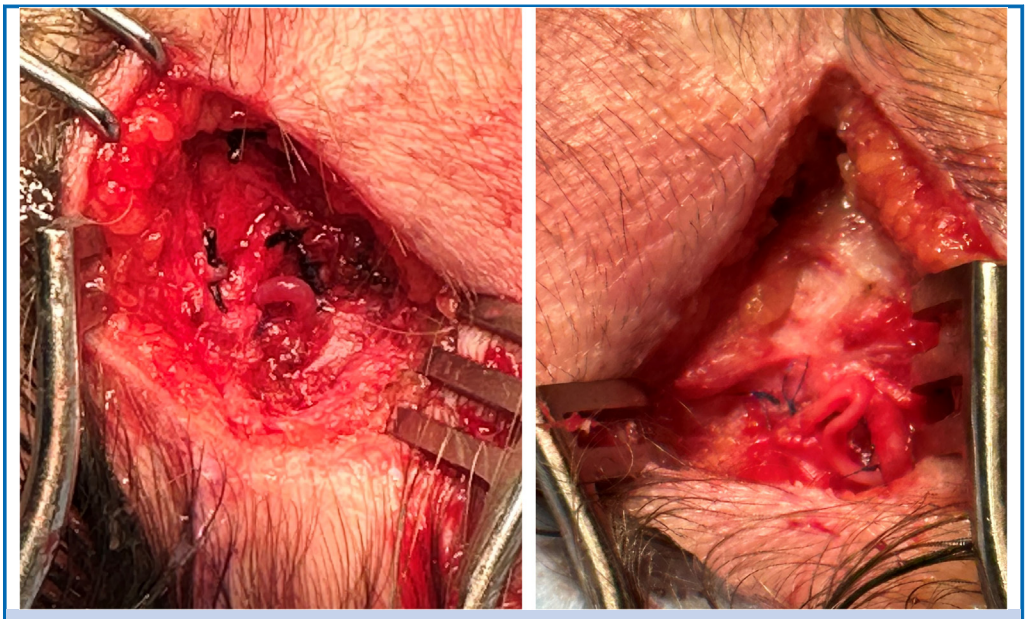

FIGURE 3. Intraoperative photos of the subcutaneous segment of the occipital artery after it has been dissected free from the surrounding tissue.

artery biopsy.[6,7] The technique to biopsy a short segment of the OA through a small incision has never been described in the literature up to now, despite having numerous neurosurgical studies for an extra-intracranial bypass through an extensive scalp incision.

This article aims to fill the gap and illustrate, in a stepwise fashion, how to perform an OA biopsy in the setting of GCA.

The course of the OA is divided into 3 segments. The most distal segment, the subcutaneous segment, runs in the connective tissue just above the galea aponeurosis, away from important structures, and can be easily dissected off. It is in average 5 cm long, allowing for >2 cm to be harvested without difficulty as has been suggested to minimize the risk of false negative pathological results.[16]

In a retrospective review of the 1190 biopsies from 1163 patients, a positive compared with negative pathologic diagnosis was associated with increased age (75.3 vs 71.3 years), ESR (57 vs 36 mm/h), C-reactive protein (51.6 vs 12.1 mg/L), and biopsy length (1.6 vs 1.2 cm).[16]

The same authors suggested a 1.5-2.0 cm biopsy specimen prefixation length as optimal, with greater lengths unlikely to provide significant additional diagnostic yield to justify risks.[16]

The surgical incision of the biopsy can be only 3-5 cm long and is precisely planned by performing an intraoperative ultrasound at the beginning of the procedure, and sterile gel can be used to keep hair out of the operative field, avoiding the need to cut hair.

Of importance, the OA is very small in diameter and can be confused with the occipital vein. To differentiate the artery from the vein we recommend using Doppler. In some cases, when the vessel caliber is small and the ultrasonic insonation of the artery difficult, we suggest harvesting and sending both artery and vein for pathological examination.

CONCLUSION

Giant cell arteritis is an autoimmune disease that can lead to permanent vision loss if not treated in a timely fashion. Currently, the temporal artery represents an important part of the diagnostic work-up. However, knowing how to perform an OA biopsy is key, as most patients with occipital headache/nuchal pain or prior negative temporal biopsy with atypical symptoms could have an isolated involvement of the OA. This article provides

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detailed information on how to perform an OA biopsy safely and effectively through a small incision.

POTENTIAL COMPETING INTERESTS

All the authors have no conflict of interest or financial disclosures.

Abbreviations and Acronyms: **CRP**, c-creative protein; **ESR**, erythrocyte sedimentation rate; **FUO**, fever of unknown origin; **GCA**, giant cell arteritis; **OA**, occipital artery; **SNL**, superior nuchal line

Correspondence: Address to Tiziano Tallarita, MD, Department of Cardiovascular Surgery, Mayo Clinic Health System, 1400 Bellinger St, Eau Claire, WI 54702 (tallarita.tiziano@mayo.edu).

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