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Dermoscopy of kaposiform hemangioendothelioma: a case report

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Abstract

Kaposiform hemangioendothelioma (KHE) is a rare vascular tumor of infancy, characterized by an infiltrative, firm, and ill-defined plaque that may be associated with Kasabach-Merritt phenomenon. While the diagnosis is primarily clinical and histopathological, dermoscopy may provide valuable insights into its microstructural features. We present the case of a newborn with congenital KHE involving the lumbar and right gluteal region, focusing on its dermoscopic characteristics. The lesion exhibited a homogeneous, grayish-blue background with no discernible vascular structures, a feature that may aid in differentiating KHE from other vascular tumors. The findings contribute to the understanding of dermoscopic patterns in KHE, highlighting its potential role in early diagnosis.

Introduction

Kaposiform hemangioendothelioma (KHE) is a locally aggressive vascular tumor typically presenting at birth or in early infancy. It is characterized by an ill-defined, indurated plaque with a violaceous to erythematous coloration. Unlike infantile hemangiomas, KHE does not undergo spontaneous regression and may be complicated by Kasabach-Merritt phenomenon, a life-threatening consumptive coagulopathy.[1] The diagnosis relies on clinical presentation and histopathology, yet dermoscopy can provide additional, non-invasive insights into its microvascular architecture. While the dermoscopic features of common vascular tumors, such as hemangiomas and vascular malformations, are well described, data on KHE remain scarce. We report a case of congenital KHE in a newborn, with a detailed dermoscopic evaluation to characterize its features.[2]

Case Report

A full-term male neonate presented at birth with a large, firm, and violaceous plaque involving the lumbar and right gluteal region (Figure 1a). The lesion was indurated upon palpation, non-compressible, and had ill-defined borders that gradually blended into the surrounding skin. Its color varied from erythematous to violaceous, with no superficial ulceration or evidence of rapid postnatal growth. The clinical differential diagnosis included KHE, deep infantile hemangioma, and a vascular malformation. Due to the lesion's infiltrative nature and firm consistency, a dermoscopic evaluation was performed to assess its microstructural patterns (Figure 1b).

The lesion also exhibited fine, white, interwoven filamentous structures across the surface, possibly corresponding to areas of stromal fibrosis.

Given the clinical and dermoscopic findings, a biopsy was performed, confirming the diagnosis of KHE. Histopathology revealed a dense proliferation of spindle-shaped endothelial cells arranged in nodules with slit-like vascular lumina. The surrounding stroma exhibited areas of fibrosis and

hemosiderin deposition, consistent with the obliterative and infiltrative nature of the tumor. Immunohistochemical staining showed strong positivity for CD31 and CD34, while GLUT1 was negative, ruling out the diagnosis of infantile hemangioma.[3] These histopathological findings correlated well with the dermoscopic features, where the grayish-bluish background likely reflected the dense endothelial proliferation and stromal thickening, while the fine white filaments corresponded to areas of stromal fibrosis.

Discussion

Dermoscopy is an established tool for evaluating vascular lesions, but its application in KHE remains underexplored. In this case, the most striking dermoscopic feature was the uniform grayish-bluish background, which differed from the bright red, reticular, or globular patterns seen in hemangiomas. The lack of visible vessels and the presence of fine white filamentous structures further supported the obliterative and fibrotic characteristics of KHE. Unlike venous malformations, which typically present with well-demarcated lacunae, KHE exhibited a homogeneous pattern with no discrete vascular elements. This dermoscopic appearance aligns with the histopathological findings of vessel compression and endothelial proliferation, suggesting that dermoscopy may serve as a useful adjunctive tool in the early identification of KHE.[2,4]

As highlighted in a recent review, the dermoscopic features of KHE remain poorly described, with most data limited to isolated case reports or anecdotal observations.[5]

Recognizing the dermoscopic features of KHE is clinically relevant, as it aids in distinguishing this aggressive vascular tumor from benign infantile hemangiomas, which often exhibit well-structured lobular or reticular vascular patterns. The early identification of KHE is crucial, given its potential association with the Kasabach-Merritt phenomenon, which requires urgent medical intervention.[1,3] While histopathology remains the gold standard for diagnosis, dermoscopic findings such as a homogeneous grayish-bluish background, absence of well-formed vessels, and fine white filaments may provide early diagnostic clues, prompting further investigations and timely management.[4]

Conclusions

This case highlights the distinctive dermoscopic features of KHE, characterized by a homogeneous grayish-bluish background, absence of visible vascular structures, and the presence of fine white filamentous patterns. These findings reflect the histopathological characteristics of the tumor, including dense endothelial proliferation, vessel obliteration, and stromal fibrosis. Although dermoscopy is not a substitute for histopathology, it may play a valuable role in the early differentiation of KHE from other vascular lesions, guiding clinical decision-making and facilitating

prompt intervention. Further studies are needed to establish standardized dermoscopic criteria for KHE and explore its potential role in monitoring disease progression and treatment response.[3]

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Figure 1. (**a**) Clinical image of the large, firm, and violaceous plaque involving the lumbar and right gluteal region; (**b**) dermoscopic image showing a uniform grayish-bluish background covering the entire lesion.