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Cholesterotic fibrous histiocytoma with no associated dyslipidemia

Dermatofibroma, also known as fibrous histiocytoma, is a relatively common benign tumor of fibrohistiocytic origin. It typically occurs as single or multiple firm nodules on the lower limbs of young women. More than 40 clinicopathologic variants of dermatofibroma have been reported according to their clinical and pathological characteristics. In particular, cholesterotic fibrous histiocytoma is a rare variant of dermatofibroma.[1] It is clinically identical to classic dermatofibroma but is characterized by cholesterol deposits within the lesion and an appearance suggests the possibility of an underlying lipid metabolism abnormality. In addition to the existing cases, we report a case of dermatofibroma with cholesterol crystal deposits observed in a patient without associated dyslipidemia.

Our patient was a 29-year-old woman with no personal and family history of hypercholesterolemia and hypertriglyceridemia. She was referred to our dermatology clinic for evaluation of a skin lesion on her right upper arm. She had noticed the tumor around the age of 10 years, and the tumor had become tender a year before presentation. On physical examination, a dome-shaped, firm nodule measuring 0.9 mm × 0.8 mm was located in an 18 mm × 10 mm reddish-brown macule (Fig. 1a). Laboratory findings included a complete blood count and biochemistry. Cholesterol (199 mg/ml) and triglyceride levels (82 mg/dl) were normal.

Based on the clinical diagnosis of dermatofibroma, the lesion was excised. The lesion consisted of a poorly demarcated dermal nodule (Fig. 1b). The overlying epidermis showed acanthosis, papillomatosis, and basal hyperpigmentation. The tumor revealed marked proliferation of spindle histiocytes and fibroblasts, sometimes in a storiform arrangement, between thick collagen bundles. No cytologic atypia or mitotic figures were observed. Groups of biconvex, needle-shaped cholesterol crystals surrounded by many foam cells were observed within the lesion (Fig. 1c). Immunohistochemically, spindle cells lacked the expression of CD34, α-smooth muscle actin, or S-100 protein. We diagnosed cholesterotic fibrous histiocytoma based on these histological findings.

Dermatofibroma presents with a variety of clinical and histological features. The main variants of dermatofibroma include cellular, aneurismal, epithelioid, and atypical fibrous histiocytomas. Many foam cells in the lesion might suggest lipidized fibrous histiocytoma, which is characterized histologically by accumulation of numerous foam cells, smaller numbers of siderophages, and stromal hyalinization, and concentrated strikingly in the lower limb, especially around the ankle.[2] However, the presence of cholesterol deposits indicate cholesterotic fibrous histiocytoma.

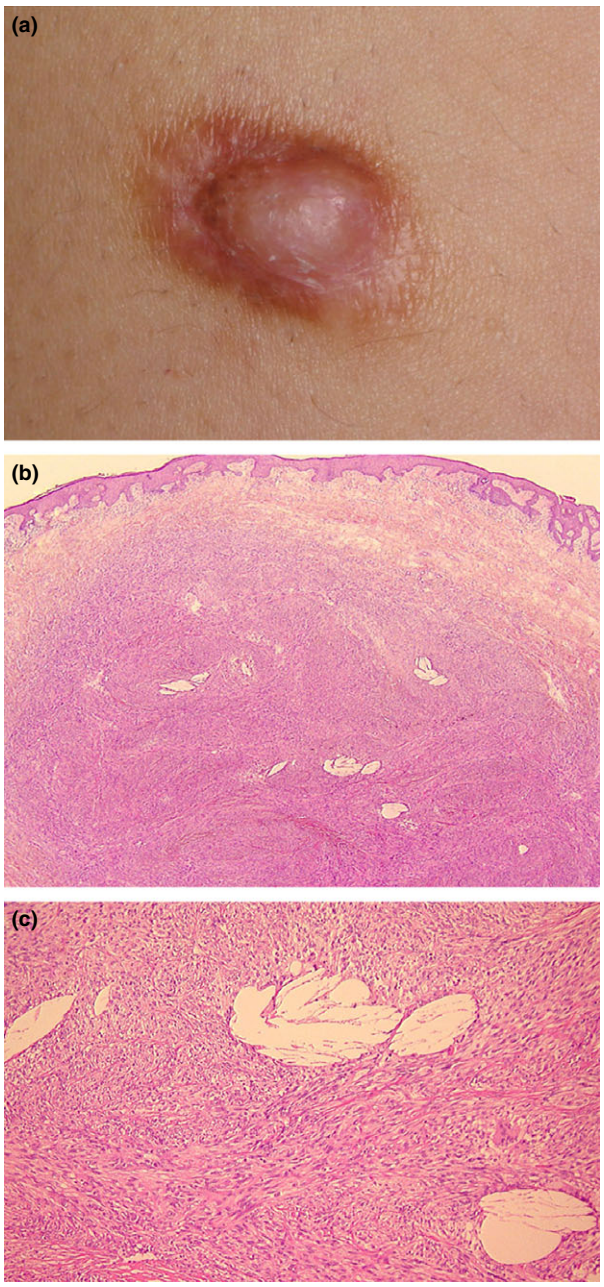

Figure 1 Clinical and histological findings. (a) The tumor was a dome-shaped, firm nodule measuring 0.9 mm × 0.8 mm located in an 18 mm × 10 mm reddish brown macule. (b) The tumor consisted of a poorly demarcated dermal nodule. The overlying epidermis showed acanthosis, papillomatosis, and basal hyperpigmentation. (c) Groups of biconvex, needle-shaped cholesterol crystals surrounded by many foam cells were observed within the lesion. Hematoxylin and eosin staining, ×40. Marked proliferation of spindle histiocytes and fibroblasts, sometimes in a storiform arrangement was found between thick collagen bundles. No cytologic atypia or mitotic figures were observed. Hematoxylin and eosin staining, ×100

The cholesterol variant of dermatofibroma, which is characterized by cholesterol crystal deposits, is very rare. To the best of our knowledge, only three cases of the variant have been reported in the English literature. The first was the case of a woman with a history of hypercholesterolemia whose histopathological study results showed cholesterol deposits and numerous foamy histiocytes.[1] The authors concluded that the histiocytic nature of the dermatofibroma enhanced by hypercholesterolemia induced the xanthomatous changes and cholesterol deposits. The second case was a patient with HIV infection but did not show any lipid metabolism abnormalities.[3] HIV-positive patients frequently have dermatofibroma and dyslipidemia, and treatments of HIV infection may lead to metabolic syndromes such as lipodystrophy and insulin resistance. Therefore, the authors suggested that a relationship between cholesterol deposits and HIV infection exists. Furthermore, a very recent paper reported the case with type 2 hyperlipidemia.[4]

Our case did not manifest dyslipidemia and HIV infection, as well as significant disorders related with cholesterol metabolism. Although the previous cases indicated the need for patients with dermatofibroma to undergo evaluation of lipid profiles and for the possibility of HIV infection, clinicians must be aware that a dermatofibroma with cholesterol deposits can occur even in healthy individuals. In addition, the presence of cholesterol clefts with foamy macrophages may imply a slowly growing process of the lesion, and this patient had noticed the current lesion around the age of 10 years. Thus, the cholesterotic deposit in some cases of cholesterotic fibrous histiocytoma might be simply histologic evidence of chronic process.

An interesting recent study has reported rare examples of metastasizing benign cutaneous fibrous histiocytomas and 11 of 16 cases were cellular variant.[5] This case is a relatively cellular lesion. Therefore, we have to see the clinical course carefully at this standpoint.

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Papular acantholytic dyskeratosis of vulva in setting of Hailey–Hailey

Acantholytic dyskeratosis is a histopathological pattern defined by a hyperkeratotic and parakeratotic epidermis with intraepidermal clefts containing acantholytic and dyskeratotic keratinocytes with the formation of corps ronds and grains.[1] These typical features are distinctive but not pathognomonic of diseases such as Darier' disease, Hailey–Hailey disease, Grover' disease, papular acantholytic dyskeratosis (PAD), and others.[1]

Hailey–Hailey disease (familial benign chronic pemphigus) is a rare intraepidermal blistering disorder inherited as an autosomal dominant trait. It is caused by mutations in ATP2C1 gene, which encodes a Golgi apparatus Ca^{2+}-ATPase protein, leading to incorrect processing of desmosomal proteins. This results in deficient cell adhesion with vesicles and erosions occurring in intertriginous sites prone to heat and friction.[2] The most commonly affected sites are the back of the neck, groin, perineum, and axilla.[3]

PAD is an exceedingly rare disorder of the intertriginous and genital areas characterized by numerous whitish papules that coalesce into plaques.[1] It primarily affects women in the vulva, perineum, and perianal areas.[4] It is most often reported as an isolated finding in patients without any history of blistering disorder. Previous reports indicate this may be a unique diagnosis from Hailey–Hailey disease.[1,4–6]

A 68-year-old woman with a history of Hailey–Hailey disease presented with complaints of a worsening blistering eruption over her inframammary chest, labia, and gluteal crease. She also complained of increased malodorous discharge. There was no family history of a similar eruption. Her disease was previously managed by her primary care physician with intermittent oral prednisone and oral antibiotics, and the eruption on her groin was treated with cryotherapy. She denied any history of abnormal Papanicolaou smears. On exam, she was found to have macerated, friable plaques on her groin, inframammary chest, and axilla with discrete whitish verrucous papules over most of the vulva (Fig. 1-2). A punch biopsy from the chest was consistent with Hailey–Hailey disease, with a negative direct immunofluorescence study. A biopsy of the vulvar lesions histologically revealed acantholytic dyskeratosis (Figs 3–4).

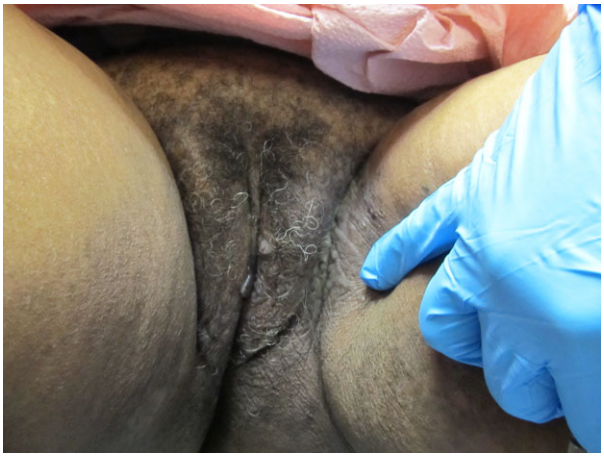

Figure 1 White verrucous papule on left labia submitted for pathology

Hailey–Hailey disease involving the vulva is not uncommon, affecting nearly 50% of females with Hailey–Hailey.[5] The clinical presentation of Hailey–Hailey disease of the vulva varies. Reports of leukoplakia, condyloma, lichenified white papules, and an association with syringomas have been made.[5] Histopathologic examination of Hailey–Hailey disease shows widespread suprabasal acantholysis with loss of intercellular bridges, most apparent at the spinous layer, which results in a characteristic "dilapidated brick-wall appearance."[5]

Clinically, PAD closely resembles Hailey–Hailey disease. However, PAD usually exhibits asymptomatic papules while Hailey–Hailey may have symptomatic vesiculation.[4] The

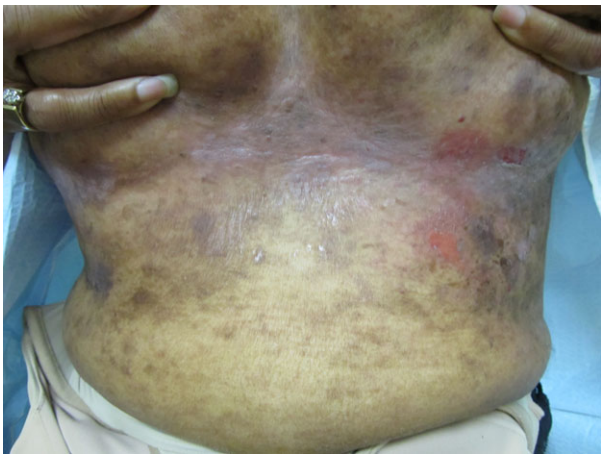

Figure 2 Inframammary area with typical findings of Hailey–Hailey disease