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Diagnostic challenges and clinical insight of medial thigh hemangiolymphangioma in adult: A rare case report

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ABSTRACT

Vascular anomalies are classified into vascular tumors and vascular malformations. Hemangiolymphangioma (HLA) is a rare vascular malformation that contains both blood and lymphatic components, most commonly diagnosed in infancy or early childhood. Adult cases, especially in atypical locations such as the medial thigh, are exceedingly rare and pose diagnostic challenges. A 27 yo female presented with a slowly enlarging, painless lump on the left medial thigh. Initially misdiagnosed as condyloma acuminata, the lesion was subsequently identified as a vascular malformation through dermoscopic visualization of characteristic vascular lacunae. Histopathological and immunohistochemical analyses confirmed the diagnosis of HLA. Surgical excision was performed with clear margins, and no recurrence was observed after 3 mo. Hemangiolymphangiomas are benign but may exhibit local infiltration and recurrence, especially after incomplete resection. Diagnosis requires a multimodal approach including clinical assessment, dermoscopy, histopathology, and immunohistochemistry. Differential diagnosis includes hemangioma, lymphangioma, and malignancies such as lymphangiosarcoma. Complete surgical excision remains the treatment of choice, with other modalities like electrocautery or cryotherapy considered in selected cases. Long-term follow-up is crucial due to the risk of recurrence. In conclusion, this rare adult case of medial thigh HLA highlights the importance of considering vascular malformations in atypical anatomical sites. Early recognition and comprehensive diagnostic evaluation facilitate appropriate management and improve patient outcomes.

ABSTRAK

Kelianan vaskular dapat dibagi menjadi tumor vaskular dan malformasi vaskular. Hemangiolymphangioma (HLA) merupakan malformasi vaskular langka yang terdiri dari komponen pembuluh darah dan pembuluh limfatik, yang paling sering didiagnosis pada masa bayi atau awal masa kanak-kanak. Kasus pada usia dewasa, terutama pada lokasi atipikal seperti paha bagian medial, sangat jarang ditemukan dan dapat menimbulkan tantangan diagnostik. Seorang wanita berusia 27 tahun datang dengan keluhan benjolan yang membesar secara perlahan dan tidak nyeri pada paha medial kiri. Lesi tersebut awalnya didiagnosis sebagai kondiloma akuminata, namun kemudian diidentifikasi sebagai malformasi vaskular melalui pemeriksaan dermoskopi yang menunjukkan lakuna vaskular yang khas. Pemeriksaan histopatologis dan imunohistokimia mengonfirmasi diagnosis HLA. Eksisi bedah dilakukan dengan margin yang bersih dan tidak ditemukan tanda-tanda kekambuhan selama pemantauan tiga bulan pascaoperasi. Hemangiolimfangioma bersifat jinak, namun dapat menunjukkan infiltrasi lokal dan kekambuhan, terutama jika tidak dilakukan reseksi secara menyeluruh. Diagnosis memerlukan pendekatan multimodal yang mencakup evaluasi klinis, pemeriksaan dermoskopi, histopatologi, dan imunohistokimia. Diagnosis banding mencakup hemangioma, limfangioma, dan keganasan seperti limfangiosarkoma. Eksisi bedah secara menyeluruh tetap menjadi pilihan terapi utama, sementara modalitas lain seperti kauterisasi atau krioterapi dapat dipertimbangkan pada kasus tertentu. Tindak lanjut jangka panjang penting dilakukan mengingat adanya risiko kekambuhan. Simpulan, kasus HLA yang jarang terjadi pada paha medial pada pasien dewasa ini menyoroti pentingnya mempertimbangkan malformasi vaskular sebagai diagnosis banding pada lesi yang muncul di lokasi anatomi yang tidak lazim. Deteksi dini dan evaluasi diagnostik yang komprehensif memungkinkan penatalaksanaan yang tepat dan meningkatkan prognosis pada pasien.

Keywords:
hemangiolymphangioma;
vascular malformation;
lymphatic vessels;
medial thigh;
dermoscopy

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INTRODUCTION

Vascular anomalies are broadly divided into vascular tumors and vascular malformations. Vascular tumors, such as hemangiomas, are characterized by endothelial proliferation, whereas vascular malformations, including venous, arteriovenous, and lymphatic malformations, are structural anomalies resulting from errors in vascular morphogenesis.[1,2] Vascular malformations are relatively uncommon, affecting approximately 0.3% of the general population, with capillary malformations being the most frequently observed subtype.[1] Vascular malformations may exhibit mixed components, often comprising both lymphatic and venous endothelium, making it difficult to classify them as purely one type of lesion.[3]

Hemangiolymphangioma (HLA) is an extremely rare vascular malformation comprised of both endothelial and lymphatic components. Approximately 40–60% of HLA are diagnosed at birth, with up to 90% identified within the first two years of life, after which the incidence declines markedly with increasing age.[4,5] Although histologically benign, HLA can exhibit aggressive behavior, including rapid growth, infiltration into adjacent structures, and a potential for local recurrence.[6] Hemangiolymphangioma can occur in multiple anatomical areas like head & neck, axilla, abdominal cavity, extremities, testis, urinary bladder, and vertebral column.[5-7] Clinically, HLA typically presents as painless, slowly enlarging masses.[7] The definitive diagnosis of HLA typically requires a multimodal approach, incorporating clinical examination, imaging studies, histopathological analysis, and immunohistochemical staining.[8]

We reported a rare case of HLA in a 27 yo female presenting with a lesion located on the medial aspect of the left thigh, adjacent to the inguinal region. Due to the unusual anatomical location and its potential to be clinically mistaken for more common inguinal pathologies such as hernia, lymphadenopathy, or condyloma acuminata, this case underscores the diagnostic challenge and highlights the importance of considering vascular malformations in the differential diagnosis of inguinal masses in adults.

CASE

A 27 yo female patient came to our outpatient clinic with a complaint of a lump on the medial aspect of her left thigh. The lesion first appeared approximately ten years prior as a small, mole-like lump on the medial left thigh, without associated symptoms such as itching or pain. At that time, the patient neither sought medical consultation nor received any treatment, and the lesion was perceived to have flattened. Four years ago, the patient noticed an increase in the size of the lesion. She sought medical attention at a primary healthcare facility and was clinically diagnosed with condyloma acuminata. However, no treatment was administered due to her pregnancy at the time. One year ago, the lesion continued to enlarge, still without pain or pruritus, although intermittent bleeding was noted upon friction.

This patient denied any history of drug allergies, food allergies, atopy, or malignancy. There was also no relevant family history of similar complaints or hereditary conditions. Her primary concern was the increasing discomfort, prompting her to seek surgical removal of the lesion. On dermatovenereological examination, a solitary, skin-colored papule with a lobulated surface was observed on the left inner thigh, measuring approximately 1.5 cm in diameter (FIGURE 1). The lesion was firm on palpation and non-tender.

The differential diagnosis for this patient was HLA, hemangioma, lymphangioma, and lymphangiosarcoma. To establish a definitive diagnosis, dermoscopic and histopathological examinations were conducted. Dermoscopy was performed using a Heine Delta 20 dermatoscope under 10x magnification in polarized mode, revealing multiple, densely distributed, round to oval, yellowish-white, reddish to purplish translucent lacunae surrounded by pale septa along with a few linear and punctate vessels and reddish to brownish scattered dots and globules (FIGURE 2).

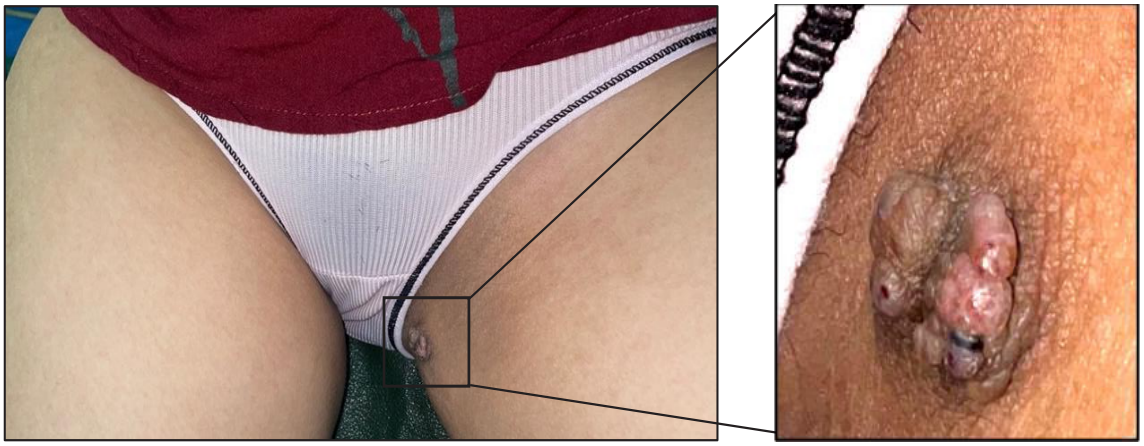

FIGURE 1. Clinical images of a skin-colored papule with a lobular surface, solitary

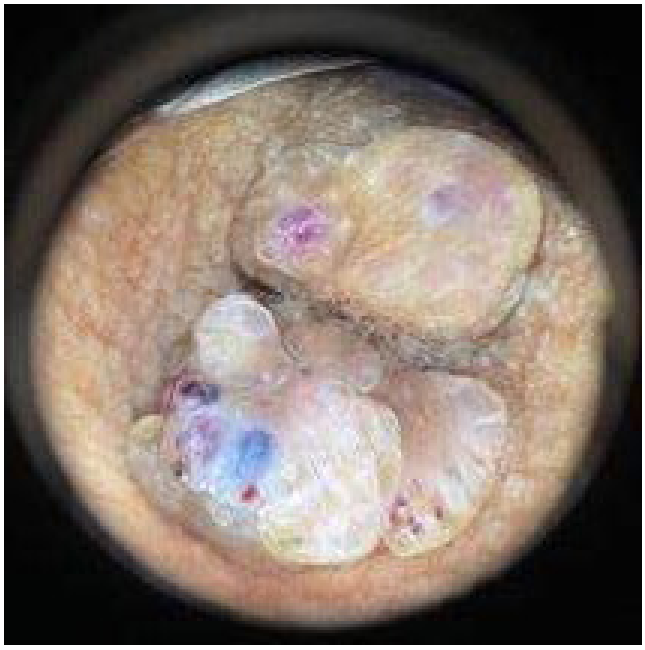

FIGURE 2. Dermoscopic examination of the lesion (Magnification 10x)

Histopathological analysis was conducted on a tissue sample fixed in 10% neutral buffered formalin and embedded in paraffin. Sections were stained with hematoxylin and eosin (H&E) using a standard protocol involving deparaffinization, hydration, hematoxylin staining, differentiation, bluing, eosin counterstaining, dehydration, and mounting. Histopathological examination showed basket weave and lamellar orthokeratosis, focal parakeratosis, irregular acanthosis, and elongated rete ridges around the dilated vessels. The superficial dermis and subcutis contained numerous dilated vessels of various shapes and sizes, with a lining endothelium without atypia, filled with amorphous proteinaceous masses, some vessels filled with erythrocytes, and surrounded by fibrous connective tissue. There was also extravasation of erythrocytes, especially in the subcutis, as well as lymphocytes, histiocytes, and neutrophils. No signs of malignancy were found (FIGURE 3).

To confirm the presence of lymphatic components, immunohistochemical staining using the D2-40 antibody (monoclonal mouse anti-human podoplanin) was performed following a standard protocol that included antigen retrieval via heat-induced epitope retrieval (HIER), incubation with the primary antibody, application of a secondary antibody, and visualization with DAB chromogen. The staining showed moderate membranous positivity in lymphatic vessels, while blood vessels remained negative (FIGURE 4).

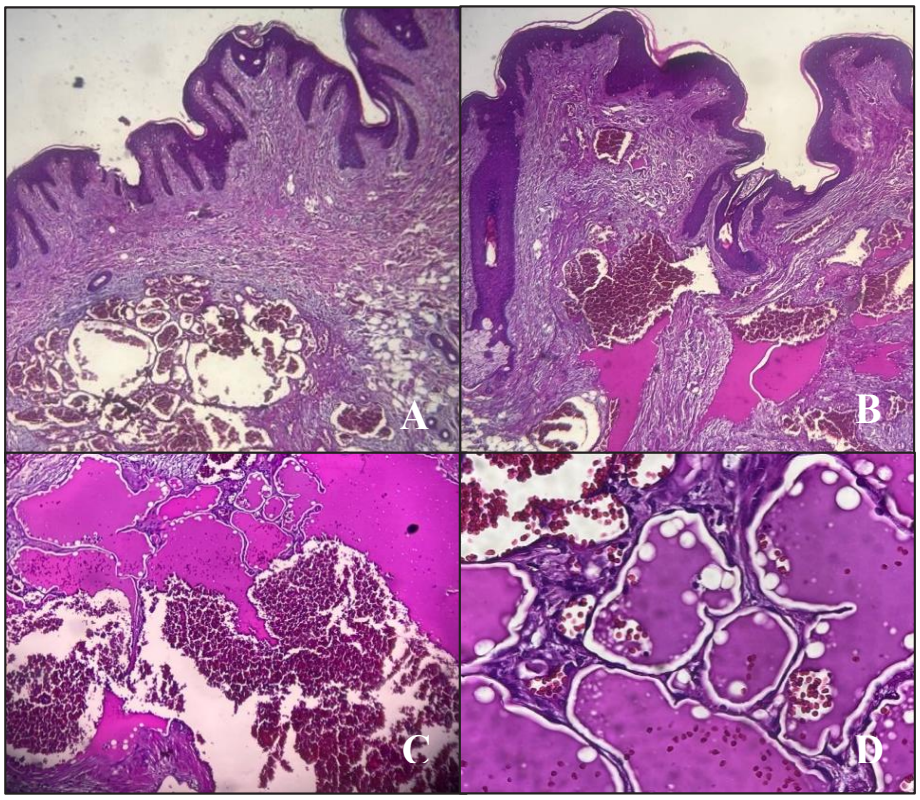

FIGURE 3. (A) H & E stained section showing lymph vessels containing red blood cells (Magnification 10x) (B) H & E stained section showing numerous vessels spaces which are filled with either blood cells or lymphatic fluids (Magnification 10x) (C) and (D) Cystically dilated spaces filled with lymph and red blood cells

Based on the clinical presentation, physical examination, dermoscopic findings, histopathology, and immunohistochemical examination, the final diagnosis was HLA. The patient then underwent surgical excision of the lesion under local anesthesia using an elliptical excision technique. The lesion was removed with a 2-mm margin of clinically normal tissue. The excised specimen was sent for histopathological evaluation, which confirmed clear surgical margins. The postoperative course was uneventful, and no complications such as bleeding, infection, or delayed wound healing were noted. On follow-up at 3 months post-surgery, there was no evidence of recurrence, and dermatological examination revealed a linear postoperative scar in the left inner thigh (FIGURE 5).

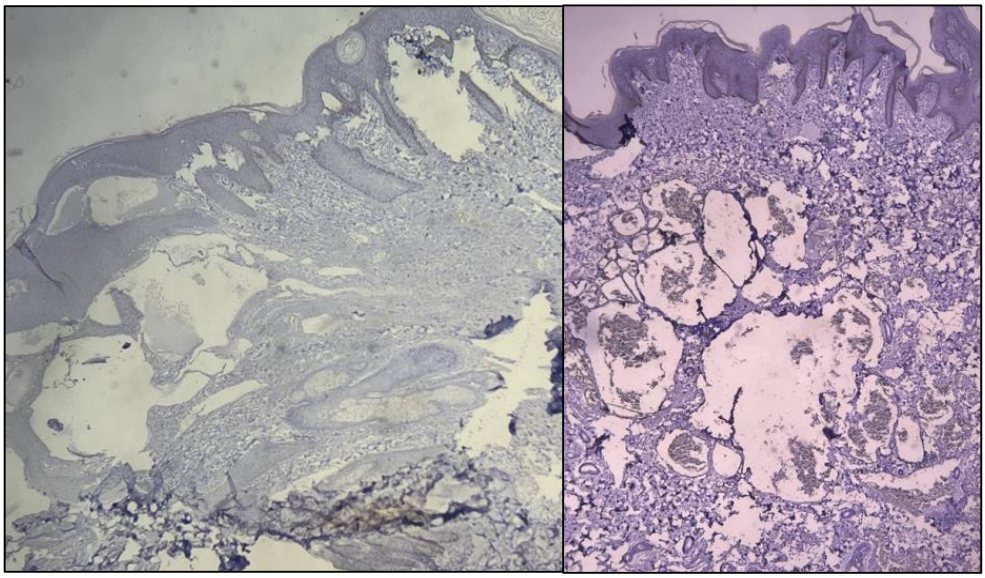

FIGURE 4. Immunohistochemical reaction showed focal positivity for D2-40

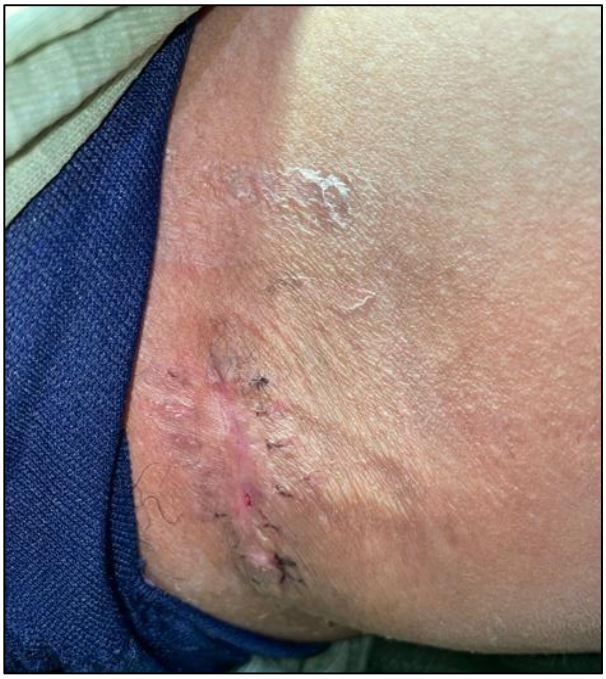

FIGURE 5. Linear postoperative scar in the left inner thigh on follow-up at 3 months post-surgery

DISCUSSION

Definition

Vascular malformations are classified based on the predominant type of vessels involved, such as arterial, arteriovenous, venous, capillary, or lymphatic malformations. Malformations involving a combination of these vessels, such as congenital lymphangiomas or other lymphatic anomalies, represent proliferations of abnormal lymphatic channels.[9] When lymphangiomas are filled with blood, they are referred as HLA, which is an extremely rare entity.[5]

Etiology and pathogenesis

The etiology and pathogenesis of HLA are not fully understood, but it is hypothesized to originate from abnormal embryological development, particularly related to the jugular lymphatic sacs and the failure of venolymphatic connections to regress during fetal development.[10] The simultaneous proliferation of both lymphatic and blood vessels may be explained by their origin from a common mesenchymal precursor.[11] Additional contributing factors, such as trauma, surgery, or hormonal changes during pregnancy, may exacerbate the growth of these malformations.[7]

Differential diagnosis

Hemangiolymphangioma may mimic other lesions based on clinical presentation and histologically.[5] The clinical presentation of HLA, especially in atypical anatomical sites such as the medial thigh, poses a diagnostic challenge. The differential diagnosis includes hemangioma, dermoid cyst, condyloma acuminata, and neurofibroma.[12] From a histopathological perspective, important differential diagnoses include hemangioma, lymphangioma, and lymphangiosarcoma.[13,14] Needle aspiration or biopsy can also be useful in differentiating hemangiolymphangioma from other fluid-filled masses.[12] Hemangiomas typically appear shortly after birth, grow rapidly, and then involute over time, distinguishing them from HLA.[3] Lymphangioma is a benign congenital malformation of lymphatic vessels, typically presenting in early childhood as lobular masses or cystic lesions, and is histologically distinguished from HLA by the absence of blood-filled spaces.[15,16] Lymphangiosarcoma is a rare, aggressive malignancy of lymphatic endothelium that typically arises in chronically lymphedematous limbs, often following surgery or radiotherapy, and is characterized by poor prognosis and distinct histopathological features.[17,18]

Histopathological features

Histologically, HLA consists of dense fibrous tissue that grows in bands between the numerous dilated vascular spaces and invades the subcutaneous fat. These spaces may contain either lymphatic fluid or red blood cells, depending on the predominance of lymphatic or blood vessel components.[14] Immunohistochemical markers play a crucial role in confirming vascular malformations, with D2-40 serving as a marker for lymphatic endothelium and CD31/CD34 for blood vessel identification.[5] Notably, lymphatic endothelial cells may also exhibit CD34 positivity.[7]. Recent literature has highlighted additional markers, such as Prox-1 and VEGFR3, for the identification of aberrant lymphatic vessels.[5] In our case, the diagnosis was confirmed with H&E staining and supported by immunohistochemistry, which revealed positive D2-40 staining in lymphatic endothelial cells.

Diagnostic utility of dermoscopy

Dermoscopy has been shown to facilitate the clinical recognition of skin tumors, especially with vascular structures.[19] Dermoscopy was a valuable non-invasive diagnostic tool in this case, providing enhanced visualization of subclinical vascular structures that are not readily apparent on routine clinical examination. Unlike standard inspection or palpation, dermoscopy allows the identification of characteristic vascular patterns, such as lacunae, red-blue coloration, or mixed vascular structures, which may suggest a diagnosis of vascular malformations, including HLA.[19] In this patient, dermoscopic findings supported a vascular etiology and guided the decision to proceed with histopathological and immunohistochemical confirmation. This aligns with previous studies that emphasize dermoscopy's utility in differentiating benign vascular lesions from other cutaneous nodules or malignancies, thereby improving diagnostic accuracy and reducing unnecessary interventions.[20]

Management and prognosis

Management of HLA depends on lesion size, location, and involvement of vital structures. Therapeutic options include laser therapy, embolization, sclerotherapy, electrocautery, radium implantation, cryotherapy, or complete surgical excision.[7] Surgical excision is considered the most definitive and effective treatment, particularly when the lesion is well-circumscribed and does not involve critical anatomical structures.[2] Electrocautery and cryotherapy may be considered for smaller or superficial lesions, particularly when surgery is contraindicated. Incomplete resection is associated with recurrence rates up to 100%, whereas complete excision leads to favorable outcomes.[7] Despite a generally benign course and rare malignant transformation, long-term follow-up is essential due to recurrence rates reported up to 21%.[4]

CONCLUSION

Hemangiolymphangioma is a rare vascular malformation involving both blood and lymphatic vessels. This report presents an HLA occurring in the left inner thigh of a 27-year-old female patient, representing an unusual anatomical location for this type of vascular malformation. The case emphasizes the importance of a comprehensive diagnostic approach combining clinical evaluation, dermoscopic assessment, and histopathological confirmation. From a clinical perspective, this report aims to enhance awareness among practitioners of the diverse presentations of HLA. It also emphasizes the importance of early recognition and complete surgical excision to minimize recurrence and improve patient outcomes.

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