Case 22
A 42 year-old woman from Bangkok

Chief complaint: Recurrent solitary erythematous mass on right thigh for 7 months

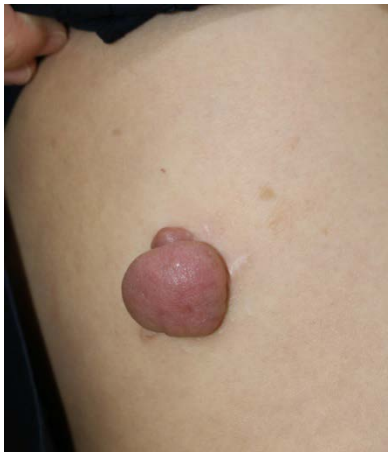


Figure 22.1

Present illness:
27 years ago, she presented with asymptomatic solitary erythematous papule on right thigh. 5 years later, the lesion increased in size, so the excision was done. The diagnosis was lymphangioma.
4 years ago, she developed recurrent solitary erythematous nodule 2.5 cm in diameter on the same site. The lesion was excised at private hospital. Diagnosis was epithelioid hemangioma which confirmed by histopathology (not free margin)
7 months ago, she developed recurrent solitary exophytic erythematous mass on the same site again, then she visited at Ramathibodi hospital.

Underlying disease: Graves' disease S/P I^{131} ablation and thyroidectomy since 1996, spondyloarthropathy and dyslipidemia

Family history: There was no family history of similar skin lesion.

Dermatologic examination: (figure 22.1)

Solitary large firm exophytic erythematous nodule on right thigh

Physical examination: Systemic examination other than skin revealed no abnormality. (no lymphadenopathy)

Histopathology: (S18-020567, skin, right thigh)

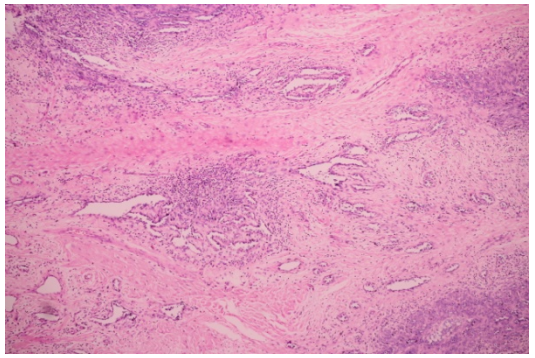


Fig. 22.2

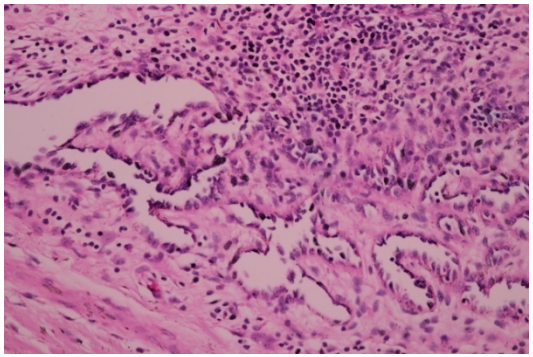
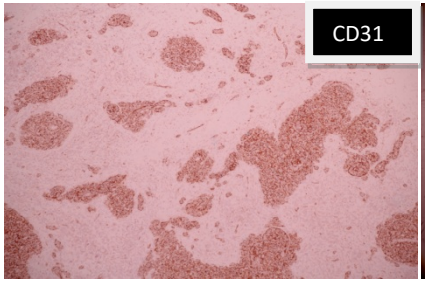


Fig. 22.3



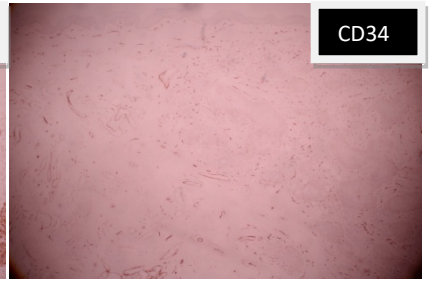


Fig. 22.4 Fig. 22.5

- Proliferation of abnormal blood vessels characterized by arborizing elongated blood vessels in association with dense lymphocytic infiltrate (fig. 22.2)
- Abnormal vessels lined by hobnail-like endothelial cells (fig. 22.3)

Immunohistochemistry: CD31+, CD34- (figure 22.4, 22.5)

Diagnosis: Retiform hemangioendothelioma

Treatment: Wide excision (not free margin), appointment for MRI right thigh and plastic surgery consultation for surgical excision

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Consultant: Saneerat Porntharukcharoen, MD

Discussion:
Retiform hemangioendothelioma (RHE) or Hobnail hemangioendothelioma is a locally aggressive vascular tumor and can be regarded as a low-grade angiosarcoma.[1] RHE commonly presents in young to middle-aged persons (varies from 6-78 years) with female predominance (male:female = 1:2).[2] Although etiology is unknown, human herpes virus type 8 has been purported to be associated with RHE.[3] It presents clinically as a slow-growing asymptomatic solitary nodule or plaque on extremities, although there are also cases on head, trunk, and penis.[4] Duration of the disease and tumor size at the time of diagnosis have been reported to range from 2 months to several years and from 1-30 cm, respectively.[5] Tumor has an indolent course with very high local recurrence and rare metastasis.

Histologically, neoplasm involves entire thickness of the dermis and often extends to subcutaneous tissue. Infiltrative neoplasm composed of elongated arborizing vessels is arranged in anastomosing pattern resembling that of the rete testis. Neoplastic vessels are lined by single layer of hobnail-like endothelial cells that protrude within the narrow lumina. The nuclei of hobnail-like endothelial cells are hyperchromatic, whereas their cytoplasm is scant. Mitotic figures are absent or scant.[4] Immunohistochemically, the hobnail-like endothelial cells lining the elongated vascular

spaces express endothelial markers, including von Willebrand factor, CD31, and CD34.[6]

The differential diagnosis of hobnail-like endothelial cells includes Hobnail hemangioma (HH), papillary intralymphatic angioendotheliomas (PILA) and RHE. HH presents as a small erythematous papule surrounded by an ecchymotic halo. Histopathological characteristics are superficial elongated vascular channel with abundant deposits of hemosiderin.[4] PILA presents as enlarging dermal or subcutaneous mass or plaque. Histopathology shows intercommunicating, thin-wall vessels lined by hobnail-like endothelial cells with intraluminal papillary projections and rosette appearance.

Surgical excision is the treatment of choice for RHE. However, accurately defining the excision margins is particularly troublesome. Indeed, RHE is associated with a high rate of local recurrence (50%), which may occur from months to several years after surgery.[5] Only 2 patients has been reported to develop regional lymph node metastasis and no patients died of their neoplasms.[6,7]

Reference:

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