Perivascular Marginal Zone Lymphoma Mimicking Temporal Arteritis

Maximilian Linxweiler, MD¹, Andrea Hasenfus, MD², Gregor Wolf, MD¹, and Bernhard Schick, MD¹

No sponsorships or competing interests have been disclosed for this article.

Keywords
marginal zone lymphoma, giant cell arteritis, temporal artery biopsy, halo sign

Received May 12, 2014; revised September 8, 2014; accepted September 26, 2014.

Giant cell arteritis (GCA) is the most common form of primary systemic vasculitis, and it mainly affects large- and medium-sized vessels in elderly people. Its clinical symptoms include temporal headaches, jaw claudication, a thickened temporal artery, and in severe conditions, visual disturbance and stroke as a consequence of inflammatory vascular obliteration. However, similar symptoms can be caused by other diseases as well, such as granulomatosis with polyangiitis (Wegener’s granulomatosis) and vessel-infiltrating malignancies. In this article, we present the first report of a perivascular marginal zone lymphoma that affected the temporal artery and manifested the typical clinical symptoms and sonographic findings of GCA. The scientific use of the patient’s tissue and clinical data was approved by the Saarland Medical Association ethics review committee.

Case Report
A 73-year-old female was referred to our clinic with a chief complaint of left-sided temporal swelling accompanied by occasional frontotemporal headaches, jaw claudication, and vertigo for 3 months. She disclaimed any visual disturbance, fevers, night sweat, and weight loss. Her medical history consisted of dietetically treated type 2 diabetes; atrial fibrillation; and a right-sided, retrobulbar, indolent non-Hodgkin lymphoma (NHL) that had been treated with radiation. Aside from solid subcutaneous swelling at the region of the left temple, the clinical examination showed no conspicuous findings. In the ultrasound examination, marked wall thickening of the left temporal artery was found as a hypoechoic halo around the perfused lumen (Figure 1). The laboratory analysis showed no conspicuous findings including normal values for C-reactive proteins, leukocytes, and erythrocyte sedimentation rate.

Considering temporal arteritis, infectious disease, and neoplasm as differential diagnoses, we decided to perform a biopsy through transparotideal partial resectioning of the left temporal artery. Intraoperatively, the vessel appeared markedly thickened with palpable induration. The histomorphological analysis of the resected tissue, which was macroscopically described as homogeneously beige-colored soft tissue with a central lumen, showed an arterial vessel with a fragmented inner elastic membrane, but it had no transmural inflammatory infiltrate with giant cells, which would be typical of GCA. However, a large lymphoid infiltrate immuring the vessel wall was detected (Figure 2) and was further

Figure 1. Ultrasound imaging of the left temporal artery demonstrating a markedly thickened vessel wall measuring 2.5 mm and 3.1 mm.

Corresponding Author:
Maximilian Linxweiler, MD, Department of Otorhinolaryngology, Head and Neck Surgery, Saarland University Medical Center, Homburg, Saarland, Germany

Email: maximilian.linxweiler@uks.eu
characterized through immunohistochemical staining. Thereby, the lymphoid cells were characterized as positive for CD20, Bcl2, and predominantly Ki67, without any expression of cyclin D1 and CD10.

Based on these findings, the working diagnosis of GCA was changed to a recurrent, perivascular marginal zone lymphoma, as the same expression pattern of biomarkers was found for the right-sided, retrobulbar NHL that was diagnosed 2 years prior to the current disease. Curative-intended involved field radiation was started after the completion of staging examinations.

**Discussion**

Ultimately, GCA is a primary systemic vasculitis that mainly involves large- and medium-sized arteries. The diagnosis is based on characteristic clinical symptoms such as temporal headaches, jaw claudication, elevated inflammatory markers, and proof of an inflammatory infiltrate in a temporal artery biopsy specimen, which is considered as the gold standard for diagnosing GCA.3

Depending on the proliferating cell clone and the clinical course of the disease, NHL can be classified as either B- or T-cell NHL that can clinically appear as indolent or aggressive. Marginal zone lymphomas belong to the group of indolent B-cell NHL and can occur as extranodal, nodal, or splenic in form. Although the marginal zone lymphoma originating from mucosa-associated lymphoid tissue represents the most common extranodal type, its manifestation in the head area is relatively rare. To our knowledge, we present the first case of a marginal zone lymphoma affecting the temporal artery. Until now, only 4 other NHLs that affected the same vessel have been described in the literature.4

What we can learn from this case is that not only the clinical symptoms of GCA but also the sonographic finding of a perivascular halo can be mimicked by other diseases such as perivascular malignancies and infectious diseases.2,5 As the quality of ultrasonography has continuously improved over the past decades, some authors have proposed the view that the histological proof of a diagnosis would not be further needed for GCA once a halo sign is found.5 However, our case showed that this sonographic finding is not pathognomonic for GCA. Therefore, practitioners must consider alternative diagnoses and pursue further workups that include temporal artery biopsy to rule out other disease processes.

**Author Contributions**

Maximilian Linxweiler, data analysis, article drafting, article revision and final approval, responsibility for content of article; Andrea Hasenfus, data analysis, article drafting, final approval, responsibility for content of article, pathology consultation; Gregor Wolf, data analysis, article drafting, final approval, responsibility for content of article, surgery; Bernhard Schick, data analysis, article drafting, article revision and final approval, responsibility for content of article, surgery.

**Disclosures**

**Competing interests:** None.

**Sponsorships:** None.

**Funding source:** None.

**References**