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Case Series

Cutaneous Intravascular papillary endothelial hyperplasia – A case series

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ABSTRACT

Intravascular papillary endothelial hyperplasia (IPEH) is rare intravascular endothelial proliferation that may mimic an angiosarcoma. They are most commonly located in veins on head and neck, fingers and trunk and usually presents as a small superficial mass. It is now considered as an exuberant form of organising thrombus. We present here a series of cases of IPEH with varied clinical presentation and clinical diagnosis which on microscopy confirmed as Intravascular papillary endothelial hyperplasia

1. Introduction

Intravascular papillary endothelial hyperplasia (IPEH) comprises approximately 2% of the benign and malignant vascular tumors of the skin and subcutaneous tissues.[1] It was first described by Masson in 1923 as vegetant intravascular hemangioendothelioma.[2,3] He reported it as an atypical papillary endothelial proliferation in an ulcerated hemorrhoid, in a 68 year old man. He considered it to be a true neoplasm with degenerative changes like necrosis and thrombosis due to inadequate blood supply. In 1976, Clearkin and Enzinger termed it as "Intravascular Papillary Endothelial Hyperplasia". They suggested that the thrombosis takes place before the papillary proliferation and that the thrombotic material provides a matrix for its development.[4] Now it is considered as an exuberant form of organizing thrombus. Albrecht and Kahn studied IPEH immuno- histochemically and suggested that it is closely related to organizing thrombi and is probably a peculiar form of it.[5] We present here a series of cases diagnosed as IPEH.

2. Case Series

2.1. Case 1

A 40 year old female presented with swelling over the right arm for three months. There was no history of pain/redness over the swelling. On examination it was a cystic swelling with a clinical diagnosis of sebaceous cyst. Excision biopsy was done. On gross examination it was a nodular tissue measuring 1.5x1x0.5 cm. Histopathological examination (Figure 1) showed a lesion arising within the vascular lumen and is seen in close association to the organising thrombus within the vessel. The lesion is arranged in a papillary pattern predominantly with a central collagenized core along with few papillae with fibrin core surrounded by single layer of endothelial cells. There was no areas of necrosis, pleomorphism or high mitosis. The features were consistent with IPEH.

2.2. Case 2

A 34 year old female presented with swelling in the right submandibular region for 2 months with clinical diagnosis of lymphadenopathy/ salivary gland neoplasm. Fine needle

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aspiration was attempted from the swelling which yielded only hemorrhagic material. Excision biopsy was done and shows features consistent with IPEH with fibrin and collagenous cores surrounded by endothelial proliferation.

2.3. Case 3

A 39 year old male presented with pedunculated mass in the right posterior thigh with clinical diagnosis of fibroepithelial polyp/ pedunculated dermatofibrosarcoma protuberans. Excision biopsy was done and showed features of fibroepithelial polyp with dilated vessel within the polyp showing thrombus with papillary endothelial hyperplasia (Figure 2).

2.4. Case 4

A 53 year old female presented with swelling in the right thumb for 2 months with clinical diagnosis of dermoid cyst. Excision biopsy was done and showed features of IPEH with collagenous papillary cores surrounded by endothelial cells associated with fibrin thrombi and proliferation of endothelial cells with mild atypia and rare mitosis.

2.5. Case 5

A 47 year old female presented with anal tag with fissure in ano and anal polyp which was excised for histopathological examination and shows presence of hemorrhoids with fissure in ano. The dilated vessels within the hemorrhoids showed fibrin thrombi with features of ipeh showing fibrin cores surrounded by endothelial proliferation.

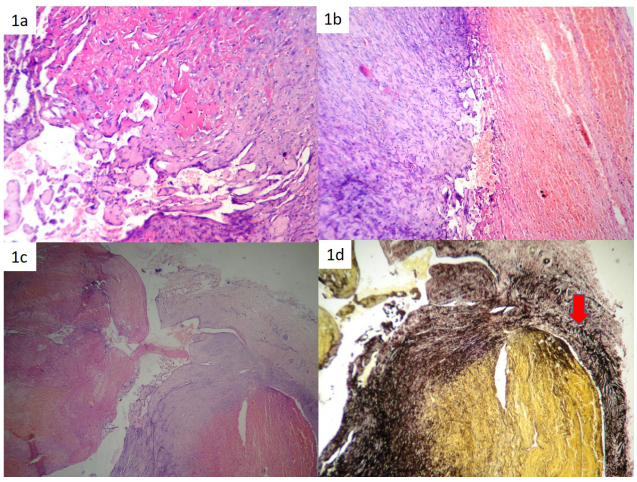

Figure 1: a: Histopathological examination from pure form showing the papillary structure with fibrin and collagen cores (H&E, 400X); b) Papillary structures seen in association with thrombus (H&E, 100X); **c,d:** The fibrocollagenous wall showing elastic lamina and elastic fibres (H&E, 400X); highlighted by Verhoeff-Van Gieson stain (VVG, 400X)

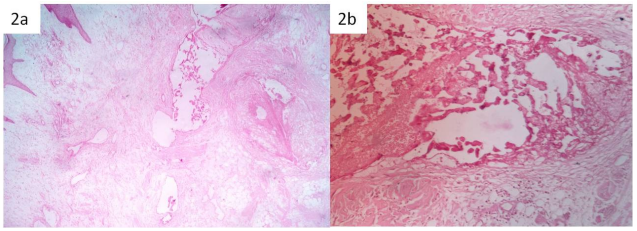

Figure 2: a: Fibroepithelial polyp showing IPEH in multiple vessels within the polyp (H&E, 100X); **b:** High power view showing the papillary structures with fibrin core (H&E, 400X)

3. Discussion

IPEH have been reported to be of three types (a) a primary, the pure form where changes are observed within a distended vessel; (b) a secondary, which is considered a secondary form that occurs in preexisting varices, hemangiomas, pyogenic granulomas, or lymphangiomas; and (c) an uncommon type in an extravascular location, in hematomas.[6] This series had three cases of primary forms within vessels and two secondary forms which were seen in dilated vessels of fibroepithelial polyp and hemorrhoids

Pure form is usually an intravascular lesion which is a usually small with an average size of two centimeters, and appears purple-red. Morphologically it appears as a multicystic mass containing clotted blood and surrounded by a fibrous pseudocapsule containing residual smooth muscle or elastic tissue of the preexisting vessel wall. They are most commonly located in veins on the head, neck, fingers, and trunk.[6] Few Case reports of IPEH are available involving head and neck, hand, skull, bladder, liver, uterus, brain and gastrointestinal tract.[1,7–11]

In this series the primary lesions presented as solitary swellings of an average of less than two centimeters as described in literature and all were cutaneous in origin involving head, arm, thigh, finger and anal region lesion.

The secondary forms are identified mostly only under microscopeand when it occur on a preexisting vascular lesion the ultimate prognosis depends on the underlying lesion. Rarely IPEH occurs extravascularly as a result of organization of a hematoma.[6] This study includes two cases of secondary forms which were picked up only on microscopic examination of a fibroepithelial polyp and hemorrhoids.

In early lesions, the endothelium grows along the contours of the thrombus and forms coarse papillae with fibrin cores. In the well-established lesion the papillae has a single layer of endothelium containing a collagenized core. This was observed even in our series, where the early lesions had predominantly fibrin core especially in secondary forms and the well established late lesions had predominantly collagenized core. Some residual fibrin thrombi could always be noted representing the evolution of the lesion

Table 1: Clinical andhistopathological details of cases

Case No.	Age (yrs)	Sex	Site	Clinical diagnosis	Gross findings	Microscopy
1.	40	Female	Right arm	Sebaceous cyst	1.5x1x0.5 cm nodular mass	Fibrin and collagenous core with endothelial proliferation. No atypia/ mitosis
2.	34	Female	Right submandibular	Lymph nde/ salivary gland neoplasm	1.8 x1.2 x 0.8 cms , nodular mass	Fibrin and collagenous core with endothelial proliferation. No atypia/ mitosis
3.	39	Female	Right thigh	Fibroepithelial polyp/ pedunculated dermatofibrosarcoma protuberans	4 x3.2 x1.6 cms , skin covered polypoidal mass	Fibrin core and endothelial proliferation. No atypia/ mitosis
4.	53	Female	Right thumb	Dermoid cyst	2 x1 x0.8 cms, nodular mass	Collagenous core with endothelial proliferation. Mild atypia, rare mitosis
5.	47	Female	Anal region	Anal polyp with fissue	1.4 x0.6x 0.5 cms , skin covered polyp	Fibrin core and endothelial proliferation. No atypia/ mitosis

from thrombus to early to well established forms. There was no evidence of pleomorphism, mitosis or necrosis in any of the cases; thus the close differential diagnosis of angiosarcoma was ruled out.

There are few studies on immunohistochemistry of IPEH supporting the hypothesis of its origin from organizing thrombus.[5,12] Akdur et al studied the histomorphological and immunohistochemical findings of ten IPEH cases. They found that all cases were intraluminal and intimately associated with the organizing thrombus, and all cases stained positive for CD31 and CD34. The staining with Factor VIII, Smooth Muscle Antigen, and type IV collagen, was variable.[12] Albrecht and Kahn observed similar immunophenotype of the endothelial cells in IPEH and organizing thrombi.[5]

The prognosis of this lesion is excellent and can be cured by simple excision in primary cases. In secondary forms the prognosis are related to the underlying lesion. In this series all the patients had good prognosis and was cured by simple excision.

4. Conclusion

IPEH is a rare benign vascular lesion which is now considered as a reactive process arising from an organizing thrombus which can be diagnosed mostly on histopathological examination. It requires adjuvant diagnostic work up only in rare cases especially in primary forms and should be differentiated from angiosarcoma which is a close differential diagnosis.

5. Source of Funding

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6. Conflict of Interest

None.

References

1. Tosios K, Koutlas IG, Papanikolaou SI. Intravascular papillary hyperplasia of the oral soft tissues: report of 18 cases and review of the literature. *J Oral Maxillofac Surg*. 1994;52(12):1263–8.
2. Garcia-Macias CM, Abad M, Alonso MJ, Flores T, Bullon A. Masson's vegetant intravascular haeman- gioendothelioma. *Acta Cytol*. 1990;34(2):175–8.
3. Masson P. Hémangioendothéliome végétant intravasculaire. *Bull Mem Soc Anat* . 1923;93:517–23.
4. Clearkin KP, Enzinger FM. Intravascular papillary endothelial hyperplasia. *Arch Pathol Lab Med*. 1976;100(8):441–4.
5. Albrecht S, Kahn HJ. Immunohistochemistry of intravascular papillary endothelial hyperplasia. *J Cutan Pathol*. 1990;17(1):16–21.
6. Weiss SW, Goldblum JR. Benign tumors and tumorlike lesions of blood vessels. In: Enzinger and Weiss 's Soft Tissue Tumors . 5th edn. St. Louis: Mosby; 2008. p. 668–71.
7. Tokyol Ç, Demir Y, Aktepe F. Cutaneous intravascular papillary endothelial hyperplasia of the hand: a case report and review of the literature. *Turk J Pediatr*. 2005;47(1):71–4.
8. Jazaerly T, Jaratli H, Sakr W, Almardini N, Urabi M, Dhar N, et al. Intravascular papillary endothelial hyperplasia of the bladder: Case report and review of the literature. *Avicenna J Med*. 2011;1(2):55–7.
9. Park KK, Won YS, Yang JY, Choi CS, Han KY. Intravascular Papillary Endothelial Hyperplasia (Masson tumor) of the Skull : Case Report and Literature. *J Korean Neurosurg Soc*. 2012;52(1):52–4.
10. Mohammadyari F, Dufan T, Ahmed YK, Dolati P. Masson Tumor of the Central Nervous System: A Case Report and Review of Literature. *Am J Case Rep*. 2022;23:937597. doi:10.12659/AJCR.937597.
11. Burguêz D, Dornelles LME, Serafini OA. Masson's Tumor- Case Report. *Oncol Cancer Case Rep*. 2021;7(10):1–2.
12. Akdur NC, Donmez M, Gozel S, Ustun H, Hucumenoglu S. Intravascular papillary endothelial hyperplasia: histomorphological and immunohistochemical features. *Diagn Pathol*. 2013;8:167. doi:10.1186/1746-1596-8-167.

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