

Case Report

Oral Hobnail Hemangioma: A Case Report

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Abstract

Hobnail hemangioma is a benign vascular lesion and intraoral of the lesions are relatively rare. Histopathologically, it shows distinctive biphasic pattern with vascular channels lined by endothelial cells showing characteristic "hobnail" cytomorphology. Since hobnail hemangioma shares similar clinical and histopathologic features with many other benign and malignant vascular tumors, accurate diagnosis is mandatory for proper treatment and prognosis. In the present study, emphasis was given to discuss the differential diagnosis and delineate the hobnail hemangioma from other vascular lesions. Further, immunohistochemical study was performed which showed strong immunopositivity for CD31 and factor VIII in all endothelial cells lining the vascular channels. CD34 was moderately immunopositive and vascular endothelial growth factor was negative.

Keywords: Dabska tumor, epitheloid hemangioendothelioma, hobnail hemangioma, Kaposi sarcoma, retiform hemangioendothelioma

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Introduction

Hobnail hemangioma is a benign vascular lesion. Originally it was called as "targetoid hemosiderotic hemangioma" (THH). THH is a clinical entity referring to the presence of ecchymotic halo surrounding a violaceous papule and shows a targetoid aspect on clinical examination. Further, expanded clinicopathologic studies have shown clearly that most vascular lesions showing histopathologic features of this distinctive neoplasm lack the characteristic clinical features of THH. Also, similar hobnail endothelial cytomorphology is observed in retiform hemangioendothelioma, patch stage Kaposi sarcoma, benign lymphangioendothelioma, Dabska tumor, and well-differentiated angiosarcoma. Hence, the renaming was proposed based on the observation that many histopathologically diagnosed hobnail hemangioma cases did not present clinical targetoid appearance.^{1,2}

Hobnail hemangioma affects mainly adults with slight male predominance. Common anatomic locations involved are the extremities, trunk, and in rare cases head and neck region. Intraoral lesions are extremely rare and till date only three cases have been reported.³ The exact pathogenesis is unknown, but it is postulated that trauma plays a vital role in the pathogenesis.²

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Case Report

A 25-year-old man was presented with complaint of a growth in the lower front region of the mandible since two years which was associated with pain since the last five months. The growth was small initially and increased to the present size gradually. The pain was continuous, non-radiating, and aggravated on taking food as it was interfered while chewing. On extraoral examination, no abnormality was detected except palpable bilateral submandibular lymphadenopathy.

Intraoral examination revealed a growth in the gingiva which was sessile, exophytic, and extending from lower labial sulcus covering the alveolar mucosa between the central and lateral incisors. Superiorly, it extended above the incisal edge of the incisors. Mediolaterally, the lesion extended from mesial aspect of the right central incisor to distal aspect of the left canine region. The color was same as the adjacent mucosa, irregular in shape, with cauliflower-like appearance (Figure 1). On palpation, the superior aspect was tender and soft in consistency, and in the inferior aspect it was firm. Bleeding was noticed from the gingival sulcus.

Based on the history and clinical examination, a provisional diagnosis of pyogenic granuloma was made and differential diagnoses were listed which included peripheral cemento-ossifying fibroma, peripheral giant cell granuloma, inflammatory fibrous hyperplasia, and Kaposi sarcoma.

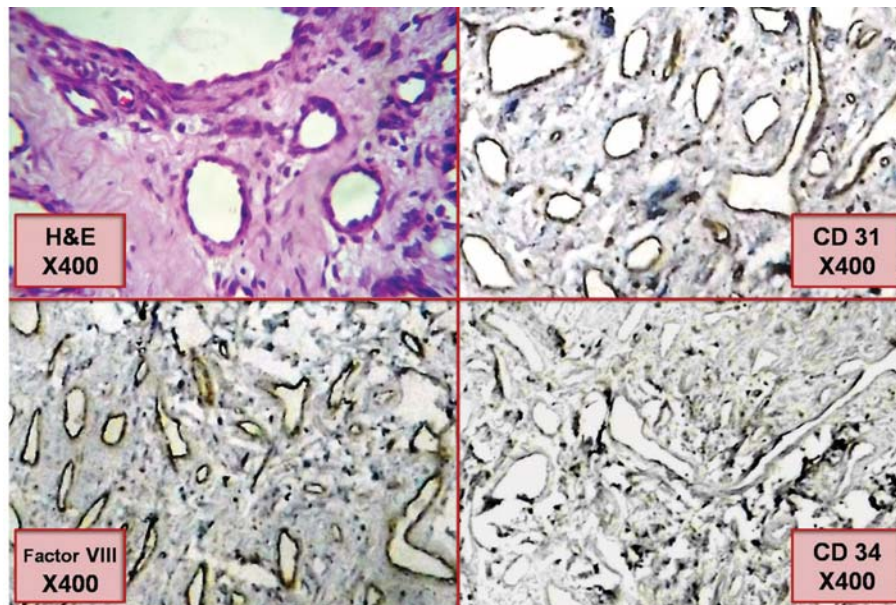
Investigations were carried out, which included intraoral periapical (IOPA) radiograph of the lower anteriors, complete hemogram, ELISA, and Biopsy. IOPA radiograph revealed an interdental bone loss between the right central and the left central incisors up to middle one-third of the root. Blood examination revealed a normal picture with nonreactivity in HIV₁ and HIV₂.

On histopathologic examination, the tissue section showed stratified squamous epithelium overlying vascular stroma. The epithelium was parakeratinized stratified squamous epithelium. Juxtaepithelially, the stromal tissue showed numerous irregular,

Table1. Specification of the antibodies used

Antibody	Clone	Isotype	Immunogen	Source	Antigenic retrieval	Protocol
CD31	JC/70A	IgG 1, kappa	Spleen membrane from a patient with hairy cell leukemia	Mouse	Citra	Overnight
CD34	Q Bend/10	IgG 1	CD34 isolated from human placental endothelial cell	Mouse	Citra	30 minutes
Factor VIII	BGX016A	IgG1, kappa	Purified human factor VIII	Mouse	Citra plus	2 hours
VEGF	Polyclone	N/A	Human recombinant VEGF 165	Rabbit	Citra	30 minutes

N/A: Not Applicable

**Figure 1.** The exophytic growth in the gingiva extending from the lower labial sulcus to lingual sulcus.**Figure 2.** Photomicrograph showing irregular vascular spaces lined by a single layer of endothelial cells with hyperchromatic nuclei protruding into the lumen (hobnail cells) (H&E X 400). Also endothelial cells are intensely immunopositive for CD31 and factor VIII, and moderately immunopositive for CD34 (X 400).

thin-walled vascular channels lined by a single layer of endothelial cells. The endothelial cells appeared to be protruding into the lumen of vascular spaces (hobnail appearance) (Figure 2). In the deeper part of the stroma, vascular spaces were slit-like and angulated and appeared to dissect through the collagen bundles. The stroma also showed chronic inflammatory infiltrate, and extravasated red blood cells. Further, we performed immunohistochemical study with CD31, CD34, factor VIII, and vascular endothelial growth factor (VEGF) (specifications of antibodies are listed in

Table 1). Immunohistochemistry revealed a strong immunopositivity for CD31 and factor VIII in all endothelial cells lining the vascular channels. CD34 was moderately immunopositive (Figure 2) and VEGF was negative.

Based on morphologic and immunohistochemical findings, the lesion was diagnosed as hobnail hemangioma. The patient is on regular follow-up and six months after the surgery, no abnormalities were found on examination.

Discussion

The characteristic histopathologic features of hobnail hemangioma include prominent proliferation of irregular, thin-walled ectatic vessels lined by prominent epithelioid endothelial cells in the superficial dermis. In the deeper dermis, the vascular spaces are more angulated and slit-like and appear to dissect through collagen bundles. There are variable concentration of inflammatory cells (primarily lymphocytes), extravasated erythrocytes, edema, and numerous perivascular hemosiderin-laden macrophages in the dermis. Intravascular papillary projections and thrombi may be observed. The prominent endothelium gives a hobnail appearance.^{2,4} The present case showed similar histopathologic features except hemosiderin deposition (Figure 2).

A hobnail endothelial cell is characterized by a high nucleus to cytoplasmic ratio and an apically placed, occasionally grooved nucleus that produces a surface bulge, accounting for the term "hobnail" or "match stick" pattern (Figure 2).¹

Immunohistochemistry revealed a strong immunopositivity for CD31 and factor VIII. CD34 was moderately immunopositive (Figure 2) and VEGF was completely negative. CD31 is the best marker for benign and malignant vascular tumors. Previous studies have documented CD31 positivity in majority of the cases and CD34 positivity in minority of the cases.⁵ The present case also showed a similar pattern of immunostaining mentioned in previous reports. Immunoreactivity for factor VIII indicates the cytoplasmic luminal features and the epithelial cellular neoplastic nature.⁶

Furthermore, a limited number of cases stained positively for VEGF and D2-40, which are the markers of vascular tumors with presumed lymphatic differentiation. Results of such studies suggested a lymphatic line of differentiation of neoplastic cells in hobnail hemangioma. However, immunoexpression of VEGF and D2-40 in angiomatous entities has alternatively been interpreted by some as a possible origin from stem cells that would be able to differentiate either into lymphatic cells or blood vessels.^{7,8} In the present case, endothelial cells were negative for VEGF. Many studies also tried to correlate the clinical variations in hobnail hemangioma with hormonal variations. But these studies failed to locate the estrogen and progesterone receptors based on immunohistochemistry.⁹

Hobnail hemangioma represents a broad spectrum of diagnoses clinically which include melanotic nevus, malignant melanoma, Kaposi sarcoma, hemangioma, and solitary angiokeratoma. Histopathologic differential diagnosis includes solitary angiokeratoma, retiform hemangioendothelioma, Kaposi sarcoma, benign lymphangioendothelioma, and well-differentiated angiosarcoma.²

Retiform hemangioendothelioma and hobnail hemangioma share similar histopathologic features showing neoplastic vascular spaces lined by a single layer of monomorphic hobnail endothelial cells. However, the morphologic hallmark of retiform hemangioendothelioma which can be differentiated from hobnail hemangioma includes long, arborizing, thin-walled vascular spaces that infiltrate into a retiform pattern reminiscent of the normal rete testis.¹⁰

Hobnail hemangioma displays slit-like vascular spaces in the deeper dermis, which is a histopathologic feature indistinguishable from that of patch stage (early lesion) Kaposi sarcoma. However, hobnail hemangioma consists of dilated blood vessels in the

superficial dermis which is lined by endothelial cells, showing hobnail cytomorphology. In contrast, patch stage Kaposi sarcoma consists of flat, hyperchromatic endothelium.¹¹ Kaposi sarcoma is immunopositive for CD34 and actin-positive pericytes, whereas hobnail hemangioma is not surrounded by complete layer of actin-positive pericytes and only few cases showed CD34 positivity.

Lymphangioendothelioma can be differentiated from hobnail hemangioma by its quite monotonous appearance throughout under low-power microscopic examination, whereas hobnail hemangioma is recognized by its characteristic biphasic growth pattern.² Well-differentiated angiosarcoma is characterized morphologically by anastomosing multilayered vascular spaces lined by atypical and proliferating active endothelial cells.⁶ Simple surgical excision is the treatment of choice and prognosis is excellent.

Hobnail hemangioma is a benign vascular lesion characterized by hobnail endothelial cytomorphology and biphasic growth pattern. Since many benign and malignant vascular tumors display similar hobnail cytomorphology, it is essential for the accurate diagnosis of hobnail hemangioma for proper treatment and prognosis. Further studies are required to assess the tissue of origin and etiology as it aids in diagnosis, treatment, and prognosis.

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