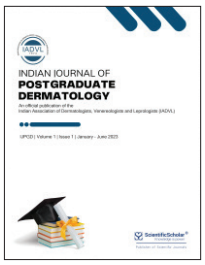
ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

https://ijpgderma.org

Indian Journal of Postgraduate Dermatology



Clinicopathologic Challenge

Nuchal Papules

Siddharth Mani[1]

[1]Department of Dermatology, Indian Naval Hospital Ship Sanjivani (INHS) Sanjivani, Kochi, Kerala, India.

***Corresponding author:**
Siddharth Mani,
Department of Dermatology,
Indian Naval Hospital Ship
Sanjivani, Kochi, Kerala, India.

smani5931@gmail.com

Received: 26 November 2023
Accepted: 20 April 2024
EPub Ahead of Print: 01 August 2024
Published: 23 August 2024

DOI
10.25259/IJPGD_114_2023

Quick Response Code:

CASE DESCRIPTION

Nuchal papules are a common clinical presentation in the dermatology outpatient department, with acne keloidalis nuchae (AKN) being the most common culprit. However, other entities such as folliculitis decalvans, hidradenitis suppurativa, dissecting cellulitis, acne conglobata, angiolymphoid hyperplasia with eosinophilia (ALHE), pyogenic granuloma, follicular cutaneous T-cell lymphoma and bacterial folliculitis may mimic AKN and confound the clinician. Herein, we present a clinicopathological challenge where the patient was initially diagnosed as AKN; however, dermoscopic and histopathological examination led to a change in the diagnosis.

A 26-year-old male reported with 6-month history of multiple painless papules and nodules over the nape of neck. The lesions gradually increased in size and number over the course of 6 months. The patient gave a history of keeping a short crop and wearing a collared shirt as a part of his job requirement, suggesting recurrent friction and trauma as an inciting factor. He denied any history of hair loss or similar lesions over his face, back, axilla or inguinal region. Physical examination revealed multiple, polysized, firm, non-tender, skin coloured to erythematous papules and nodules over the nape of neck [Figure 1]. There was no lymphadenopathy. Based on history and clinical examination, the patient was diagnosed with a case of AKN and empirically managed with four weekly intralesional injections of triamcinolone. However, the patient

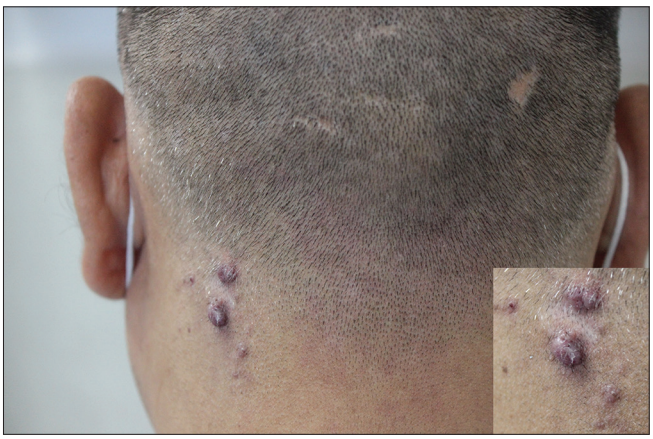


Figure 1: Multiple, polysized, erythematous to hyperpigmented papules and nodules on the nape of neck.

did not respond and the lesions kept on increasing in size and number. The patient reported an episode of bleeding from the lesion, which led us to reconsider the diagnosis. Dermoscopy revealed white–pink structureless areas, irregular white lines with interspersed red clods, serpentine vessels and white scales [Figure 2a]. A recent lesion showed dilated vessels [Figure 2b] (DermLite DL4 × 10; 3 Gen, San Juan Capistrano, CA, USA).

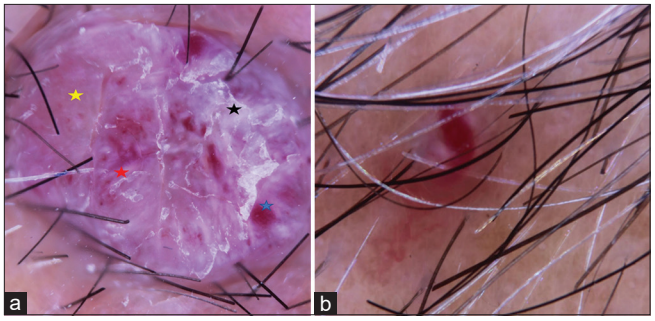


Figure 2: (a) Dermoscopy reveals white pink structureless areas (yellow star), irregular white lines with interspersed red clods (blue star), serpentine and dotted vessels (red star) and white scales (black star). Red clods indicate vascular dilation (DermLite DL4 × 10; 3 Gen, San Juan Capistrano, CA, USA). (b) Presence of dilated vessel in recent onset lesion. (DermLite DL4 × 10; 3 Gen, San Juan Capistrano, CA, USA).

HISTOPATHOLOGICAL FINDINGS

His peripheral eosinophil count and immunoglobulin E levels were normal. Excision biopsy revealed dilated blood vessels in the dermis with endothelial cells appearing as epithelioid cells in a background of multiple eosinophils and lymphocytes [Figure 3].

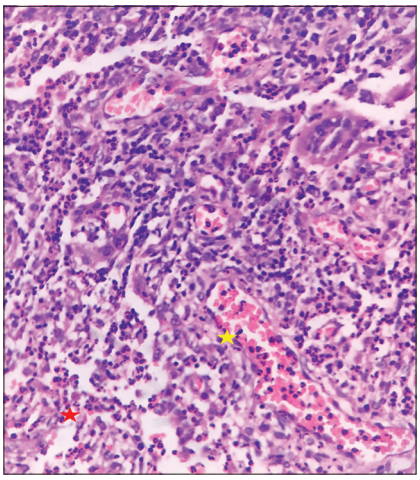


Figure 3: Dilated blood vessels in dermis with endothelial cells appearing as epithelioid cell (yellow star) and presence of plenty of eosinophils and lymphocytes (red star) (H and E, ×40). H&E: Haematoxylin and eosin.

DIAGNOSIS

Based on clinical, dermoscopic and histopathological findings, the patient was diagnosed as a case of ALHE.

DISCUSSION

Nuchal papules are a common clinical presentation in the dermatology outpatient department, with AKN being the most common culprit. However, other entities such as folliculitis decalvans, hidradenitis suppurativa, dissecting cellulitis, acne conglobata, ALHE, pyogenic granuloma,

Table 1: Difference between ALHE and Kimura's disease.

Features	Kimura's disease	ALHE
Initial description	1937 by Kim and Szeto Kimura *et al*. in 1948[5]	Wells and Whimster in 1969[1]
Age	Younger age of onset. Usually in 2nd decade	3rd–4th decade
Sex	Male>Female	Female>Male
Lymphadenopathy	More common. May involve cervical, epitrochlear, inguinal and posterior auricular nodes.	Less common. May involve cervical nodes.
Extracutaneous features	Common. May show Raynaud's phenomenon or bony involvement. Occasionally the condition has been described with ulcerative colitis, asthma and temporal arteritis	Rare
Blood/Peripheral eosinophilia	Marked	Rare. Usually in <10% cases
Elevated IgE	Frequent	Rare
Histopathology	Lymphoid follicular hyperplasia with reactive germinal centres. Presence of eosinophilic and lymphocytic infiltrate	Florid vascular proliferation with epithelioid-looking endothelial cells
Immunohistochemistry	IgE reticular staining	CD 31, CD34 and FVIII in vascular component
Duration of disease	Long	Short

ALHE: Angiolymphoid hyperplasia with eosinophilia, IgE: Immunoglobulin E

follicular cutaneous T-cell lymphoma and bacterial folliculitis may mimic AKN and confound the clinician. Our case was diagnosed on the basis of dermoscopy and histopathology.

ALHE was first described in 1969 by Wells and Whimster.[1] At present, ALHE also known as epithelioid haemangioma has been classified as a benign vascular tumour by the International Society for the Study of Vascular Anomalies in 2018. The tumour is very rare in India; however, its exact incidence or prevalence has not been reported. It usually presents with papules or nodules on the head, especially around the ears.[2] Lesions limited to the nape of neck have been rarely reported. ALHE lesions represent localised proliferations of capillaries around medium-sized vessels with an epithelioid endothelial cell lining and are surrounded by associated lymphocytic and eosinophilic infiltrates as seen in our case.[3]

ALHE can imitate multiple conditions, thereby leading to a diagnostic difficulty. Kimura's disease is another entity that was earlier considered to be closely related to ALHE, but now, it is widely recognised that both are separate entities [Table 1].[4-6] Although surgical excision is the treatment of choice, other modalities such as cryotherapy, electrofulguration, imiquimod and retinoids have been tried with success.[6,7] Our patient underwent serial surgical excision with no relapse over a 6-month follow-up.

The take home message from this case includes:

1. ALHE also known as epithelioid haemangioma is a rare benign vascular tumour
2. Clinically, it may mimic AKN and pose a diagnostic difficulty
3. History of bleeding from any lesion should alert the treating doctor regarding the presence of vascular tumour or malformation
4. Dermoscopy is an important tool in diagnosing vascular tumours/malformation
5. Histopathologically ALHE is characterised by endothelial cells appearing as epithelioid cells in a background of multiple eosinophils.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Wells GC, Whimster IW. Subcutaneous Angiolymphoid Hyperplasia with Eosinophilia Br J Dermatol 1969;81:1-14.
2. Urabe A, Tsuneyoshi M, Enjoji M. Epitheloid Hemangioma Versus Kimura's Disease. A Comparative Clinicopathologic Study. Am J Surg Pathol 1987;11:758-66.
3. Kitamura H, Ito S, Kuwana N, Yutani C. Epithelioid Hemangioma of the Temporal Artery Clinically Mimicking Temporal Arteritis. Pathol Int 1999;49:831-5.
4. Chong WS, Thomas A, Goh CL. Kimura's Disease and Angiolymphoid Hyperplasia with Eosinophilia: Two Disease Entities in the Same Patient. Int J Dermatol 2006;45:139-45.
5. Kimura T, Yoshimura S, Ishikawa E. On the Unusual Granulation Combined with Hyperplastic Changes of Lymphatic Tissue. Trans Soc Pathol Jpn 1948;37:179-80.
6. Ahmad SM, Wani GM, Khursheed B, Qayoom S. Angiolymphoid Hyperplasia with Eosionophilia Mimicking Cylindromas: A Rare Case Report. Indian J Dermatol 2014;59:423-5.
7. Reddy PK, Prasad AL, Sumathy TK, Shivaswamy KN, Ranganathan C. An Overlap of Angiolymphoid Hyperplasia with Eosinophilia and Kimura's Disease: Successful Treatment of Skin Lesion with Cryotherapy. Indian J Dermatol 2015;60:216-9.

How to cite this article: Mani S. Nuchal Papules. Indian J Postgrad Dermatol. 2024;2:138-40. doi: 10.25259/IJPGD_114_2023