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CASE REPORT

Congenital and Multiple Hobnail Hemangiomas

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Hobnail hemangioma (targetoid hemosiderotic hemangioma) is a vascular tumor affecting the limbs or trunk. Characteristically, the lesion has a "targetoid" clinical feature and dilated vascular spaces lined by hobnail endothelial cells at histologic examination. The age of onset is widely variable, form 5～67 years, typically occurring in young or middle-aged persons. It is usually apparent as a small solitary lesion. However, multiple lesions are identified sometimes. Herein, we report two cases of hobnail hemangioma in 7-year-old and 15-year-old males. Of note, the former case had a congenital lesion and the latter, multiple acquired lesions, which are both rare atypical presentations of the disease. **(Ann Dermatol 23(4) 539～543, 2011)**

-Keywords-
Congenital, Hemangioma, Multiple

INTRODUCTION

Hobnail hemangioma (targetoid hemosiderotic hemangioma) is a benign vascular tumor that typically presents as a small, single lesion on the skin of the trunk or limb of a young or middle-aged adult[1,2]. The lesion has a characteristic 'targetoid' appearance, in which a violaceous papule is surrounded by an ecchymotic or brown ring that can expand or subsequently disappear, with persistence of the central papule. In the Korean literature, there are 10 case reports. We report two additional cases of hobnail hemangioma revealed by typical clinical and histological appearance. Remarkably, one of them was a congenital lesion, never described before, and the other was a multiple hobnail hemangioma, which is also an atypical presentation.

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CASE REPORT

Case 1

A 7-year-old boy presented with a dark brown papule with ecchymotic halo on the left upper back. According to the patient's mother, it had been there as a black macule since birth and recently the size had increased. The patient had no subjective symptoms and denied any history of trauma. On examination, the skin lesion was a 4 mm-sized, well-demarcated dark brown, irregularly shaped papule surrounded by a thin, pale area and a peripheral ecchymotic ring (Fig. 1). Histologic examination revealed dilated vascular channels with hobnail endothelial cells protruding into the lumen and occasional intra-

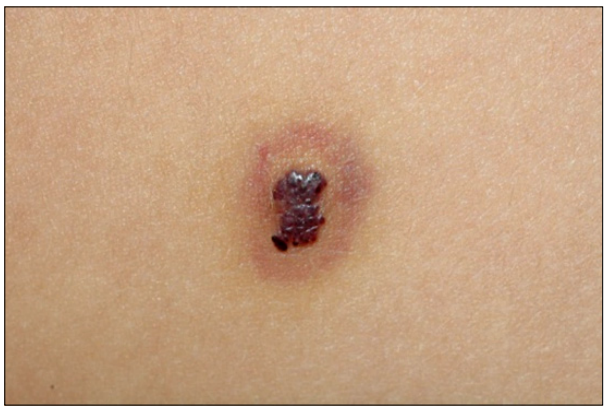

Fig. 1. A dark brown papule with ecchymotic halo on left upper back (case 1).

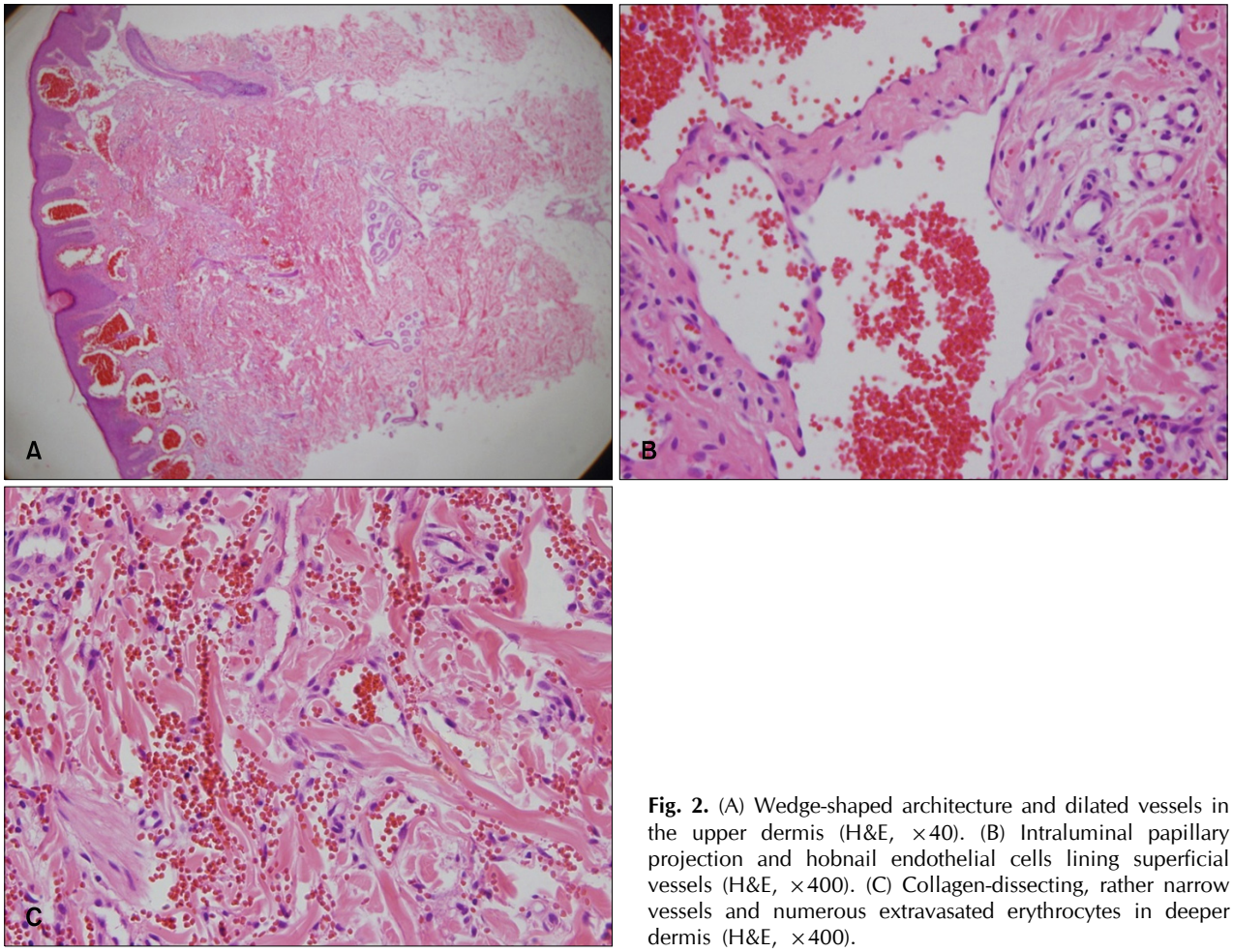

Fig. 2. (A) Wedge-shaped architecture and dilated vessels in the upper dermis (H&E, ×40). (B) Intraluminal papillary projection and hobnail endothelial cells lining superficial vessels (H&E, ×400). (C) Collagen-dissecting, rather narrow vessels and numerous extravasated erythrocytes in deeper dermis (H&E, ×400).

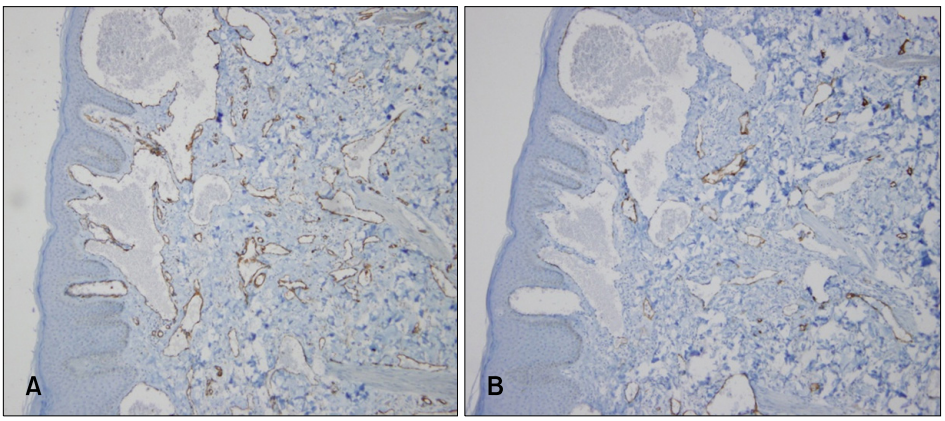

Fig. 3. Strongly positive reaction with CD31 (A) and focal positive reaction with D2-40 (B) (×100).

luminalpapillaryprojections in the upper dermis. Deeper in the dermis, the vascular channels were thinner and seeme d to dissect the collag en bundles. There were numerous extravasated erythrocytes (Fig. 2). Immunohistochemistry with monoclonal antibodies revealed strongly positive reaction with CD31 and focal positive reaction with CD34 and D2-40 (Fig. 3). The lesion disappeared completely following punch biopsy, and no recurrence was observed.

Case 2

A 15-year-old boy presented with two dusky red to brown plaques on the medial and lateral aspect of the left knee that appeared 12 and 4 years ago, respectively. The pa-

tient had no subjective symptoms and denied any history of trauma. The patient said that these lesions had been indurated small papules but recently the size had increased and the color darkened progressively. Physical examination revealed two dusky erythematous plaques about 2 cm in size with surrounding ecchymotic macular rings (Fig. 4). A 4-mm skin biopsy was performed from the superolateral lesion to rule out verrucous hemangioma and Kaposi's sarcoma. Histologic examination was very similar to that of the first case, and additionally hemosiderin deposition was widely present in the dermis (Fig. 5). HHV-8 staining was negativ e. The lesions improved substantially following intermittent triamcinolone intralesional injections and pulsed dye laser treatment.

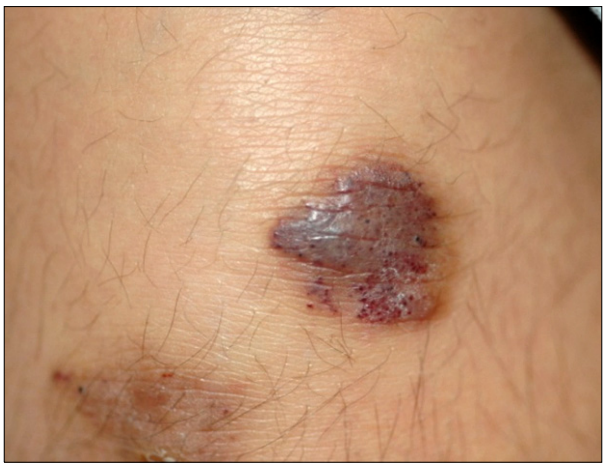

Fig. 4. Two dusky red to brown plaques with surrounding ecchymotic macular rings on the left knee (case 2).

DISCUSSION

Hobnail hemangioma was first reported by Santa Cruz and Aronberg in 1988[3]. A typical clinical appearance is a small solitary lesion consisting of a 2～3 mm-sized brown to violaceous papule surrounded by a thin, pale area and a peripheral ecchymotic ring. The characteristic targetoid appearance is due to peripheral hemorrhage and subsequent deposition of hemosiderin. However, these featuresareonlypresent in a small percentage of cases and, most often, the clinical appearance is that of a red-blue or brown papule.

The etiology of hobnail haemangioma is unknown, but traumas to a pre-existing hemangioma and the influences of sex steroid hormones have been proposed[4-6]. Interestingly, some lesions in females change during the menstrual cyce or pregnancy.

Published reports of hobnail hemangioma reveal an equal gender incidence, lwith an age range of presentation of 5～67 years. It is identified more frequently in younger persons[3]. There are 10 case reports in the Korean literature[7-16] (Table 1). In those cases, skin lesions developed at 3～32-years-of-age. Nine patients had a solitary lesion, but one patient had two papules on the upper back and arm[8]. Five lesions arose in the lower extremities, four on the trunk and two on the upper extremities. Our 'case 1' is the youngest patient reported in Korea so far a nd is the first patient that has had the skin lesion since birth. 'Case 2' is the second case with two lesions after the first one reported in the Korean literature[8].

Histologically, characteristic features are irregularly dilated vesHsels lined by hobnail endothelial cells in the superficial dermis and collagen-dissecting, rather narrow vessels in deeper dermis. The hobnail endothelial cell has

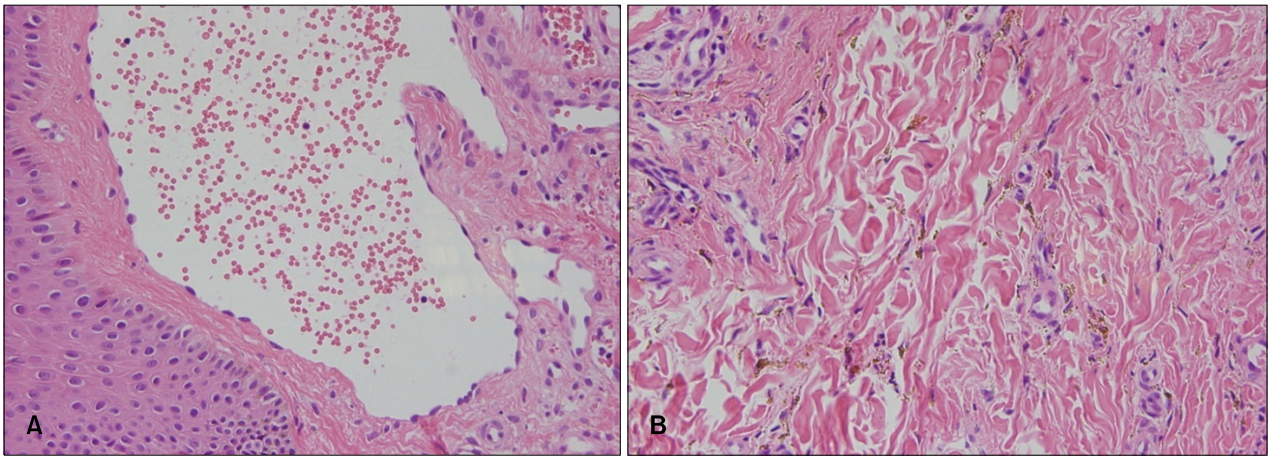

Fig. 5. (A) Intraluminal papillary projection and hobnail endothelial cells lining superficial dilated vessels (H&E, ×400). (B) Hemosiderin deposits in deeper dermis (H&E, ×400).

Table 1. Reported cases of hobnail hemangioma in the Korean literature

No.	Cases	Sex/Age	Duration	Location	Clinical finding	Immunohistochemistry & histochemistry	Outcome
1	Moon et al.[7]	M/23	5 years	Lower back	TP	Peals potassium ferrocyanide stain: hemosiderin deposits	Excision: disease free
2	Lee et al.[8]	M/28	5 years	Upper back & Rt. arm	TP	Peals potassium ferrocyanide stain: hemosiderin deposits	Punch biopsy: disease free
3	Seo et al.[9]	M/26	4 years	Lt. arm	Reddish papule	Factor VIII related antigen: positive CD34: positive	Excision: disease free
4	Oh et al.[10]	F/21	5 weeks	Lt. shin	Dusky red patch	Factor VIII related antigen: positive; *Ulex europaeus* agglutinin I: negative; CD34: strongly positive	Excision: recurrence
5	Park et al.[11]	F/33	1 year	Lt. abdomen	TP	Peals potassium ferrocyanide stain: hemosiderin deposits	Excision: disease free
6	Cho et al.[12]	F/29	1 year	Lt. shoulder	TP	Gomori iron stain: hemosiderin deposits; CD34: strongly positive	Excision: disease free
7	Yun et al.[13]	F/12	1 year	Rt. thigh	TP	Factor VIII related antigen: positive; CD31, CD34: positive	Excision: disease free
8	Roh et al.[14]	F/10	5 years	Lt. thigh	TP	Factor VIII related antigen: positive; CD34: positive	Excision: disease free
9	Yun et al.[15]	F/17	10 years	Rt. leg	TP	CD34: positive	Follow-up loss
10	Kang et al.[16]	F/8	5 years	Lt. heel	TP	CD34: positive; smooth muscle actin: positive	Follow-up loss
11	Present case 1	M/7	7 years	Upper back	TP	CD34: positive; CD31, D2-40: focal positive	Punch biopsy: disease free
12	Present case 2	M/15	12 years	Lt. knee	Dusky red plaques	Iron stain: hemosiderin deposits	TA ILI and pulsed dye laser: improved

M: male, F: female, Rt: right, Lt: left, TP: targetoid papule.

scanty cytoplasm and rounded nuclei that protrude into theumina. Focally, intraluminal papillary projections can be seen in the superficial blood vessels. The vascular channels in the deeper dermis become much less conspicuous and eventually disappear completely. The whole architecture is wedge l-shaped, with a prominent superficial component. In the later stages, extensive stromal hemosiderin deposits are commonly seen.

The clinical differential diagnoses include melanocytic nevus, dermatofibroma, hemangioma and insect bite reaction. The microscopic differential diagnoses are the patch stage of Kaposi's sarcoma, retiform hemangioendothelioma, solitary angiokeratoma, progressive lymphangioma and eosinophilic hemangioma. It is especially important to distinguish hobnail hemangioma from the patch stage of Kaposi's sarcoma. Factors favoring Kaposi's sarcoma are the presence of plasma cells, spindle-shaped cells and apoptotic endothelial cells[17].

The tumor origin is controversial. Santonja and Torrelo[18] suggested a vascular origin for hobnail hemangioma since it had positive reaction with Factor VIII-related antigen, CD31 and CD34. However, Franke et al.[19] proposed that this tumor has a lymphatic origin because it revealed positive reaction with D2-40 and CD31, and a negative reaction with CD34. In our first case, immunohistochemistry revealed a mild focal positive reaction with CD34 and D2-40 and strong positive reaction with CD31. These results imply that the origin is vascular endothelial cells, not lymphatic endothelial cells.

To our knowledge, all reported cases are acquired type. Here, we report two cases of hobnail hemangioma that are peculiar in that one is congenital and the other is multiple.

REFERENCES

1. Mentzel T, Partanen TA, Kutzner H. Hobnail hemangioma ("targetoid hemosiderotic hemangioma"): clinicopathologic and immunohistochemical analysis of 62 cases. J Cutan Pathol 1999;26:279-286.
2. Guillou L, Calonje E, Speight P, Rosai J, Fletcher CD. Hobnail hemangioma: a pseudomalignant vascular lesion with a reappraisal of targetoid hemosiderotic hemangioma. Am J Surg Pathol 1999;23:97-105.
3. Santa Cruz DJ, Aronberg J. Targetoid hemosiderotic hemangioma. J Am Acad Dermatol 1988;19:550-558.
4. Morganroth GS, Tigelaar RE, Longley BJ, Luck LE, Leffell DJ. Targetoid hemangioma associated with pregnancy and the menstrual cycle. J Am Acad Dermatol 1995;32:282-284.
5. Ortiz-Rey JA, González-Ruiz A, San Miguel P, Alvarez C,

Iglesias B, Antón I. Hobnail haemangioma associated with the menstrual cycle. J Eur Acad Dermatol Venereol 2005;19: 367-369.
6. Christenson LJ, Stone MS. Trauma-induced simulator of targetoid hemosiderotic hemangioma. Am J Dermatopathol 2001;23:221-223.
7. Moon TK, Chun YS, Chun SI, Chung KY. A case of targetoid hemosiderotic hemangioma. Korean J Dermatol 1999;37: 627-630.
8. Lee JR, Lee SW, Choi GS, Lee SC, Kim YK. A case of targetoid hemosiderotic hemangioma. Ann Dermatol 2001; 13:228.
9. Seo PG, Kang HA, Kim HO, Kim CW. A case of hobnail hemangioma. Korean J Dermatol 2001;39:1144-1147.
10. Oh ST, Lee SD, Kim SU, Jang IG, Cho BK. A case of hobnail hemangioma. Ann Dermatol 2002;14:45.
11. Park SY, Kim KH, Kim CW, Kim BC, Lee KS. A case of targetoid hemosiderotic hemangioma. Korean J Dermatol 2003; 41:795-798.
12. Cho KM, Kim TJ, Hwang KY, Lee JS, Lee SY, Park YL. A case of hobnail hemangioma. Korean J Dermatol 2003;41:1677-1680.
13. Yun JH, Lee JY, Yoon TY. A case of targetoid hemosiderotic hemangioma. Korean J Dermatol 2007;45:197-199.
14. Roh BH, Whang KU, Kim YM, Cho MK, Park YL, Lee JS. A case of childhood hobnail hemangioma. Korean J Dermatol 2007;45:979-982.
15. Yun JH, Kim KR, Kim MK, Lee JY, Yoon TY. A case of targetoid hemosiderotic hemangioma. Korean J Dermatol 2008; 46:554-556.
16. Kang YS, Kim SW, Park SH, Lee UH, Park HS, Jang SJ. A case of hobnail hemangioma that occurred together with angiolipoma. Korean J Dermatol 2009;47:718-721.
17. Chor PJ, Santa Cruz DJ. Kaposi's sarcoma. A clinicopathologic review and differential diagnosis. J Cutan Pathol 1992; 19:6-20.
18. Santonja C, Torrelo A. Hobnail hemangioma. Dermatology 1995;191:154-156.
19. Franke FE, Steger K, Marks A, Kutzner H, Mentzel T. Hobnail hemangiomas (targetoid hemosiderotic hemangiomas) are true lymphangiomas. J Cutan Pathol 2004;31:362-367.