

ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA WITH LATER ONSET - CASE REPORT

HIPERPLAZIE ANGIOLIMFOIDĂ CU EOZINOFILIE CU DEBUT TARDIV - CAZ CLINIC

Virgil Pătrașcu¹, Raluca Ciurea², Marius Eugen Ciurea³, Andreea - Oana Enache¹

¹Dermatology Department, University of Medicine and Pharmacy of Craiova, Romania

²Pathology Department, University of Medicine and Pharmacy of Craiova, Romania

³Plastic and Reconstructive Surgery Department, University of Medicine and Pharmacy of Craiova, Romania

Corresponding author:

Virgil Pătrașcu, Professor, MD, PhD,
University of Medicine and Pharmacy,
Petru Rareș Street, No 2-4, 200345, Craiova, Romania,
phone: 004-0724273676, e-mail: vm.patrascu@gmail.com

Conflict of interest: The authors do not have any conflict of interest to declare.

Open Access Article

Abstract

Keywords:

angiolymploid hyperplasia with eosinophilia, pathogenesis, treatment.

Angiolymploid hyperplasia with eosinophilia is a benign vasculo-proliferative disease, predominantly found in middle-aged women (20-50 years old) and with exceptionally cases reported in children or elderly. We report the case of a male patient who developed multiple lesions of angiolymploid hyperplasia with eosinophilia in the seventh decade of life. A 63-year-old man, without pathological antecedents presents with 6 nodular reddish lesions, 2-4 mm in size, moderately itchy, located in the left retroauricular region. Based on anamnesis, clinical examination, histopathological exam and other laboratory tests, we established the diagnosis of Angiolymploid hyperplasia with eosinophilia. After complete excision, immediate repair of the defect was performed using advancement flaps from the declive aria. After a 9-months follow-up there was no evidence of recurrence. Considering the possible internal organ involvement, we recommend a quarterly follow-up of the patient for an early diagnosis of other pathological conditions that can lead to a poor vital prognosis.

Cite this article:

Virgil Pătrașcu, Raluca Ciurea, Marius Eugen Ciurea, Andreea - Oana Enache. Angiolymploid hyperplasia with eosinophilia with later onset - case report. RoJCED 2016; 3(2):144 - 147

Rezumat

Cuvinte-cheie:

hiperplazia angiolimfoidă cu eozinofile, patogenie, tratament.

Hiperplazia angiolimfoidă cu eozinofile este o afecțiune vasculo-proliferativă benignă, întâlnită cu precădere la femei de vârstă medie (20-50 ani) și în mod excepțional la copii sau vârstnici. Noi prezentăm cazul unui bărbat la care boala a debutat în decada a VII-a de viață cu leziuni multiple. Pacient în vârstă de 63 ani, fără antecedente patologice, se spitalizează pentru 6 formațiuni nodulare de culoare roșietică, cu dimensiuni cuprinse între 2 și 4 mm, însoțite de prurit moderat, localizate retroauricular stâng. În urma anamnezei, examenului clinic, examenului histopatologic și a celorlalte investigații paraclinice am stabilit diagnosticul de Hiperplazie angiolimfoidă cu eozinofile. Am efectuat excizie completă și acoperirea lipsei de substanță prin lambouri cutanate avansate din partea declivă. Nu au existat semne de recidivă la 9 de luni de la intervenție.

Având în vedere posibila afectare organică, considerăm necesară supravegherea trimestrială a bolnavului pentru a depista din timp alte condiții patologice care pot întuneca prognosticul vital al acestuia.

Introduction

Angiolymphoid hyperplasia with eosinophilia (ALHE) was first described in 1969 by Wells and Whimster⁽¹⁾. ALHE is a benign vasoproliferative disease, being predominantly encountered in middle-aged women (20-50 years old) and, exceptionally, in children or elderly^(2,3).

We report the case of an elderly man who developed multiple lesions of ALHE in the seventh decade of life.

Case report

A 63-year-old man, without significant pathological history, presents with 6 itchy reddish lesions, 2-4 mm in size, located in the left retroauricular region. Lesions had appeared 12 months earlier without a history of trauma to the affected area (figs 1, 2). The regional lymph-nodes were not enlarged. No abnormality was detected in general physical examination.

All routine laboratory investigations including complete blood count, renal and liver function tests, peripheral blood eosinophils count and total serum Ig E level were within normal limits. Magnetic resonance imaging (MRI) excluded an underlying organic cause.

Histopathological examination of the skin lesions showed marked dermal vascular proliferation, lined by endothelial cells and perivascular infiltration with eosinophils and lymphocytes (figs 3, 4).

Based on anamnesis, clinical examination, histopathology and other laboratory tests we established the diagnosis of „Angiolymphoid hyperplasia with eosinophilia”.

After complete excision, immediate repair of the defect was performed using advancement flaps from the declive part.

At 9-months follow-up there was no evidence of recurrence.



Figures 1, 2.

Angiolymphoid hyperplasia with eosinophilia showing multiple lesions in the left retroauricular region

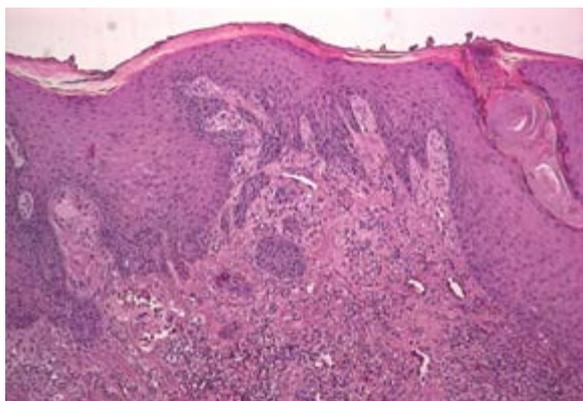


Figure 3. Acanthosis of the epidermis and proliferation of vascular channels in the deep dermis (HE stain, x40)

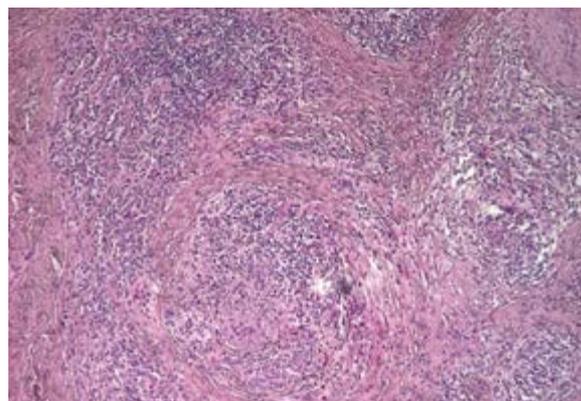


Figure 4. Dense, chronic, inflammatory infiltrate rich in eosinophils in dermis (HE stain, x100)

Discussion

ALHE has been described with different names including: epithelioid hemangioma, atypical pyogenic granuloma, pseudopyogenic granuloma, histiocytoid hemangioma⁽⁴⁾.

Most people who develop ALHE are aged between 20 and 50, with an average age of 30-33 at the time of diagnosis⁽⁵⁾. Rare cases have been described in children and elderly patients. The disease is more common in women than in men.

The pathogenesis of ALHE is complex and not fully understood.

Some authors consider ALHE as a vascular reaction secondary to some complex immunologic mechanisms⁽⁶⁾.

Olsen and Helwig consider that arteriovenous shunt is the main etiopathogenetic mechanism observed in 46 % of the cases⁽⁷⁾.

Kempf et al. showed the predominance of T lymphocytes and the rearrangement of TCR (T-cell receptor) in 5 patients with ALHE, leading to the supposition that ALHE is a low-grade neoplastic disease⁽⁸⁾.

A variety of factors have been incriminated in ALHE appearance such as: insect bites, local trauma (onset of the lesions ranges from 7 months to 20 years), infections (HIV, HTLV and HHV8), hyperestrogenism (in pregnancy or with oral contraceptive use), after vaccination⁽⁹⁾.

The disease is clinically characterized by single or multiple purplish red or brownish papules and subcutaneous nodules, with a range of 0.2-8 cm. The surface of the lesion may be smooth, ulcerated or crusted secondary to scratching⁽¹⁰⁾.

ALHE appears as a single lesion in 80% of patients but multiple disseminated cutaneous lesions were reported⁽¹¹⁾.

ALHE is most commonly found on the cephalic extremity, particularly on the auricular pavilions and periauricular regions, situation also present in our case.

Extracutaneous involvement is rare and has been reported in salivary glands, bone, colon, liver, orbit, spleen, lung, cardiovascular system⁽¹²⁾.

According to published studies, systemic eosino-

philia of 6-34% and regional lymphadenopathy may be found in 20% of patients. Serum levels of IgE are usually normal⁽¹³⁾.

Follicular mucinosis occurring along with ALHE has been described but without a clear association with mycosis fungoides⁽¹⁴⁾.

Also, peripheral T-cell lymphoma has been reported in patients with ALHE, thus it is hypothesized that ALHE might represent an early stage of T-cell lymphoma⁽¹⁵⁾.

The diagnosis is based on histopathology, which highlights the proliferation of small blood vessels and the presence of inflammatory infiltrate containing predominantly lymphocytes and eosinophils. Blood vessels are lined by enlarged endothelial cells with eosinophilic cytoplasm and large round nuclei. The cells are mostly cuboidal, with occasional "hobnail cells" appearance. In early stages of ALHE, the vascular component predominates, whereas in late stages of the disease lymphocytes become more prominent⁽¹⁶⁾.

Immunohistochemistry typically reveals a predominance of T lymphocytes and occasionally B cells that usually form the lymphoid follicles⁽¹⁷⁾.

ALHE should be distinguished from Kimura disease, initially thought to represent the same disease spectrum. They are now known to represent separate entities.

Kimura disease is characterized by deep soft tissue and subcutaneous lesions, lymphadenopathy, eosinophilia and elevated serum Immunoglobulin E⁽¹⁸⁾.

ALHE must also be differentiated from: bacillary angiomatosis, Kaposi sarcoma, eruptive keratoacanthomas, molluscum contagiosum, eruptive xanthomas, multiple trichoepitheliomas, cutaneous lymphomas/ pseudolymphomas, cutaneous metastases.

ALHE is a chronic disease, with a strong tendency to relapse after various medical therapies or surgery. Exceptionally, ALHE has shown spontaneous resolution⁽¹⁹⁾.

Complete surgical excision is the treatment of choice in ALHE with fewer lesions. In most cases,

relapse occurs as a result of incomplete surgical excision, being observed in 33% -50% of the cases. Better aesthetic results were obtained using Mohs surgery⁽²⁰⁾.

In a recent article, Wang et al. reported a case of ALHE treated with tacrolimus 0.1% ointment with the disappearance of lesions after 14 weeks of treatment⁽²¹⁾.

Other treatments that have been reported include: curettage and cautery, irradiation, laser therapy, corticosteroids (systemic, topical or intralesional preparation), topical tacrolimus 0,1%, topical imiquimod, indomethacin farnesil, intralesional interferon alfa-2a, treatment with oral retinoids and pentoxifylline, chemotherapeutic agents (vinblastine, bleomycin, fluorouracil)^(22,23).

Conclusion

Considering the possible organic involvement, we recommend as necessary a quarterly follow-up of the patient for an early diagnosis of other pathological conditions that can lead to a poor vital prognosis.

 This work is licensed under a Creative Commons Attribution 4.0 Unported License. The images or other third party material in this article are included in the article's Creative Commons license, unless indicated otherwise in the credit line; if the material is not included under the Creative Commons license, users will need to obtain permission from the license holder to reproduce the material. To view a copy of this license, visit <http://creativecommons.org/licenses/by-nc/4.0/>

Bibliography

- Karabudak O, Taskapan O, Bozdogan O, Dogan B. Angiolymphoid hyperplasia with eosinophilia: atypical appearance in an older patient. *Indian Journal of Dermatology*. 2008;53(3):144-145.
- Zaraa I, Mlika M, Chouk S, Chelly I, Mokni M, Zitouna M et al. Angiolymphoid hyperplasia with eosinophilia: A study of 7 cases. *Dermatology Online Journal*, 2011;17(2).
- Kim SM, Yoon J, Yoon T.J. Angiolymphoid Hyperplasia with Eosinophilia on the Palm. *Annals of Dermatology*. 2010;22(3):358-361.
- P. Doloi and S. Khanna. Angiolymphoid Hyperplasia with Eosinophilia—A Case Report. *International Journal of Otolaryngology and Head & Neck Surgery*, Vol. 1 No. 2, 2012, pp. 44-47.
- C.A. Moran and S. Suster. Angiolymphoid Hyperplasia with Eosinophilia (Epithelioid Hemangioma) of the Lung: A Clinicopathologic and Immunohistochemical Study of Two Cases. *American Journal of Clinical Pathology*, Vol. 123, No. 5, 2005, pp. 762-765.
- Devi B, Jena S, Kar D, Patro S, Behera B. Epithelioid Hemangioma: A Report of Two Cases. *Indian Journal of Dermatology*. 2014;59(5):510-512.
- Olsen TG, Helwig EB. Angiolymphoid hyperplasia with eosinophilia. A clinicopathologic study of 116 patients. *J Am Acad Dermatol*. 1985 May;12(5 Pt 1):781-96.
- Kempf W, Haeflner AC, Zepter K, Sander CA, Flaig MJ, Mueller B, et al. Angiolymphoid hyperplasia with eosinophilia: Evidence for a T-cell lymphoproliferative origin. *Hum Pathol* 2002;33:1023-9.
- Zarrin-Khameh N, Spoden JE, Tran RM. Angiolymphoid hyperplasia with eosinophilia associated with pregnancy: A case report and review of the literature. *Arch Pathol Lab Med*. 2005;129:1168-71.
- Trindade F, Haro R, Requena L. Giant angiolymphoid hyperplasia with eosinophilia on the chest. *J Cutan Pathol*. 2009 Apr;36(4):493-6.
- Farhadian, Joshua A; Shvartsbeyn, Marianna; Meehan, Shane A; &Urbanek, Richard W. (2015). Angiolymphoid hyperplasia with eosinophilia. *Dermatology Online Journal*, 21(12). doi. 29531.
- Moran CA, Suster S. Angiolymphoid hyperplasia with eosinophilia (epithelioid hemangioma) of the lung: A clinicopathologic and immunohistochemical study of two cases. *Am J Clin Pathol*. 2005;123:762-5.
- Aggarwal A, Keluskar V. Epithelioid hemangioma (angiolymphoid hyperplasia with eosinophilia) in the oral mucosa. *Indian J Dent Res* 2012;23:271-4
- Joshi R. Angiolymphoid hyperplasia with follicular mucinosis. *Indian J Dermatol Venereol Leprol*. 2007;73(5):346-347.
- Andreea J, Galle C, Magdorf K, et al. Severe atherosclerosis of the aorta and development of peripheral T-cell lymphoma in an adolescent with angiolymphoid hyperplasia with eosinophilia. *Br J Dermatol*. 2005;152(5): 1033-1038
- R Guo, A C P. Gavino. Angiolymphoid Hyperplasia With Eosinophilia. *Archives of Pathology & Laboratory Medicine* May 2015, Vol. 139, No. 5, pp. 683-686.
- Mukherjee B, Kadaskar J, Priyadarshini O, Krishnakumar S, Biswas J. Angiolymphoid Hyperplasia with Eosinophilia of the Orbit and Adnexa. *Ocul Oncol Pathol* 2016;2:40-47
- Sah P, Kamath A, Aramanadka C, Radhakrishnan R. Kimura's disease - An unusual presentation involving subcutaneous tissue, parotid gland and lymph node. *Journal of Oral and Maxillofacial Pathology: JOMFP*. 2013;17(3):455-459.
- Satpathy A, Moss C, Raafat F, Slator R. Spontaneous regression of a rare tumour in a child: angiolymphoid hyperplasia with eosinophilia of the hand: case report and review of the literature. *Br J Plast Surg*. 2005 Sep; 58(6):865-8.
- Miller CJ, Ioffreda MD, Ammirati CT. Mohs micrographic surgery for angiolymphoid hyperplasia with eosinophilia. *Dermatol Surg*. 2004 Aug;30(8):1169-73.
- Wang S, Li W. Angiolymphoid hyperplasia with eosinophilia successfully treated with tacrolimus ointment. *J Eur Acad Dermatol Venereol*. 2010 Feb;24(2):237.
- Baghestani S, Firooz A, Ghazisaidi MR. A refractory case of angiolymphoid hyperplasia with eosinophilia successfully treated by surgery. *J Dermatolog Treat*. 2011;22:49-51.
- Lebo S, Balato A, Cirillo T, Balato N. A Long-Term Follow-Up of Angiolymphoid Hyperplasia with Eosinophilia Treated by Corticosteroids: When a Traditional Therapy is Still Up-to-Date. *Case Reports in Dermatology*. 2011;3(1):64-67.