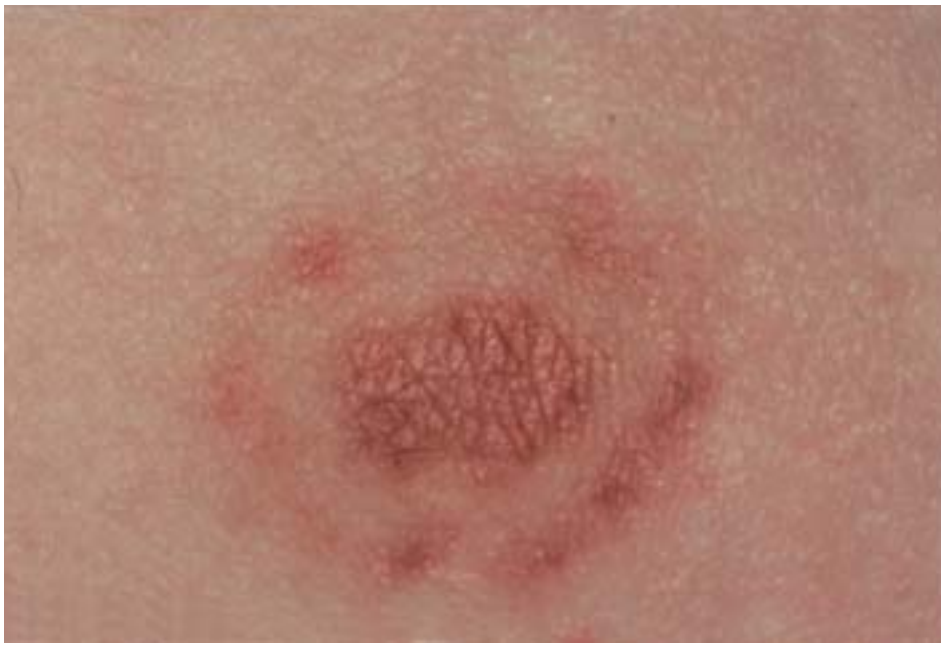
What Is Your Diagnosis?



A 45-year-old white woman presented with a solitary lesion on her lower back that had been present for approximately 2 years. The lesion changed monthly with menses. The patient described a brown papule that would enlarge, become deep purple in color, and develop a bruiselike rim with the onset of menses. Although the lesion was tender during menses, the tenderness and bruiselike rim would resolve following menses, and the lesion would revert back to a brown color. The patient was in good health and denied a history of previous trauma to the area or a history of abnormal vaginal bleeding or abdominal or pelvic pain. Physical examination revealed a 9×6 mm firm tan papule with a surrounding pale area and peripheral ecchymotic rim, resulting in a targetoid appearance. There were no other significant findings on cutaneous examination.

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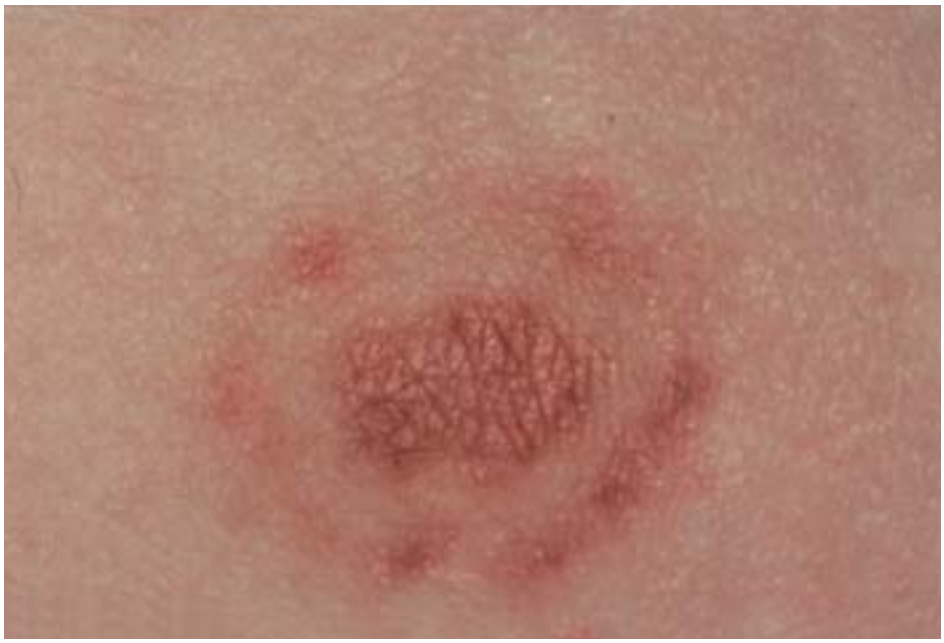
The Diagnosis: Targetoid Hemosiderotic Hemangioma



Targetoid hemosiderotic hemangiomas (THHs) were originally described in 1988 by Santa Cruz and Aronberg,[1] who reported a series of patients with single vascular lesions with a distinctive targetoid appearance and characteristic histopathology. Despite some histopathologic similarities to malignant vascular neoplasms, these lesions followed a benign clinical course. Although now well recognized, a recent review of the literature revealed a total of only 46 reported cases of THHs.[2] THHs appear most commonly on the trunk or lower extremities and are found in both genders equally. Most occur in young or middle-aged persons.[2]

The typical appearance of THH is that of a brown or violaceous papule or nodule surrounded initially by a pale area and later by an ecchymotic rim, resulting in a targetoid appearance. Over time, the ecchymotic rim expands peripherally and ultimately disappears, leaving the central papule.[1] The lesion often is misdiagnosed clinically as a melanocytic nevus, hemangioma, or dermatofibroma; lesions in the process of evolution may even be mistaken for melanoma.

The histopathology of THH is variable and depends on the stage of evolution. A biphasic pattern is characteristic. Superficially, dilated vascular spaces lined by protuberant endothelial cells displaying a hobnail appearance are evident. Intraluminal papillary projections lined by a single layer of endothelial cells also may be present. Deep to this, a proliferation of narrow, angulated, and slit-like vascular spaces dissect between the collagen bundles.[2] Fibrin thrombi, erythrocyte extravasation, and hemosiderin deposition are variable, as is a mild lymphocytic infiltrate. With maturity, the vascular spaces appear collapsed, and the stroma appears more fibrous with increased erythrocyte extravasation and hemosiderin deposition.

This histologic appearance of THH may be similar to that of early Kaposi sarcoma (KS). Features that may aid in distinguishing this lesion from KS include the prominent hemosiderin deposition and protuberant or "hobnailed" endothelial cells in dilated superficial vessels seen in THH versus the adnexocentricity seen in KS.[3]

The histopathologic differential diagnosis of THH includes hobnail hemangioma (HH), another recently described tumor. HH and THH share many similar features, including dilated vascular channels superficially and a collagen-dissecting pseudo-

angiosarcomatous pattern deeper, with the endothelial lining displaying hobnail cytomorphology. However, in HH a variable but often minimal amount of hemosiderin deposition is present. In addition, the HH lesion demonstrates a nontargetoid clinical appearance.[4] One study reported a series of 62 cases with histology similar to that of THH. However, in each of these cases, the lesion had a nontargetoid clinical appearance.[5] Despite the histologic similarity, the relationship between THH and HH remains unclear.

Trauma is thought to play a major role in the pathogenesis of THH.[6] The ecchymotic rim of THH correlates to hemosiderin deposits, a defining histopathologic feature.[2] Trauma to the vascular component of the central papule may result in extravasated erythrocytes and subsequent hemosiderin deposition. Trauma to a preexisting hemangioma resulting in thrombi also would explain histologic features such as intraluminal projections and fibrin thrombi.[7]

An interesting subset of THH consists of lesions with episodic and cyclic changes. These changes, though uncommon, have been reported to occur without preceding trauma,[2,8] and the cause remains unknown. One report described a father and son with THH, both exhibiting cyclic changes with no apparent cause.[8] Other cases occurring in women have been hypothesized to be mediated by hormonal factors. Carlson et al[2] reported one case, similar to the one reported here, in which cyclic changes occurred during menses, suggesting the diagnosis of endometriosis. However, these changes may have been due to fluctuating estrogen levels. Estrogen has been demonstrated to promote vascular permeability and fragility,[9] which may account for the extravasated erythrocytes and subsequent hemosiderin deposition characteristic of THH. A case termed *targetoid hemangioma* displayed similar clinical features, including cyclic changes in the ecchymotic rim occurring during menses.[9] Although the histologic features varied from THH, the similar clinical presentation lends support to the diagnosis of THH.

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