Scalp and Tongue Necrosis as Rare Manifestations of Giant Cell Arteritis Complicated with a Preauricular Abscess

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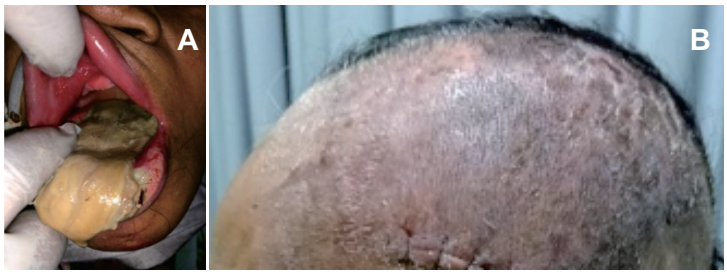

Figure 1. Tongue necrosis (A) and scalp necrosis over the temporoparietal area (B) of the patients at hospital admission.

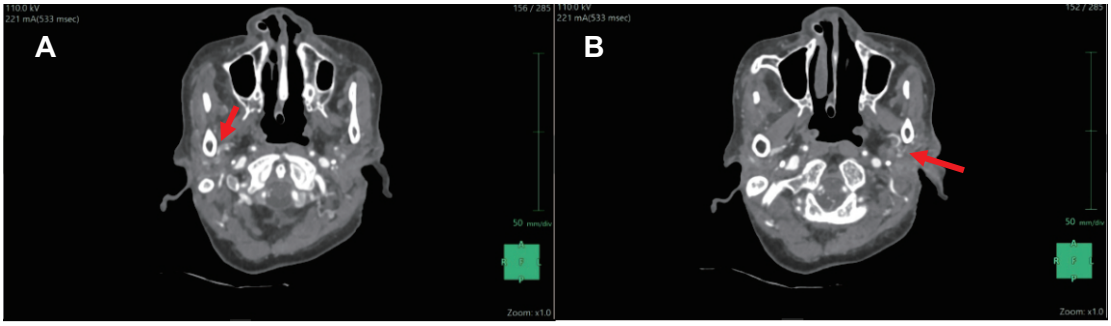

Figure 2. No contrast filling in the (A) right and (B) left temporal arteries (red arrows).

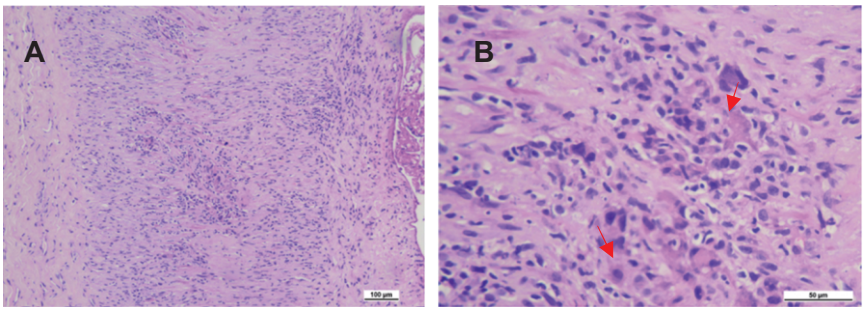

Figure 3. Histology of the temporal artery biopsy (hematoxylin and eosin).
Temporal artery biopsy showing an infiltration of inflammatory cells, such as lymphocytes and histiocytes, within the tunica media. Original magnification x100 (A) and x400 (B). At low power (A), the tunica media exhibits an irregular arrangement of muscle fibres. At high power (B), some histiocytes exhibit an epithelioid appearance, forming granulomas with interspersed multinucleated giant cells **(red arrows).**

Giant cell arteritis (GCA) is a primary large-vessel vasculitis that primarily affects large- to medium-sized arteries, particularly the cranial branches of the carotid arteries, and is characterized by granulomatous inflammation of the arterial walls.[1] GCA frequently manifests in individuals over 50 years of age, with the highest incidence observed in the eighth decade of life and a female predominance (female-to-male ratio of 3:1).[2] Immune tolerance within the arterial walls is a precursor to a wide array of interconnected aberrant immunological responses that involve both the innate and adaptive immune systems, thereby facilitating the onset and progression of GCA.[3]

Clinical manifestations of GCA are diverse, with the most common features comprising abrupt-onset headaches, temporal artery abnormalities, weakness or malaise, and scalp tenderness.[4] Inflammation of the vessel wall can lead to stenosis and subsequent ischemic complications. Among these, rare occurrences of scalp and tongue necrosis warrant particular attention, as they indicate a more severe disease phenotype and are associated with an increased risk of mortality.[5] Scalp necrosis has also been associated with an increased incidence of tongue necrosis, and the two conditions may occur simultaneously.[6] The rarity of these necrotic features often obscures the clinical presentation of GCA, thereby hindering prompt identification and delaying essential treatment.[7] Here, we present a rare case of concurrent scalp and tongue necrosis complicated by a preauricular abscess. To the best of our knowledge, this is the first documented case in Indonesia describing simultaneous scalp and tongue necrosis in a patient with GCA.

A 64-year-old woman with a history of hypertension and dyslipidemia was referred to a tertiary hospital in Surabaya, presenting with jaw claudication, scalp tenderness, and necrosis of both the scalp and tongue. Her current symptoms were preceded by a two-week history of worsening bitemporal headaches and general malaise. One week before admission, she developed dysarthria and dysphagia. She also reported pain with mastication consistent with jaw claudication. This was followed by an acute episode of nonreducible mandibular protrusion, for which she underwent surgical repositioning of the temporomandibular joint. Upon admission, she was hypertensive (150/90 mmHg), while other vital signs were within the normal range. She denied fever, visual disturbances, or other systemic complaints. Oral examination identified whitish plaques over the dorsal tongue and lower lip, along with previously documented areas of tongue necrosis (Figure 1A). The scalp was mottled, with a thick crust and necrotic lesions (Figure 1B). Additionally, active purulent discharge was observed originating from the left preauricular region. Laboratory evaluation showed a marked acute-phase response, with an elevated C-reactive protein (CRP) of 19.28 mg/dL, an Erythrocyte Sedimentation Rate (ESR) exceeding 140 mm/h, and leukocytosis (13,750 cells/mm^3). The rapid onset of tongue

necrosis, presenting with pallor and without signs of tumorous invasion, ruled out malignancy as a primary differential diagnosis. Given the constellation of symptoms, specifically the jaw claudication, temporal headache, and significantly elevated inflammatory markers, a rheumatology consultation was sought to evaluate for GCA.

Doppler ultrasound of the carotid arteries demonstrated absent flow in the frontal and parietal branches of the superficial temporal arteries, suggestive of stenosis. This finding was corroborated by computed tomography (CT) angiography, which showed no contrast filling in either temporal artery, suggesting bilateral stenosis (Figure 2). Stenosis of the right distal lingual artery was also observed. A concurrent soft-tissue ultrasound identified a left preauricular abscess measuring 0.39 × 1.2 × 3.1 cm. Serological testing for anti-neutrophil cytoplasmic antibodies (ANCA) was negative. Given the clinical presentation of jaw claudication, scalp tenderness, and temporal headache, supported by laboratory markers and imaging evidence of temporal artery involvement, GCA was strongly suspected. This diagnosis was maintained despite the presence of scalp and tongue necrosis, an atypical and rare ischemic manifestation of GCA. Meeting the 2022 ACR/EULAR criteria, the patient was immediately started on high-dose prednisone (1 mg/kg/day). Concurrently, an antibiotic regimen was administered to manage the left preauricular abscess identified on examination. Subsequently, the patient underwent surgical management that included debridement of the preauricular abscess, temporal artery biopsy, and partial excision of necrotic tongue tissue. The postoperative course was marked by pain at the surgical sites, which was managed with appropriate analgesics. Following clinical improvement, the patient was discharged in stable condition after a 10-day hospitalization. Nutritional requirements continued to be met through nasogastric tube feeding. The discharge medication regimen included a tapering course of oral prednisone, a completed course of oral antibiotics, and analgesics.

During the outpatient clinic follow-up visit, there was a significant reduction in temporal headache and jaw claudication. Laboratory investigations showed a downward trend in inflammatory markers, with a CRP level of 3.9 mg/dL and an ESR of 86 mm/h. Histopathological analysis of the 1.5-cm temporal artery biopsy specimen confirmed the diagnosis of GCA. The biopsy displayed classic GCA features: an infiltration of inflammatory cells, such as lymphocytes and histiocytes, within the tunica media with interspersed multinucleated giant cells (Figure 3). Simultaneously, the resected tongue tissue revealed extensive necrosis with preserved architecture and a mixed inflammatory infiltrate (lymphocytes, neutrophils, histiocytes, and plasma cells). Microbiological evaluation identified Staphylococcus aureus susceptible to the empiric antibiotics administered. Given the favorable clinical and laboratory evolution, the corticosteroid regimen was tapered to prednisone 20 mg/day.

GCA is an inflammatory vasculitis that affects medium- to large-sized vessels and occurs in individuals aged 50 years and older. In this case, the patient presented with cranial symptoms, including jaw claudication, along with the rare ischemic manifestations of scalp and tongue necrosis. As a manifestation of GCA, tongue necrosis falls within the spectrum of ischemic mucocutaneous presentations, which also notably includes necrosis of the lip and scalp.[1] An atypical presentation of tongue necrosis can delay diagnosis because many differential diagnoses must be excluded: underlying malignancy, radiation and chemotherapy exposure, and infectious etiologies.[2] The patient also exhibited scalp tenderness, a clinical feature reported in approximately 40% of GCA cases and clinically significant due to its strong association with scalp, tongue, or lip necrosis.[3]

Ultrasound examination showed absent flow in the branches of the superficial temporal artery, consistent with CT angiography findings of non-enhancing bilateral temporal arteries indicative of stenosis and simultaneous involvement of the right lingual artery. According to the 2022 ACR/EULAR classification criteria for GCA[4], the patient fulfilled diagnostic criteria, as she experienced scalp and tongue necrosis,

although this is not part of the classification criteria. A temporal artery biopsy (TAB) of ≥ 1 cm was obtained and revealed epithelioid transformation forming granulomatous structures with multinucleated giant cells—features characteristic of GCA. The combination of jaw claudication, temporal artery abnormalities, and a biopsy length of ≥1 cm is associated with an increased probability of obtaining a diagnostic temporal artery biopsy.[5]

Tongue necrosis is considered an unfavorable prognostic indicator in GCA, driven by vascular obliteration and ischemia.[6] Because improvement in ischemic symptoms typically occurs within 24 to 72 hours of initiating steroid therapy, high-dose glucocorticoids should be promptly administered upon clinical suspicion, even before biopsy confirmation. The 2021 ACR/Vasculitis Foundation guidelines recommend that patients with active GCA receive an initial high dose of prednisone at 1 mg/kg/day, up to a maximum of 80 mg or its equivalent, to induce remission of active disease.[7] Although conservative management is typically preferred, surgical intervention may be required based on the severity of tissue damage. Upon presentation, the patient exhibited a preauricular abscess with purulent discharge. This case highlights that infectious complications, such as a preauricular abscess, may coincide with GCA even in treatment-naïve patients who have not yet received steroids or immunosuppressants. The interplay between infection and GCA pathogenesis can be attributed to several factors. It is plausible that immune dysregulation renders GCA patients more susceptible to infectious agents.[8] In addition, because GCA primarily affects older individuals, age-related immune decline (immunosenescence) likely contributes to increased susceptibility.[9] Recent literature also challenges the notion of vascular sterility, suggesting that a resident vascular microbiome may also contribute to inflammation.[10] A multimodal approach was implemented to preserve residual tissue and facilitate recovery. The patient received high-dose prednisone (1 mg/kg/day) and empirical antibiotics, along with surgical debridement of a preauricular abscess and resection of necrotic tongue tissue. This aggressive management strategy yielded a favorable clinical outcome.

GCA presents a diverse array of clinical symptoms and findings attributable to the various affected vessels. This case describes a rare and severe presentation of GCA, characterized by the rapid onset of scalp and tongue necrosis leading to extensive tissue loss accompanied by a preauricular abscess. A high index of suspicion is imperative for diagnosing GCA in patients over 50 years of age who present with tongue necrosis, particularly when no alternative explanation is identified. Although the diagnosis relies on a combination of clinical features, laboratory markers, and imaging studies, temporal artery biopsy remains the diagnostic gold standard. Obtaining an adequately long arterial segment is recommended to improve diagnostic yield. This case underscores the importance of early recognition and the initiation of concomitant corticosteroids and adjunctive measures to achieve prompt disease control in patients with GCA.

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CONFLICT OF INTERESTS

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Jennette JC, Falk RJ, Bacon PA, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum. 2013;65(1):1–11.
2. Pugh D, Karabayas M, Basu N, et al. Large vessel vasculitis. Nat Rev Dis Prim. 2023;7 (1):93.
3. Ciccia F, Macaluso F, Mauro D, Nicoletti GF, Croci S, Salvarani C. New insights into the pathogenesis of giant cell arteritis: are they relevant for precision medicine? Lancet Rheumatol. 2021;(3):e874-85.
4. Osiowski A, Osiowski M, Stolarz K, Klepinowski T, Taterra D. Headache as the most common manifestation of giant cell arteritis?: A systematic review with meta-analysis. Rheumatol Int. 2025;(45):47.
5. Sobrinho RAB de S, de Lima KCA, Moura HC, Araújo MM, de Assis CMRB, Gouveia PA da C. Case report tongue necrosis secondary to giant cell arteritis: A

case report and literature review. Case Rep Med. 2017;6327437.
6. Fongaufier C, Guffroy A, Lutz JC. Tongue and scalp necrosis: Simultaneous initial complications revealing giant cell arteritis. J Rheumatol. 2018;45:873–4.
7. Jalaledin DS, Ross C, Makhzoum J-P. Rare ischemic complications of giant cell arteritis: Case series and literature review. Am J Case Rep. 2022;(23):e937565.
8. Prieto-Pena D, Castaneda S, Atienza-Mateo B, Blanco R, González-Gay MA. A review of the dermatological complications of giant cell arteritis. Clin Cosmet Investig Dermatol. 2021;14:303–12.
9. Chehem FCD, de Mornac D, Feuillet F, et al. Giant cell arteritis associated with scalp, tongue or lip necrosis: A French multicenter case-control study. Semin Arthritis Rheum. 2024;64:152348.
10. Ponte C, Grayson PC, Robson JC, et al. 2022 American College of Rheumatology/ EULAR classification criteria for giant cell arteritis. Ann Rheum Dis. 2022;81:1647–53.
11. Reis LA dos, Faustino I, Vargas P, Santos-Silva A, Lopes M. Lingual necrosis leading to the diagnosis of giant cell arteritis. Case Hist Rep. 2021;41:408–10.
12. Chung SA, Langford CA, Maz M, et al. 2021 American College of Rheumatology/ Vasculitis Foundation Guideline for the Management of Antineutrophil Cytoplasmic Antibody–Associated Vasculitis. Arthritis Care Res. 2021;73 No. 8:1088–105.
13. Rhee RL, Grayson PC, Merkel PA, Tomasson G. Infections and the risk of incident giant cell arteritis: a population-based, case-control study. Ann Rheum Dis. 2017;76(6):1031–5.
14. Mohan S V., Liao YJ, Kim JW, Goronzy JJ, Weyand CM. Giant cell arteritis: Immune and vascular aging as disease risk factors. Arthritis Res Ther. 2011;13:231.
15. Clifford A, Hoffman GS. Evidence for a vascular microbiome and its role in vessel health and disease. Curr Opin Rheumatol. 2015;27:397–405.