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Case Report / Olgu Sunumu

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# Intra-arterial angiolymphoid hyperplasia with eosinophilia mimicking temporal arteritis: A case report and review of the literature

## Temporal arteritin nadir bir taklitçisi intra-arteriyel eozinofilik anjiyolenfoid hiperplazi: Olgu sunumu ve literatürün gözden geçirilmesi

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**Abstract**

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a rare benign disease of unknown etiology, characterized by abnormal vascular proliferation. ALHE usually presents with painful, itchy, or asymptomatic lesions in the neck region, particularly around the ears. Clinically, it is usually around the ear, at the border of the scalp. It can be seen as single or multiple nodules or papules lesions on the neck and neck. Medium-sized peripheral muscular artery involvement is rare. A 41-year-old male patient, presented to a rheumatology clinic for 1 year history of a soft painless swelling in his bilateral temporal region. He sustained no preceding trauma to the area and had no additional symptoms. On physical examination, temporal arteries were prominent, but there was no tenderness. The patient had no clinical signs of temporal arteritis. Temporal artery Doppler ultrasound was performed, which revealed an echogenic asymmetric vessel wall thickening in the bilateral superficial temporal arteries. Temporal artery biopsy yielded the final diagnosis of ALHE, a rare vascular involvement of unknown etiology. ALHE is a condition that should be kept in mind in the differential diagnosis of temporal arteritis.

**Keywords:** Temporal arteritis, angiolymphoid hyperplasia with eosinophilia, vascular proliferation

**Öz**

Eozinofilik anjiyolenfoid hiperplazi (EALH) farklı klinik ve histopatolojik özelliklere sahip, nadir görülen benign vazoproliferatif bir hastalıktır. En sık görülen klinik prezentasyon, baş ve boyun bölgesindeki dermal ve subkütan ağrısız nodüllerdir. Orta büyüklükteki periferik müsküler arter tutulumu nadirdir. Kırk bir yaşında erkek hasta, 1 yıldır bilateral temporal bölgede ağrısız şişlik şikayeti nedeniyle romatoloji polikliniğine başvurdu. Ek semptomu olmayan hastanın, temporal bölgede travma öyküsü yoktu. Fizik muayenesinde temporal arterler belirgindi, ancak hassasiyet yoktu. Temporal arterit klinik bulguları olmayan hastanın temporal arter Doppler ultrasonografisinde bilateral yüzeyel temporal arter damar duvarı ekojenitesinde asimetrik kalınlaşma saptandı. Temporal arter biyopsisiyle, etiyolojisi bilinmeyen nadir bir vasküler tutulum olan EALH tanısı konuldu. Temporal arterit ayırıcı tanısında EALH de akılda tutulmalıdır.

**Anahtar Kelimeler:** Temporal arterit, eozinofilik anjiyolenfoid hiperplazi, vasküler proliferasyon

## Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) is a benign vasoproliferative disorder of unknown etiology.[1] The majority (70%) of affected patients are women in their third to the fifth decade of life. Affected patients may have a family history. ALHE frequently involve the skin of the head and neck, particularly in the periauricular area and the scalp. Systemic involvement is rare in ALHE.[2-5]

Being a rare disorder that may mimic several conditions such as lipoma, sebaceous cyst, Dupuytren's disease, or giant

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cell arteritis[2,6,7], ALHE may impose a diagnostic challenge with a risk of misdiagnosis or delayed diagnosis.[8-11]

Here, we present a case of intra-arterial ALHE mimicking temporal arteritis with prominent temporal arteries.

## Case Report

A 41-year-old male patient was referred to the rheumatology outpatient clinic with the suspected diagnosis of temporal arteritis due to bilateral swelling in the temporal region. The patient described a painless, non-pruritic swelling in the temporal region bilaterally lasting for 1 year along with no history of trauma, bruising, bleeding, or a chronic disease. The patient had no signs/symptoms related to polymyalgia rheumatica, while jaw claudication or visual symptoms were also absent.

Physical examination revealed a painless subcutaneous nodule on the surface of prominent temporal arteries bilaterally in the temporal region. No murmurs were noted in the temporal and carotid arteries, and there was no significant pulsation in the temporal arteries (Figure 1). Previous cranial computed tomography had revealed the thickening of the vessel wall in the superficial frontal branches of both temporal arteries, which was in the foreground on the right side. Temporal artery Doppler ultrasound was performed, which revealed an echogenic asymmetric vessel wall thickening in the bilateral superficial temporal arteries (Figure 2). Hemogram findings, eosinophil count, erythrocyte sedimentation rate, C-reactive protein levels and blood biochemistry findings were within the normal limits. An excisional biopsy of the temporal artery was performed. In the histological examination, an intravascular proliferation was noted that was composed of epithelioid endothelial cells, lymphocytes, and many eosinophils. There were no multinucleated giant cells or granulomatous inflammation. Immunohistochemical staining showed positive immunoreactivity for CD 68 (focal), CD 31, CD 34 and elastin (focal) expression. Histological analysis of the lesion revealed a diagnosis of eosinophilic angiolymphoid hyperplasia involving branches of the superficial temporal artery (Figures 3 and 4). Although the patient was referred to the relevant departments for surgical excision, medical treatment, laser therapy, or cryotherapy, he refused the treatment.

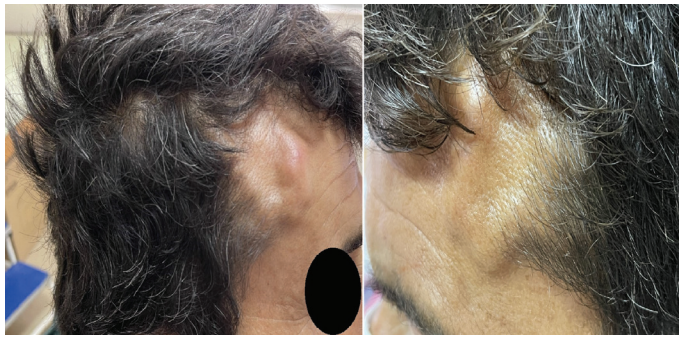

**Figure 1.** Enlargement of the right and left superficial temporal artery

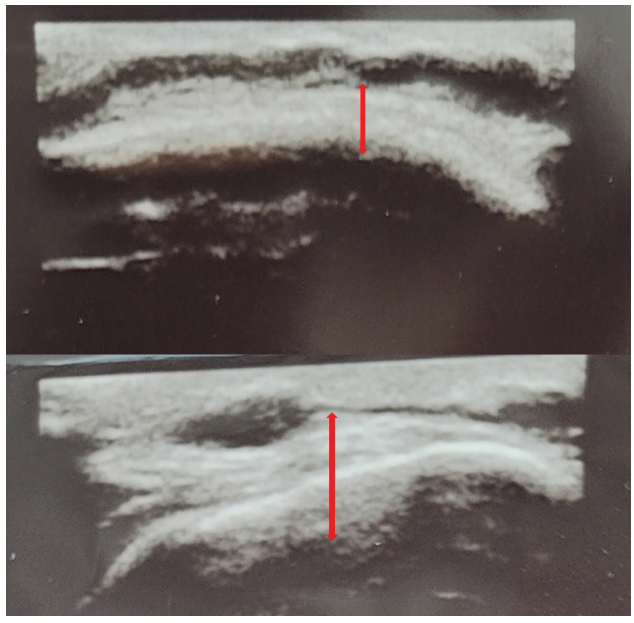

**Figure 2.** Ultrasound image of a branch of left superficial tempral artery showing asymmetric thickening of the vessel wall, producing a halo around the lumen

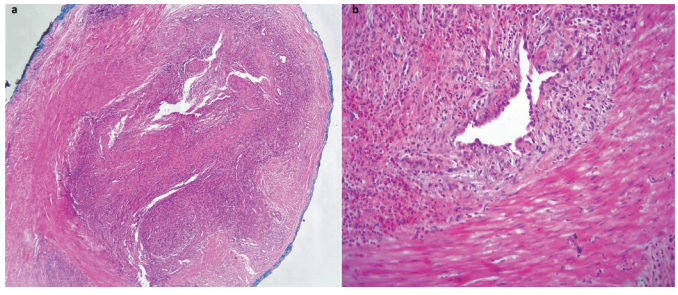

**Figure 3A.** Arterial structure with peripheral inflammation obliterating the lümen (x40 HE), **B.** Prominence and neovascularization of endothelial cells in the arterial wall (X200 HE)

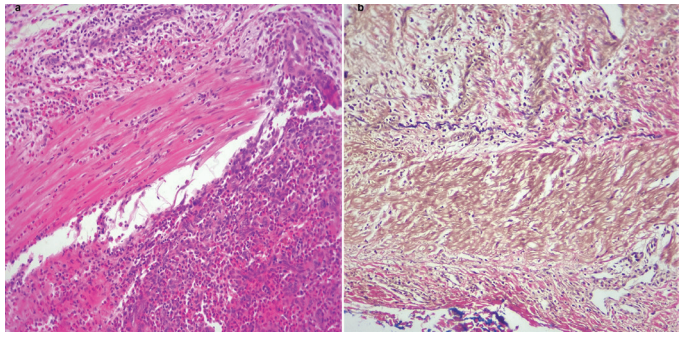

**Figure 4A.** Inflammatory cells characterized by mixed inflammation in the entire vessel wall (X200 HE), **B.** Verhoeffvan Gieson staining showing the internal elastic lamina (x200 HE)

## Discussion

ALHE is an uncommon disorder with its exact prevalence remains unknown, while several cases have been reported worldwide.[12,13] Although some authors consider ALHE a reactive process, it has also been suggested to represent a neoplastic process.[14,15] ALHE lesions usually present as single or multiple nodules in the head and neck region, especially in the periauricular area.[1,3] The lesions may be associated with pruritis, pain and ulceration, or an overlying pulsation.[16-19] In a review by Adler et al.[5], no gender-based differences were noted in the prevalence of ALHE along with an asymptomatic course in 15.4% of patients, while the most common localizations were considered as ear and periauricular area (36.3%), followed by face (28.2%) and scalp (17.3%).

Our patient had bilateral superotemporal lesions with no accompanying symptoms and was referred to our clinic with a suspected diagnosis of temporal arteritis due to bilateral prominent temporal arteries. In the current examination, Doppler ultrasound also revealed an echogenic asymmetric localized vessel wall thickening in the bilateral superficial temporal arteries, while the definitive diagnosis was made by the histopathological evaluation.

For ALHE cases involving the temporal region, the temporal arteritis is suggested to be considered in the differential diagnosis.[12] ALHE lesions can be seen adjacent to an artery, their development within the muscular artery is very rare.[20] Previous case reports indicated that the involved arteries comprise the temporal artery[2,6,12,14,20-31], radial artery[32-34], facial artery[35], post-auricular artery[36], popliteal artery[37], brachial artery[38,39], occipital artery[39], ulnar artery[9,40] and axillary artery[41] among these, cases of ALHE developed within the temporal arteries (Table 1).

Temporal arteritis is a vasculitis that can involve large- and medium-sized vessels. Presenting symptoms in patients with temporal arteritis may include tenderness in the temporal region, jaw claudication, localized headache, skin necrosis, eye involvement (i.e., ischemic optic neuropathy), fever, malaise, constitutional symptoms, polymyalgia rheumatica-like symptoms, arthralgia, myalgia and skin necrosis, whereas these symptoms are not expected in intra-arterial ALHE.[6,20,28] Nonetheless, temporal arteritis and intra-arterial ALHE may have some common manifestations, and the histopathological examination is required for definitive diagnosis.[2] Histologically, ALHE show varying degrees of nodular and diffuse lymphocytic infiltrates with eosinophils and an angio-proliferative lesion characterized by plump endothelial cells.[4] Mimicking vascular conditions include epithelioid hemangioendothelioma, Kimura's disease, angiosarcoma, eosinophilic granulomatosis with

**Table 1.** Angiolymphoid hyperplasia with eosinophilia (ALHE) of the temporal artery

| Authors | Case number | Age | Sex | Location | Artery | Eosinophil Counts |
|---|---|---|---|---|---|---|
| Grishman et al.[21] (1995) | 1 | 48 | F | Bitemporal area | Temporal artery | 2.800/mm$^3$ (24%) |
| Vadlamudi and Schinella[22] (1998) | 2 | 20 | M | The left forehead | Temporal artery | Unspecified |
| Kitamura et al.[6] (1999) | 3 | 68 | F | Right temporal area | Temporal artery, superior branch | Unspecified |
| Poilpre et al.[23] (1999) | 4 | 35 | F | Left temporal area | Temporal artery | Unspecified |
| Aurello et al.[24] (2003) | 5 | 40 | M | Right temporal area | Temporal artery, superior branch | Unspecified |
| Sandstad et al.[25] (2003) | 6 | 44 | M | Right temporal area | Temporal artery | Unspecified |
| Chopra et al.[26] (2007) | 7 | 46 | M | Left temporal area | Temporal artery | Unspecified |
| Koubaa et al.[27] (2008) | 8 | 34 | M | The right forehead | Temporal artery branch | Unspecified |
| Grum et al.[28] (2010) | 9 | 37 | M | Left temporal area | Superficial temporal artery | 17% (count not unspecified) |
| Hsiao and Wu[20] (2012) | 10 | 45 | M | Left temporal area | Temporal artery | Unspecified |
| | | 35 | M | The left forehead | Temporal artery, frontal branch | Unspecified |
| Giovagnorio and Miozzi[12] (2013) | 11 | 60 | M | Right temporal area | Superficial temporal artery | Normally specified (count not unspecified) |
| Imran[29] (2016) | 12 | 29 | M | Left temporal area | Temporal artery branch | Normally specified (count not unspecified) |
| Warner et al.[14] (2017) | 13 | 59 | F | Left temporal area | Frontal branch of the superficial temporal artery | Normally specified (count not unspecified) |
| Ansari et al.[30] (2019) | 14 | 32 | M | Right temporal area | Temporal artery | Unspecified |
| Li et al.[2] (2020) | 15 | 20 | M | Right temporal area | Temporal artery | Normally specified (count not unspecified) |
| Nakajima et al.[31] (2021) | 16 | 54 | F | Right temporal area | Temporal artery | Unspecified |
| The present case (2021) | 17 | 41 | M | Bilateral temporal area | Superficial temporal artery | 320/mm$^3$ (4%) normally detected |

polyangiitis. Conditions which may involve the scalp and subcutaneous tissue in the head region, such as Kaposi's sarcoma, metastasis, lymphoma and pyogenic granuloma, should also be considered in the differential diagnosis.[2,4]

In this case, temporal arteritis was suspected because of the prominence of the temporal arteries. The patient's age, normal acute phase response parameters, the absence of fever and constitutional symptoms, as well as lack of visual symptoms or symptoms related to polymyalgia rheumatica were the findings not supportive for the temporal arteritis diagnosis. In this study, although the halo sign in the temporal arteries was not typically seen on Doppler ultrasound imaging, there was echogenic asymmetric vessel wall thickening in the bilateral superficial temporal arteries. In fact, the halo sign, which is hypoechoic thickening of the temporal artery wall in Doppler ultrasound imaging, is frequently used in clinical practice in the diagnosis of temporal arteritis. Halo sign can also be evident, but less frequently, in other conditions such as infection, polymyalgia rheumatica and malignancy.[8,42] The finding of halo sign on ultrasound in ALHE has been previously reported in one case report. [8] The halo sign in temporal arteritis is not always related to an inflammatory process and may refer to an edema in the vessel wall rather than an inflammatory process.[8,43] In the case of intra-arterial ALHE, the halo sign can be seen if the vessel lumen is not completely obliterated. Indeed, the halo sign in intra-arterial ALHE has been associated with the presence of marked thickening of the vessel wall due to perivascular infiltrate.[8]

## Conclusion

In conclusion, our findings indicate that ALHE, as a rare disorder, should be considered in the differential diagnosis of temporal arteritis, as it may mimic temporal arteritis in terms of clinical appearance and Doppler ultrasound findings.

### Ethics

**Informed Consent:** Written informed consent was obtained from the patient for the anonymized information to be published in this article.

**Peer-review:** Externally peer-reviewed.

### Authorship Contributions

Concept: S.İ., A.E., Design: S.İ., A.E., Data Collection or Processing: S.İ., A.E., G.K., Ç.T., Analysis or Interpretation: S.İ., A.E., G.K., Ç.T., Literature Search: S.İ., A.E., G.K., Ç.T., Writing: S.İ., A.E., G.K., Ç.T.

**Conflict of Interest:** No conflict of interest was declared by the authors.

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## References

1. Amin A, Umashankar T, Dsouza CO. Intra-Arterial Angiolymphoid Hyperplasia with Eosinophilia: A Rare Case Report of Peripheral Medium Sized Muscular Artery Involvement. J Clin Diagn Res 2015;9:16-7.
2. Li E, Sinard J, Distefano A, Sohrab M. Angiolymphoid Hyperplasia with Eosinophilia with Clinical Presentation Concerning for Juvenile Temporal Arteritis. Ocul Oncol Pathol 2020;6:25-30.
3. Kakşi SA, Balevi A, Yüksel M, Özdemir M, Cakir A. Kriyoterapi ile kümelenen eozinofilili Anjiolenfoid hiperplazi. Türkiye Klinikleri Dermatoloji 2016;26:50-3.
4. Fite-Trepat L, Martos-Fernandez M, Alberola-Ferranti M, Pablo-Garcia-Cuenca A, Bescosatin C. Angiolymphoid Hyperplasia with Eosinophilia Involving the Occipital Artery: Case Report and Review of Literature. J Clin Diagn Res 2017;11:21-3.
5. Adler BL, Krausz AE, Minuti A, Silverberg JI, Lev-Tov H. Epidemiology and treatment of angiolymphoid hyperplasia with eosinophilia (ALHE): A systematic review. J Am Acad Dermatol 2016;74:506-12.
6. Kitamura H, Ito S, Kuwana N, Yutani C. Epithelioid hemangioma of the temporal artery clinically mimicking temporal arteritis. Pathol Int 1999;49:831-5.
7. Ahmad SM, Wani GM, Khursheed B, Qayoom S. Angiolymphoid hyperplasia with eosinophilia mimicking multiple cylindromas: a rare case report. Indian J Dermatol 2014;59:423.
8. Arnander MW, Anderson NG, Schönauer F. The ultrasound halo sign in angiolymphoid hyperplasia of the temporal artery. Br J Radiol 2006;79:184-6.
9. Krapohl BD, Machens HG, Reichert B, Mailänder P. A rare vasoproliferative lesion: angiolymphoid hyperplasia with eosinophilia of the hand. Br J Plast Surg 2003;56:168-70.
10. Lie JT, Gordon LP, Titus JL. Juvenile temporal arteritis. Biopsy study of four cases. JAMA 1975;234:496-9.
11. Lim CB, Khanduja V, Aleong JA, Douek M. A bloody mess: scalp lump misidentified. J R Soc Med 2005;98:64-5.
12. Giovagnorio F, Miozzi F. US and color Doppler findings in angiolymphoid hyperplasia with eosinophilia involving the superficial temporal artery. J Med Ultrason (2001) 2013;40:69-71.
13. Goldsmith W, Gilchrest K, Leffell P. Angiolymphoid Hyperplasia with Eosinophilia and Kimura Disease. In: Fitzpatrick TB, editor. Dermatology in general medicine. McGraw-Hill, Health Professions Division. New York: 2008;313-4.
14. Warner N, Marks JG, Cartee TV. Successful Treatment of Angiolymphoid Hyperplasia With Eosinophilia With Arterial Ligation and Pulsed Dye Laser. Dermatol Surg 2017;43:981-3.
15. Lee WJ, Kim MS, Lee MW, Choi JH, Moon KC, Koh JK. Angiolymphoid hyperplasia with eosinophilia associated with arteriovenous malformation. Clin Exp Dermatol 2009;34:272-3.
16. Guo R, Gavino AC. Angiolymphoid hyperplasia with eosinophilia. Arch Pathol Lab Med 2015;139:683-6.

17. Wells GC, Whimster IW. Subcutaneous angiolymphoid hyperplasia with eosinophilia. Br J Dermatol 1969;81:1-14.

18. Olsen TG, Helwig EB. Angiolymphoid hyperplasia with eosinophilia. A clinicopathologic study of 116 patients. J Am Acad Dermatol 1985;12:781-96.

19. Fetsch JF, Weiss SW. Observations concerning the pathogenesis ofepithelioid hemangioma (angiolymphoid hyperplasia). Mod Pathol 1991;4:449-55.

20. Hsiao HT, Wu YH. Intra-arterial angiolymphoid hyperplasia with eosinophilia of the temporal artery: Report of two cases and review of the literature. Dermatalogica Sinica 2012;30:79-100.

21. Grishman E, Wolfe D, Spiera H. Eosinophilic temporal and systemic arteritis. Hum Pathol 1995;26:241-4.

22. Vadlamudi G, Schinella R. Traumatic pseudoaneurysm: A possible early lesion in the spectrum of epithelioid hemangioma/ angiolymphoid hyperplasia with eosinophilia. Am J Dermatopathol 1998;20:113-7.

23. Poilpre M, Belleannee G, Trouette H, Lemaistre S, De Mascarel A. A nodular lesion of the left temporal artery. Ann Pathol 1999;19:53-4.

24. Aurello P, Cicchini C, D'Angelo F, Di Gioia CR, D'Amati G. Angiolymphoid hyperplasia with eosinophilia: A rare artery lesion. Anticancer Research 2003;23:3069-72.

25. Sandstad E, Aksnes H, Sund S, Reinholt FP. Recurrent angiolymphoid hyperplasia with eosinophilia mimicking temporal arteritis associated with nephrotic syndrome. Clin Nephrol 2003;59:206-11.

26. Chopra P, Handoo A, Parakh R. Epithelioid hemangioma of the temporal artery: a case report. Indian J Pathol Microbiol 2007;50:595-8.

27. Koubaa W, Verdier M, Perez M, Wechsler J. Intra-arterial angiolymphoid hyperplasia with eosinophilia. J Cutan Pathol 2008;35:495-8.

28. Grum F, Hufendiek K, Franz S, et al. High-resolution color-coded sonography in angiolymphoid hyperplasia with eosinophilia presenting as temporal arteritis. Circulation 2010;121:1045-6.

29. Imran AA. Angiolymphoid Hyperplasia With Eosinophilia–A Rare Entity. Biomedica 2016;32:257-60.

30. Ansari G, Pakozdi A, Etomi O. Angiolymphoid hyperplasia with eosinophilia mimicking temporal arteritis. Rheumatology Advances in Practice 2019;3:rkz024.001

31. Nakajima K, Kuroda E, Takeuchi S, Kadono T, Miyagaki T. Recurrent angiolymphoid hyperplasia with eosinophilia with CCR4 expression. Dermatol Sinica 2021;39:99-100.

32. Morton K, Robertson AJ, Hadden W. Angiolymphoid hyperplasia with eosinophilia: report of a case arising from the radial artery. Histopathology 1987;11:963-9.

33. Sandbank J, Zelikovski A, Haddad M, Sternberg E. Angiolymphoid hyperplasia with eosinophilia (epithelioid hemangioma). J Cardiovasc Surg 1991;32:370-2.

34. Khaira HS, Deshmukh NS, Vohra RK. Angiolymphoid hyperplasia presenting as a radial artery aneurysm. Eur J Vasc Endovasc Surg 1999;17:178-9.

35. Kimura Y, Tsutsumi T, Kuroishikawa Y, Kishimoto S. Angiolymphoid hyperplasia with eosinophilia arising from the facial artery. J Laryngol Otol 2003;117:570-3.

36. Cornelius RS, Biddinger PW, Gluckman JL. Angiolymphoid hyperplasia with eosinophilia of the head and neck. AJNR Am J Neuroradiol 1995;16:916-8.

37. Ghotbi R, Sotiriou A, Mlinaric M. Epitheloides Hämangiom der A. poplitea. Der Chirurg 1999;70:1494-6.

38. Arnold M, Geilen CC, Coupland SE, et al. Unilateral angiolymphoid hyperplasia with eosinophilia involving the left arm and hand. J Cutan Pathol 1999;26:436-40.

39. Vandy F, Izquierdo L, Liu J, Criado E. Angiolymphoid hyperplasia involving large arteries. J Vasc Surg 2008;47:1086-9.

40. Ozcanli H, Ozenci AM, Ozcanli C, Ibis S, Gurer IE. Angiolymphoid hyperplasia: a case of a rare arterial involvement and successful recurrence treatment with laser therapy. J Eur Acad Dermatol Venereol 2007;21:1106-7.

41. Bhat SP, Moorthy N, Ramalingam R, Jayapal M, Nanjappa MC. Angiolymphoid hyperplasia with eosinophilia presenting as a giant axillary artery aneurysm. Vascular 2010;18:49-52.

42. Nesher G, Shemesh D, Mates M, Sonnenblick M, Abramowitz HB. The predictive value of the halo sign in color Doppler ultrasonography of the temporal arteries for diagnosing giant cell arteritis. J Rheumatol 2002;29:1224-6.

43. Schmidt WA, Kraft HE, Vorpahl K, Volker L, Gromnica-Ihle EJ. Color duplex ultrasonography in the diagnosis of temporal arteritis. N Engl J Med 1997;337:1336-42.