RESIDENT’S FORUM

A solitary papule on the scalp

Case report

A man aged 37 years presented with a 2-year history of solitary papule on the left scalp. The lesion was asymptomatic but enlarged gradually. The patient did not have cutaneous hyperpigmentation, hypertrichosis, peripheral neuropathy, muscle weakness, peripheral edema, or weight loss. A physical examination revealed a single pea-sized, elastic-to-firm erythematous papule with mild telangiectasia on his left scalp (Figure 1). Under the diagnosis of an appendage tumor, an excisional biopsy was performed.

Histologically, it revealed an intradermal vascular tumor (Figure 2A). The tumor was composed of multilobulated capillary proliferation with papillary architectures, and it grew within a dilated vascular space. The lining endothelial cells were plump and filled with many hyaline globules (Figure 2B). Periodic acid Schiff (PAS) stain also highlighted the hyaline globules (Figure 2C). Cellular atypia was not observed and human herpesvirus 8 (HHV-8) stain was negative. There was no recurrence during 5 months of follow-up.

Figure 1 A pea-sized, elastic-to-firm erythematous papule with mild telangiectasia on the left scalp.

Figure 2 (A) Intravascular capillary proliferation with papillary architecture (hematoxylin and eosin stain (H & E), 20×). (B) the endothelial cells were plump and filled with many hyaline globules (H & E, 200×); (C) the hyaline globules were stained positively with periodic acid Schiff stain (H & E, 200×).
Diagnosis

Papillary hemangioma (PH).

Discussion

PH is a novel vascular proliferative lesion described by Fletcher and colleagues in 2007.1 It usually presents as a small, solitary, non-tender, bluish skin papule on the head and neck area of patients with a mean age of 57 years. On microscopic examination, it is primarily situated in the dermis with some extension into the superficial subcutaneous fat. At low-power magnification, the lesion appeared as an intravascular papillary capillary growth within ectatic thin-walled dermal blood vessels and surrounded by dermal pilosebaceous units. At high-power magnification, the papillae were focally covered by swollen endothelial cells that contained numerous hyaline globules, which sometimes results in an indented nucleus or a burst of the cytoplasm. Immunohistochemical stain shows the lining cells of the capillaries within the papillary stalks, and those cells covering the outer surface of the papillary stalks, including the swollen cells with hyaline granules, were reactive to CD31 and CD34. Electron microscopic examination revealed that hyaline granules were consistent with electron-dense giant lysosomes that contained organelle debris and fat vacuoles, also called thanatosomes.2 Dysfunction of the autophagocytic-lysosomal pathway is thought to play a role in the pathogenesis.1

The histopathologic pattern of intravascular proliferation of capillary-sized blood vessels makes glomeruloid hemangioma (GH) the major differential diagnosis. Suurmeijer2 addressed several distinctions between PH and GH: (1) PH is solitary, located on the head and neck area, while GH is multiple and is usually on the trunk or extremities, (2) GH is usually related with polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome, whereas PH is not observed in any patient with POEMS syndrome, and (3) microscopically, PH is larger and papillary-predominant, whereas GH is smaller with a glomeruloid structure. Collagen type IV has been recommended as the best immunohistochemical stain because it highlights the glomeruloid architecture and thin basal membranes of GH, which is in contrast to the thick basal membrane-like matrix of PH.2 However, the eosinophilic globules in glomeruloid hemangioma and the hyaline globules in papillary hemangioma seem to be undifferentiable under histologic or immunohistochemical examination.3 The nature between these two conditions remains to be elucidated.

Conclusion

In our case, an intravascular pyogenic granuloma (PG)-like structure under low-power magnification suggested other differential diagnoses, such as intravascular papillary endothelial hyperplasia, PG-like Kaposi’s sarcoma, and angiosarcoma. Intravascular papillary endothelial hyperplasia is characterized by papillary proliferation of endothelial cells frequently associated with organizing thrombus, but it does not contain eosinophilic hyaline globules. PG-like Kaposi’s sarcoma may be histologically indistinguishable from PG under a hematoxylin and eosin stain, so an HHV-8 stain is helpful for differentiation.4 Further, our patient’s lesion does not show nuclear pleomorphism, hyperchromatism, atypical mitosis, or collagen bundle dissection, so angiosarcoma is less likely. The distinctive histologic presentation of PH and its intravascular PG features makes this case unique.

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