Retention hemangioendothelioma of the lower limbs: a case-based review

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Summary
Retention hemangioendothelioma is a rare entity included in the spectrum of vascular tumors. It is considered an uncommon vascular neoplasm of intermediate or borderline malignancy (between entirely benign hemangiomas and highly malignant angiosarcomas). Retention hemangioendothelioma is clinically characterized by exophytic nodules and plaques in the dermal or subcutaneous compartments, which grows slowly involving more often lower limbs, upper limbs and trunk. Histologically, it shows a typical vascular pattern characterized by microscopic feature of arborizing, elongated, thin-walled cavernous blood vessels dissecting the dermal collagen. In this article, we report a case of retention hemangioendothelioma in a 86-year-old male and review the literature concerning this rare entity.

KEY WORDS: retention hemangioendothelioma; vascular tumors; skin neoplasms.

Introduction
Retention Hemangioendothelioma (RH) is a rare entity included in the spectrum of vascular tumours. It is considered an uncommon vascular neoplasm of intermediate or borderline malignancy (between entirely benign hemangiomas and highly malignant angiosarcomas). RH is clinically characterized by exophytic nodules and plaques in the dermal or subcutaneous compartments, which grows slowly involving more often lower limbs, upper limbs and trunk.

Case report
We describe the case of a 86-year-old male patient with one year history of slowly enlarging nodules and plaques of both the lower limbs (Figure 1a, b). The patient had no constitutional symptoms such as weight loss, fever or decreased appetite. His recent medical history showed stroke, hypertension, ischemic heart disease and deep vein thrombosis. Examination of the proximal lower extremity revealed purple nodules and Kaposi like plaque skin lesions. Physical examination of the distal lower limbs extremity revealed visible soft tissue mass with edema (Figure 1b). The mass was soft and spongy on palpation. Laboratory examinations including HHV8 and HIV serology status were negative. A biopsy obtained from the tumoral mass on the lower limb was submitted for histological evaluation. This analysis revealed a characteristic infiltrative growth pattern, involving the entire dermis but sparing the subcutis. Histologic examination of the tumor showed a typical vascular pattern characterized by microscopic feature of arborizing, elongated, thin-walled cavernous blood vessels dissecting the dermal collagen (Figure 2a). This vascular pattern resembles the architecture of the rete testis. These arborizing blood vessels were lined by monomorphic endothelial cells with prominent nuclei showing a characteristic hobnail-like or thrombosis-like appearance (Figure 2b). Rare solid endothelial areas were present in the superficial part of the lesion. In some areas an evident lymphocytic infiltrate was present. Cytologic atypia was minimal and mitotic figures were rare. The immunohistochemical analysis showed tumor cells reacting with endothelial markers, notably CD31 (Figure 2c) and CD34. Smooth muscle actin stained the vascular wall, but was negative in the tumor infiltrate (Figure 2d). There was no clinical or radiological evidence of regional lymph node involvement. Computed tomography scan showed no evidence of metastatic disease. The patient was initially evaluated for surgical resection but was deemed unresectable because of extent of the disease, age and comorbidities. Therefore no treatment was made and after 6 months of follow-up no progression of the disease was observed.

Discussion
We hereby report this case of RH because of its rarity and the uncommon clinical presentation. RH is a rare vascular neoplasm of intermediate borderline malignancy characterized by a net-like growth pattern.
pattern, a high rate of local recurrence and low frequency of metastasis (1-3). The first description of this tumor was made by Calonje et al. (1). To date only thirty-six cases (4) of RH have been reported in the literature. RH has been described as a neoplasm with a wide clinical heterogeneity. Therefore, the clinical recognition of RH is extremely difficult because of its non-specific appearance with differential diagnosis.

Figure 1 - (a) A 86-year-old male presenting with extended RH of the lower limbs bilaterally. (b) Multilobulated violaceous plaque on the foot. The mass feels soft and spongy on palpation.

Figure 2 - (a) Histopathological image of the tumor shows a vascular pattern with multiple interconnecting, elongated arborizing blood vessels involving the dermis and subcutis. (b) The same section under high magnification shows that arborizing blood vessels are arranged in retiform pattern; the endothelial cells is characterized by prominent protuberant nuclei having a characteristic hobnail-like appearance. (c) The diffuse immunohistochemical positivity with CD31 confirmed the endothelial origin of the neoplastic proliferation, but not for smooth muscle actin. (d) Smooth muscle actin highlights the vascular wall, but negative in the tumor cells.
comprising a variety of benign and malignant tumors (5). There is a wide variability in the age of patients diagnosed with RH; usually it occurs in the second to fourth decade of life with a mean age of 36 years (1,2). RH has also been reported in patients with less than 10 years of age (6). The tumor shows a predilection for females. Morphologically RH can occur in different areas: lower limbs, scrotum, upper arms, neck, mandible, external auditory canal, scalp. Other vascular tumors that can show similar clinical features include Dabska’s tumor, targetoid hemosiderotic hemangioma, hobnail hemangioma, epitheloid hemangioendothelioma, polymorphous hemangioendothelioma of lymph node, Kaposi Sarcoma (KS), cutaneous lymphoma, blue rubber bleb nevus syndrome, dermatofibrosarcoma protuberans, Amelanotic Melanoma (AM), bacillary angiomatosis and cutaneous metastases from solid tumors.

Dermoscopy has been proposed to improve the clinical evaluation of vascular skin tumors, enabling the visualization of morphologic structures that might be critical for the differential diagnosis (7). However the dermoscopic criteria of RF and other endotheliomas require further investigation and dermoscopic findings should be always be integrated with clinical information, patient age and patient history (5). Skin biopsy, therefore is at present the only diagnostic method available for an appropriate diagnosis of RH (8, 9). RH should be considered a low grade angiosarcoma with high incidence of recurrence, consequently in cases where it is possible, a wide surgical excision with histopathologically proven tumor-free margins is the optimal choice (10). Amputation of toe, partial penectomy and amputation of finger have been described in the setting of RH therapy (11). Unresectable RH can be treated with external beam radiation therapy and chemotherapy. In addition, adjuvant immunotherapy with recombinant interferon alpha has been utilised.

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References