CASE REPORT
Volume - 9 | Issue - 7 | July - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA OF FOREARM MIMICKING SARCOMA: A DIAGNOSTIC CHALLENGE ON FROZEN SECTION

Pathology

Dr Vaishali Walke* Associate Prof, Pathology, Indira Gandhi Govt Medical College, Nagpur. Maharashtra India *Corresponding Author

Dr Sonali Datar Assistant Prof, Pathology, Indira Gandhi Govt Medical College, Nagpur. Maharashtra

Dr. Roshni Jacob Senior Resident, Pathology, Indira Gandhi Govt Medical College, Nagpur. Maharashtra

Dr Lubhna Ansari Senior Resident, Pathology, Indira Gandhi Govt Medical College, Nagpur. Maharashtra

ABSTRACT

Angio-lymphoid hyperplasia with eosinophila (ALHE) is a benign vascular proliferation clinically characterized by single or multiple reddish-brown, nodules occurring predominantly in head and neck region in young adults. We report a case of a 30 year young male who presented with a large forearm mass of one year duration. The clinical impression was soft tissue sarcoma. Wide local resection was planned and specimen received for intra operative frozen section diagnosis and to comment on resection margins. The diagnosis offered was benign vascular neoplasm with uninvolved surgical margins. The subsequent permanent sections of excised specimen revealed, proliferation of small blood vessels lined by endothelial cells with a histiocytoid morphology protruding into the lumen. Numerous eosinophils and lymphocytes present surrounding the blood vessel in stroma. The diagnosis of Angiolyomphoid Hyperplasia with Eosinophilis was offered. Here we discuss the unusual clinical presentation and diagnostic challenges faced in intraoperative consultation of this common entity.

KEYWORDS

Angiolymphoid Hyperplasia with Eosinophilia, epitheloid hemangioma, soft tissue tumor

INTRODUCTION:

The term angiolymphoid hyperplasia with eosinophilia known for variety of synonyms as epitheloid hemangioma, inflammatory angiomatous nodule, atypical granuloma, pseudo pyogenic granuloma and histiocytoid hemangioma was first coined by Wells and Whimster. It is an uncommon, benign to reactive vaso-proliferative disease presenting as painless, vascular nodules in the dermal and subcutaneous tissues of head and neck, particularly around the ear. [1] It has also been reported in scalp, lip, orbit, conjunctiva, extremities and colon. [2] It is superficial in nature; however cases with muscular and bony involvement are reported. [3] There is predilection for women although a male preponderance has been noted in selected Asian studies. ALHE is seen most commonly in Asians followed by Caucasians and presents commonly in patients aged 20-50 years with mean age of onset of 30-33 years. [4] ALHE may persist for years but serious complications like malignant transformation have never been reported. [5] On histopatholgy it is characterized by presence of a florid proliferation of blood vessels lined by plump histiocytoid endothelial cells admixed with dense inflammatory infiltrate comprising of lymphocytes, eosinophils and plasma cells. [4] Lesions are typically small ranging from 0.5 - 3 cm with larger size being a rare manifestation. [5] We report an unusually large mass clinically simulating sarcoma due to deeper involvement and the diagnostic challenges on intraoperative consultation.

CASE PRESENTATION:

A 30 year young male presented to surgery clinic with a gradually increasing, large, painless mass on right forearm just below the antecubital fossa of one year duration. There was no history of similar complains in past. On local examination, the mass was 10x8cm, non tender, firm, and immobile. The overlying skin was normal with no obvious discoloration, ulceration, crusting. No regional lymphadenopathy was noted. MRI showed involvement of brachioradialis muscle and brachial artery. Clinico radiological diagnosis was soft tissue sarcoma. Wide local excision was planned with intraoperative consultation for confirmation of diagnosis and to comment on resection margins. The surgical margins were free from tumor infiltration and frozen sections from tumor revealed proliferation of vascular channels separated by variable amount of stroma. (Figure 1) The vessels are medium sized thick walled and the surrounding stroma shows inflammatory cells. (Figure 2) The intra operative diagnosis of benign vascular neoplasm with clear margins was offered. The subsequent excised specimen measured 10x8x5.5cm, externally capsulated, multinodular, greyish white. On cutting, variable size nodules ranging from 1x1cm to 4x3cm, soft to firm, yellowish with central hemorrhagic areas were noted. (Figure 3) The tumor displayed characteristic two major components: proliferating thick walled blood vessels and inflammatory component. (Figure 4) These vessels were lined by plump endothelial cells showed hobnailing and cytological features of histiocytes. (Figure 5) The surrounding stroma contained plenty of eosinophils admixed with lymphocytes and plasma cell. Focally lymphoid collection was noted. The involvement of adjacent muscle and fat tissue was evident. (Figure 6) The diagnosis offered on permanent section was Angiolymphoid hyperplasia with eosinophilia, in contrast to the clinical suspicion of malignancy.

DISCUSSION:

Many hypotheses regarding the etiology of Angiolymphoid hyperplasia exist. The main contention behind etiology remains controversial as to whether it's a neoplastic or atopic hypersensitivity reaction (unusual reactive process) with the latter receiving more support. [6] Several reports indicate that ALHE may be secondary to complex immunological mechanism, trauma, infection or insect bite; however some authors consider that arterio-venous shunt is the main etiopathogenetic mechanism observed in 42% of cases. [7] It is a disfiguring lesion characterized by isolated or small numbers of fleshy to plum colored papules predominantly on the head (81%), the most common locations being periauricular area (36.3%), face (28.2%) and scalp (17.3%). [1,2] Other locations such as trunk, lower extremities, hands, penis, oral mucosa and colon are rare but reported. [8] The diameters quoted in the literature range from 1-10 cm with larger lesions being extremely rare. [9] Our patient presented with a huge mass to the extent that clinician thought of a possibility of malignancy which was substantiated by radiology revealing involvement of the underlying muscle and artery. In such situation it was intra operative consultation that helped to narrow down the diagnosis. We were able to delineate the tumor as benign vascular neoplasm; however we could not offer a definitive diagnosis on frozen section. The challenges we faced are thick sections that can be attributed to the fat rich tissue submitted and rapid H&E stain. The staining may be inadequate due to thick quality sections and hence might not have picked up the granules of eosinophils, and therefore we missed this one of the important component essential for diagnosis. These are some the limitations of frozen section study as quoted by previous authors. [10,11] However histopathology remains the mainstay of diagnosis and is characterized by branching vessels with protuberant endothelial cells resembling histiocytes, lymphocyticinfiltrate and numerous eosinophils. These histiocytoid endothelial cells show abundant eosinophilic or clear cytoplasm with vesicular nuclei. The cells are mostly cuboidal with occasional hobnailing which is related to the presence of cytoplasmic vacuoles in these cells causing cytoplasmic protrusion into the lumina. The proportions of these three components vary from case to case. In some instances, vessels predominate and are embedded in a sclerotic or myxoid stroma associated with mild lymphocytic infiltrate and few eosinophils. In others, all three components maybe found but lymphocytic infiltrate is dense with germinal center formation. [12,13] The

main differential on histopathology is Kimura disease whose typical presentation is of large subcutaneous mass in periauricular or submandibular region. It's a systemic immune mediated inflammatory process that presents with lymphadenopathy, eosinophilia, and increased serum level of IgE and associated with renal disease.[13] Histologically it is characterized by florid lymphoid hyperplasia with germinal centre formation, surrounded by eosinophilic infiltrates and microabscess and arborizing vascular proliferation.[14] The other differential can be, epitheloid hemangio endothelioma which displays cords of endothelial cells having intracytoplasmic neolumina in a myxoid matrix with absence of lymphoid aggregates. The tumor cells show nucleomegaly and pleomorphism. [13] The rare differential can be angiosarcoma which displays anastomosing vascular channels lined by tumor cells showing obvious malignant features the common sites being head and neck followed by liver, breast. [13,15] Surgical excision is the most effective treatment for ALHE and when excision is complete, recurrences are rare. [16]

CONCLUSION:

The Relevance Of This Case Is Due To Rare Clinical Presentation As Huge Soft Tissue Mass That Mimics Neoplastic Process And The Difficulties Encountered While Interpreting Frozen Sections. The Knowledge And Awareness About These Challenges Will Help Pathologist To Arrive At A Definitive Diagnosis So That Treatment Can Be Planned Accordingly.

Images:

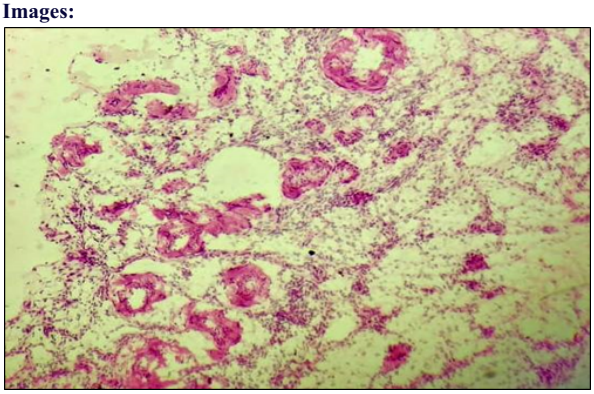

Figure 1: Frozen section: vascular proliferation consisting of thick walled blood vessels surrounded by loose oedematous stroma (H&E, 10X)

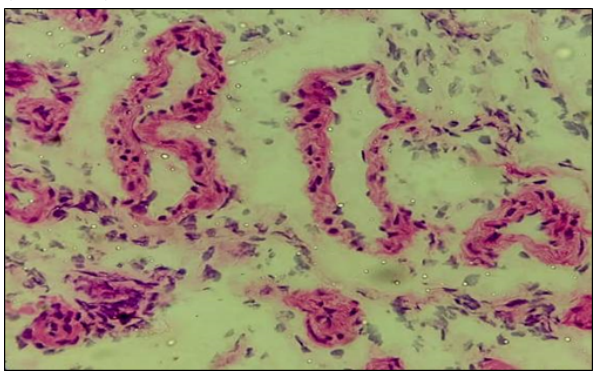

Figure 2: Frozen section: Blood vessels lined by plump endothelial cells surrounding stroma showing scanty inflammatory cells (H&E, 40X)

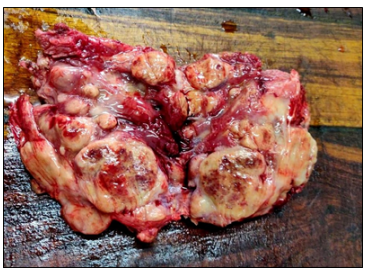

Figure 3: Irregular, multinodular, soft, yellowish white mass with hemorrhagic areas. (gross)

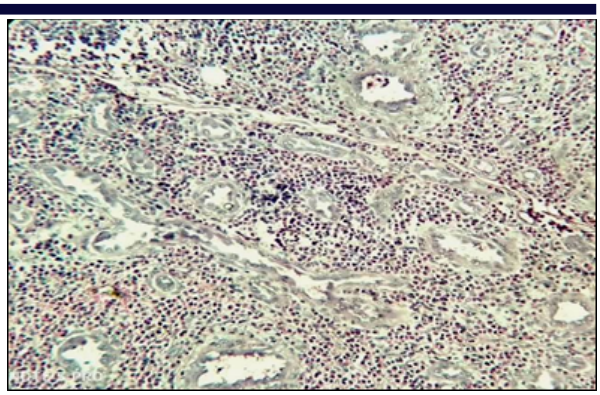

Figure 4: Proliferating medium sized thick walled vascular channels surrounded by dense inflammation (HP; H&E, 10X)

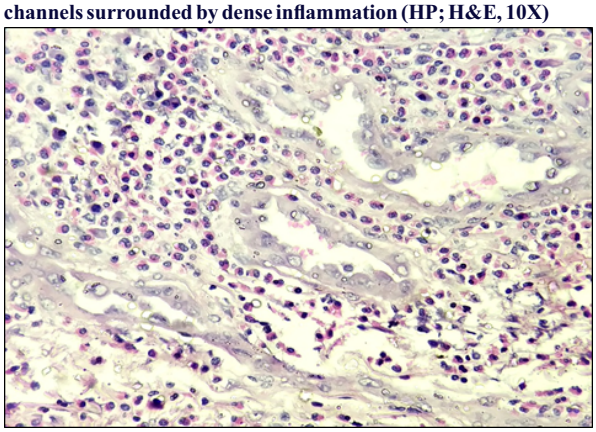

Figure 5: Blood vessels are lined by plump endothelial cells showing hobnailing and the stroma showing dense inflammation rich in eosinophils (HP; H&E, 40X)

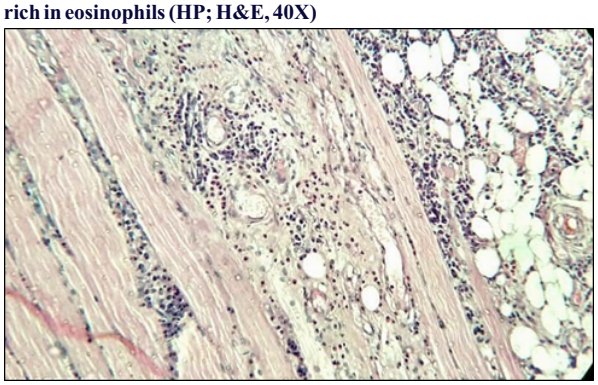

Figure 6: Involvement of underlying muscle and fat is also evident (HP; H&E,10X)

REFERENCES:

1. Wells GC, Whimster IW. Subcutaneous angiolymphoid hyperplasia with eosinophilia. Br J Dermatol. 1969;81(1):1.14.
2. Kurihara Y, Inoue H, Kiryu H, Furue M. Epitheloid hemangioma (angiolymphoid hyperplasia with eosinophilia) in zosteriform distribution. Indian J Dermatol. 2012;57(5):401-03.
3. Vitantonio HD, Paulis DD, Ricci A, Raysi SD, Marzi S, Maestro MD et al. Angiolymphoid hyperplasia with eosinophilia and entrapment of ulnar nerve. Surg Neurol Int. 2016;7(5):160-63.
4. Taylor SK, Meyerle JH, Glusac EJ. Angiolymphoid hyperplasia with eosinophilia (e Medicine Website) December 11, 2008. Available from: 2008.
5. Al-Muharraqi MA, Faqi MK, Uddin F, Ladak K and Darwish A. Angiolymphoid hyperplasia with eosinophilia (epitheloid hemangioma) of face: An unusual presentation. 2011;2(8):258-60.
6. Rajendran R, Padmakumar SK, Kothawar S, Balaraman Nm. Angiolymphoid hyperplasia with eosinophilia (ALHE). J Oral Maxillofac Pathol. 2005;9:24–26.
7. Doloi PK, Khanna S. Angiolymphoid hyperplasia with eosinophilia – A case report. Int J Otolaryngol and head and neck surgery. 2012;1:44-47.
8. 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(3):506-12.
9. Hussain JA, Zamel H Al and Nawaz B. Angiolymphoid hyperplasia with eosinophilia (epitheloid hemangioma) of the external auditory canal, an unusual presentation in an adult female: A case report. J Surg Case Rep. 2018(7):1-3.
10. Ahmad Z, Barakzai MA, Idrees R, Bhurgri Y. Corelation of intra- operative frozen section consultation with final diagnosis at a referral centre in Karachi, Pakistan. IJPM 2008;51:469-73
11. Chand P, Amit S, Gupta R, Agarwal A. Errors , limitations and pitfalls in the diagnosis of central and peripheral nervous system lesions in intra operative cytology and frozen

sections. J Cytol 2016;33:93-7.
12. Carter D, Greenson JK, Reuter VE, Stoler MH, Mills SE. Sternberg's Diagnostic Surgical Pathology. 6th ed. Philadelphia: Lippincott Williams & Wilkins; 2015.2928p
13. Guo R, Gavino AP, Angiolymphoid Hyperplasia with Eosinophilia. Archives of Pathology & Laboratory Medicine. 2015;139(5):683-86.
14. Suzuki H, Hatamochi A, Horie M, Suzuki T, Yamazaki S. A case of angiolymphoid hyperplasia with eosinophilia (ALHE) of upper lip. J Dermatol. 2005;32(12):991.95.
15. Wang L, Lao IW, Yu L and Wang J. Clinicopathological features and prognostic factors in angiosarcoma: A retrospective analysis of 200 patients from a single Chinese medical Institute. 2017;14(5):5370-78.
16. Santosa C, Wardhana M, Saputra H. Angiolymphoid hyperplasia with eosinophilia with clinical pictures of keratoacanthoma: A rare case report. Clin Case Rep. 2019;7(1):189-92.