Case Report

Angiomyomatous Hamartoma; A True Lesion or a Vascular Compensatory Hyperplasia with Muscularization? : An Interesting Case Report

Rakesh Kumar Gupta[1], Kavita Gaur[1], R K Jindal[3], Ravindra Kumar Saran[1]*, Sachin Agrawal[2]

[1]Department of Pathology, G B Pant Institute of Postgraduate Medical Education and Research, Delhi, India
[2]Department of Radiology, G B Pant Institute of Postgraduate Medical Education and Research, Delhi, India
[3]Department of Surgery, Maulana Azad Medical College, New Delhi, India

Keywords: *Angiomyomatous, Hamartoma, Lymph Node, Lymphedema.*

ABSTRACT

Angiomyomatous hamartoma is a rare entity of unknown etiology preferentially involving lymph nodes of the inguinal region. We report a case of 20-year-old young male presenting with unilateral lower limb lymphedema. Doppler study of the limb revealed venous flow in the node. Histopathology of the lymph node showed a peculiar replacement of normal hilar anatomy by a haphazard conglomerate of thick walled vessels containing red blood cells and lymphocytes. The present report explores the pathogenesis of angiomyomatous hamartoma with review of literature.

***Corresponding author:**
Dr. Ravindra Kumar Saran, MD, DNB (Pathology) Professor, Dept of Pathology, Academic Block, G B Pant Institute of Post Graduate Medical Education and Research, Jawaharlal Nehru Marg, New Delhi -110002. INDIA.
Phone: +91 97185 99074
E-mail: ravindraksaran@hotmail.com

Introduction

Angiomyomatous hamartoma (AMH) was first described by Chan et al in 1992.[1] It is a rare entity primarily involving inguinal lymph nodes and characterized by replacement of lymphoid follicles by haphazardly arranged combination of smooth muscle bundles, fibrous tissue and thick walled blood vessels. Other less commonly involved lymph nodes are popliteal, cervical, and sub-mandibular. [2-4] Allen et al [5] first described the presence of an adipose tissue component in AMH, while Magro et al [6] termed such variant as an angiomyolipomatous hamartoma (AMLH).

Case report

A 20-year-old young male presented to our hospital with the chief complaints of recurrent subcutaneous swelling of the left lower limb and mild pain in both the limbs since six years. The frequency of these episodes gradually increased over time. No history of injury or filariasis was documented. The patient is a professional tailor and uses his right leg vigorously to propel the footpad of his sewing machine. On examination, bilateral superficial inguinal lymph nodes were palpable, non-tender, and firm in consistency. Right sided lymph node was solitary and measured 4x3 cm; while left sided node measured 1.5x1 cm.Varicose veins were absent. A fine needle aspiration was attempted which yielded inadequate material for any conclusive remark. The colour doppler revealed enlarged lymph nodes with thickened capsule displaying venous flow within the nodes (Fig 1). Bilateral femoral vessels were patent and showed normal flow. As the right inguinal nodes were much larger in comparison to the left, excision was planned on right side.

Pathological findings: A lymph nodal specimen measuring 3x1.5x0.4 cm was received. Externally the capsule was intact and thickened. The cut-section was firm and fibrosed. Microscopy showed thickening of lymph node capsule and replacement of the hilum by a disorganized mass comprising of vascular channels with thick smooth muscle bundles traversing haphazardly in fibro-adipose tissue. Both RBCs and lymphocytes were noted in the lumina of vessels. These vessels were interpreted as abnormal thick walled lymphatic afferent channels. Normal calibre veins and thick walled veins were also noted along with many capillaries. Fibrosis was extending into the peripheral cortex with few remnant follicles in the sub-capsular region (Fig 2). No necrosis, granulomas or parasite were found. CD34 and HMB-45 immunohistochemistry for assessing number of pericytes and to rule out angiomyolipoma respectively were non-contributory. CD31 and smooth muscle actin (SMA) were positive in endothelial cells, and smooth muscle cells of the abnormal vessels respectively (Fig 3). The case was reported as hyperplastic muscularization of large afferent lymphatics of inguinal node secondary to recurrent lower limb edema with ultrasound doppler evidence of veno-lymphatic anastamosis.

Discussion

AMH is a rare benign entity of unknown aetiology. Till date about 35 cases have been reported in medical literature. Earlier cases of inguinal node AMH with concomitant lymphedema of ipsilateral limb have been reported. [7, 8] Bourgeois et al [7] suggested that the destruction of nodal sinuses by vascular and stromal alterations may represent impairment of the lymphatic flow and contribute to the development of lymphedema. However, Sakurai et al [9] hypothesized that impaired lymphatic flow is not just a consequence of hamartoma but may actually play a role in its pathogenesis. We also support this hypothesis as in our case due to higher levels of physical activity in the right limb for occupational reasons this may have led to an increased overall venous and lymphatic drainage. Consequently, right side inguinal lymph nodes were markedly enlarged to compensate the flow, resulting in less edema of the right leg. Channer et al postulated that a haphazard smooth muscle component within the hilum results from the reparative reaction to inflammation which was later supported by Chan et al. [1, 10] Kim et al [11] in their case reported a popliteal nodal AMH in association with pigmented villonodular synovitis (PVNS) and concluded that macrophages, cytokines and growth factors released from the PVNS lesion reached the adjacent popliteal lymph nodes through the draining lymphatics leading to the development of AMH. However, none of the aforementioned case reports described any evidence of inflammatory reaction within the lymph nodes. We too have not found inflammatory reaction within the lesion. In this case the colour doppler showed prominent dilated

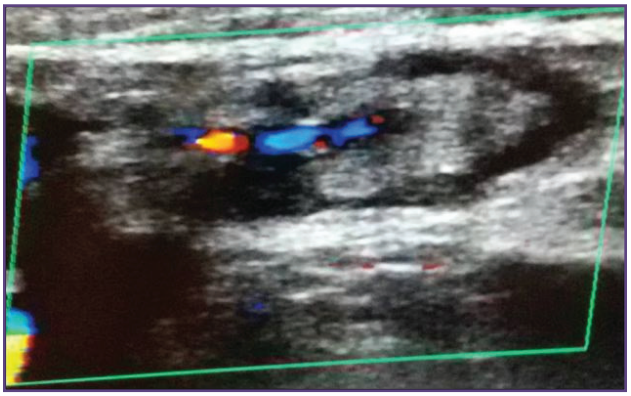

Fig. 1: Colour doppler image showing a large lymph node measuring 3x1.5 cm with echogenic centre and cluster of color signals in the hilum of the node.

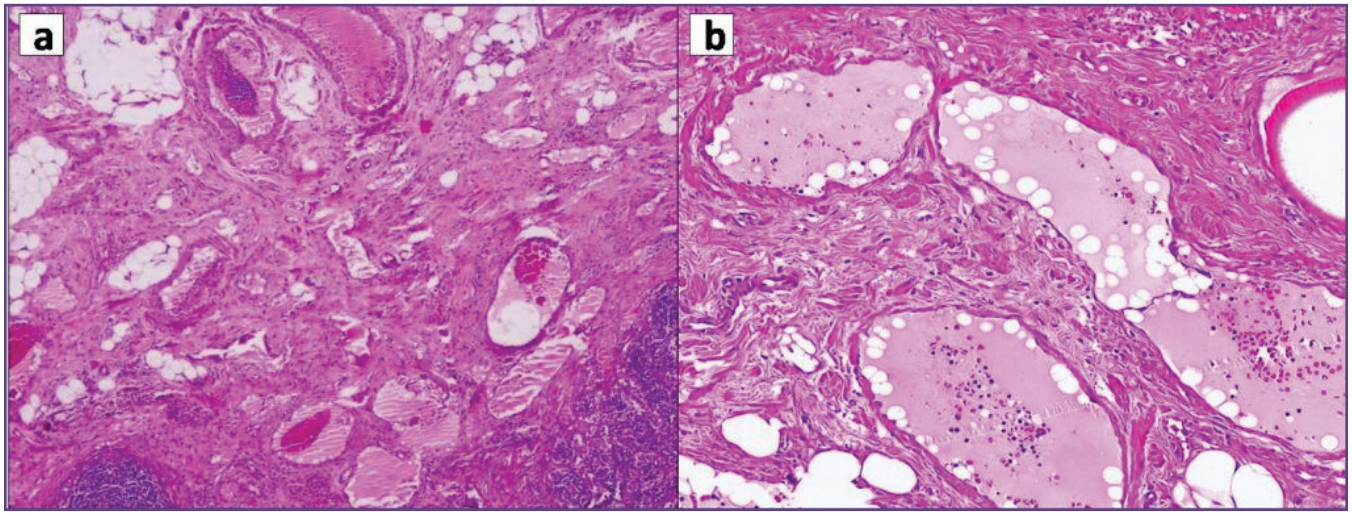

Fig. 2: Photomicrographs of the lymph node biopsy showing compressed peripheral remnant lymphoid follicles and expanded hilum with numerous haphazardly arranged dilated and thickened lympho-vascular channels surrounded by smooth muscle bundles, fibrosis and adipose tissue (a HEx40), high power image of the hilar area showing dilated tortuous vessels filled with both RBCs and lymphocytes (b HEx100).

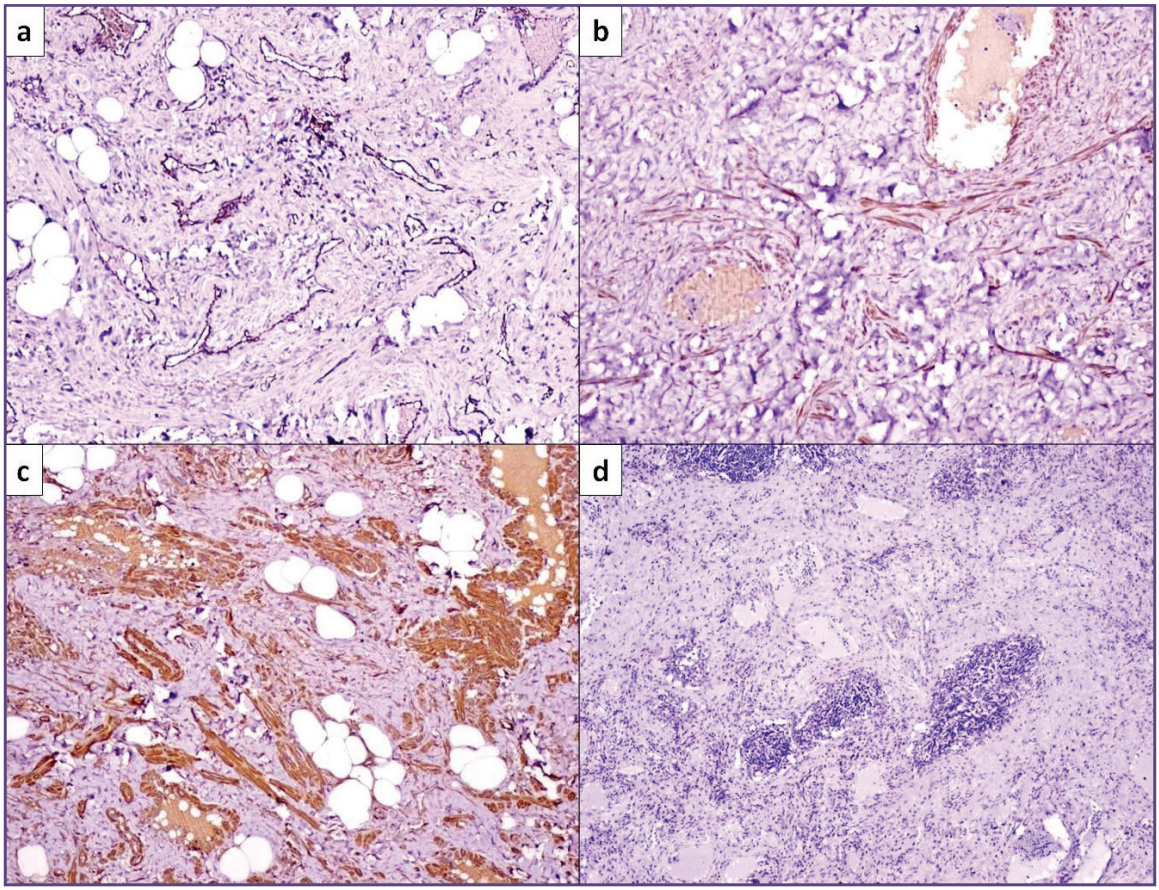

Fig 3: Immunohistochemistry images showing. a: CD31 positivity in the endothelial cells of the capillaries and small vascular channels, b: Desmin stain highlighting traversing thick smooth muscle bundles, c: SMA stain showing positivity in both smooth muscle and endothelial cells, d: HMB-45 stain negativity in all the components.

vasculature with increased flow within the node which on histopathology revealed dilated haphazard muscularized afferent lymphatics, likely causing lymphedema.

Adipose tissue within AMH may represent an associated additional hamartomatous component or result from metaplastic change.

Lymphangiomyomatosis usually involve lungs but may also rarely involve lymph nodes. Angiomyolipoma (AML) usually involves retroperitoneal lymph nodes, which should be differentiated from AMH. [12] In comparison to AMH, the smooth muscle cells show positive immunostaining for HMB-45 in lymphangiomyomatosis and AML. Surgical resection is the best treatment and till date no recurrence has been reported. Though earlier reports have not highlighted the exact clinical significance, AMH should always be considered as a remote possibility causing inguinal swelling with or without lymphedema.

To conclude, it is difficult to put this case in a watertight category of AMH. This lesion might result from physiological compensatory changes in lymphatic flow. The resultant pathology is same and accompanied with alteration in lympho-vascular flow, compensatory hyperplasia and muscularization of hilar lymphatic vessels. Present knowledge and literature review did not support clearly one or the other theory. The pathway of draining channels and changes thereafter acknowledged both the view points and hence it should not be put in one category.

Acknowledgement
We would like to acknowledge Miss Dolly for technical help in immunohistochemistry.

Funding
None

Competing Interests
Not declared

References
1. Chan JK, Frizzera G, Fletcher CD and Rosai J. Primary vascular tumors of lymph nodes other than Kaposi's sarcoma. Analysis of 39 cases and delineation of two new entities. Am J Surg Pathol. 1992;16:335-350.
2. Ghosh P, Saha K, Ghosh AK. Vascular transformation of bilateral cervical lymph node sinuses: a rare entity masquerading as tumor recurrence. J Maxillofac Oral Surg. 2015;14:397-400.
3. Prusac IK, Juric I, Lamovec J, Culic V. Angiomyomatous hamartoma of the popliteal lymph nodes in a patient with Klippel-Trenaunay syndrome: case report. Fetal Pediatr Pathol. 2011;30:320-4.
4. Barzilai G, Schindler Y, Cohen-Kerem R. Angiomyomatous hamartoma in a submandibular lymph node: a case report. Ear Nose Throat J. 2009;88:831-2.
5. Allen PW, Hoffman GJ. Fat in angiomyomatous hamartoma of lymph node. Am J Surg Pathol. 1993;17:748-749.
6. Magro G, Grasso S. Angiomyomatous hamartoma of the lymph node: case report with adipose tissue component. Gen Diagn Pathol. 1997;143:247-249.
7. Bourgeois P, Dargent JL, Larsimont D, et al. Lymphoscintigraphy in angiomyomatous hamartomas and primary lower limb lymphedema. Clin Nucl Med. 2009;34:405-9.
8. Piedimonte A, De Nictolis M, Lorenzini P, Sperti V, Bertani A. Angiomyomatous hamartoma of inguinal lymph nodes. Plast Reconstr Surg. 2006;117:714-6.
9. Sakurai Y, Shoji M, Matsubara T, et al. Angiomyomatous hamartoma and associated stromal lesions in the right inguinal lymph node: a case report. Pathol Int. 2000;50: 655-659.
10. Channer JL, Davies JD. Smooth muscle proliferation in the hilum of superficial lymph nodes. Virchows Arch A Pathol Anat Histopathol. 1985;406:261-70.
11. Hyun-Soo Kim, Ki Yong Na Jae-Hoon Lee, Nam Su Cho, Gou Young Kim, Sung-Jig Lim. Angiomyomatous hamartoma of popliteal lymph nodes occurring in association with diffuse pigmented villonodular synovitis of knee. The Korean Journal of Pathology. 2011;45:S58-61.
12. Dzombeta T, Francina M, Matković K, et al. Angiomyolipomatous hamartoma of the inguinal lymph node--report of two cases and literature review. In Vivo. 2012;26:459-62.