Dermoscopic Features of Urticaria Pigmentosa in a Nigerian Child: A Case Report

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ABSTRACT

Urticaria pigmentosa (UP) is the most common subtype of cutaneous mastocytosis (CM), which is a rare benign proliferation of mast cells in the skin. Urticaria pigmentosa presents with red-brown macules, papules, and plaques often located on the trunk, back, and thighs. Lesions of UP, although non-melanocytic may be confused with melanocytic lesions, such as melanocytic nevus and melanoma. Dermoscopy can serve as a useful tool for screening melanocytic and non-melanocytic lesions.

We present a two-year-old girl with several hyperpigmented lesions resembling multiple congenital nevi which turned out to be UP. Dermoscopy showed a dark brown pigment network on a reddish background. A punch biopsy was done which revealed infiltration of the papillary dermis by inflammatory cells which have abundant eosinophilic cytoplasm (mast cells). The patient was counselled and placed on loratadine, an antihistamine. This case describes urticaria pigmentosa in a Nigerian child and its dermoscopic findings, and this highlights the importance of dermoscopy in distinguishing between urticaria pigmentosa (non – melanocytic) and melanocytic mimics.

Keywords: Dermoscopy, Urticaria pigmentosa, Cutaneous mastocytosis, Dermatoscopy

Caractéristiques Dermatoscopiques de l'Urticaire Pigmentaire chez un Enfant Nigérian : un Rapport de Cas

ABSTRAIT

L'urticaire pigmentaire (UP) est le sous-type le plus courant de mastocytose cutanée (CM), qui est une prolifération bénigne rare de mastocytes dans la peau. L'urticaire pigmentaire se présente avec des macules, des papules et des plaques rouge-brun souvent situées sur le tronc, le dos et les cuisses. Les lésions d'UP, bien que non mélanocytaires, peuvent être confondues avec des lésions mélanocytaires, telles que le naevus mélanocytaire et le mélanome. La dermoscopie peut être un outil utile pour dépister les lésions mélanocytaires et non mélanocytaires.

Nous présentons une fillette de deux ans présentant plusieurs lésions hyperpigmentées ressemblant à de multiples nævus congénitaux qui se sont avérés UP. La dermoscopie a montré un réseau pigmentaire brun foncé sur fond rougeâtre. Une biopsie à l'emporte-pièce a été réalisée qui a révélé une infiltration du derme papillaire par des cellules inflammatoires qui possèdent un cytoplasme éosinophile abondant (mastocytes). Le patient a été conseillé et mis sous loratadine, un antihistaminique. Ce cas décrit l'urticaire pigmentaire chez un enfant nigérian et ses résultats dermoscopiques, ce qui met en évidence l'importance de la dermoscopie pour distinguer l'urticaire pigmentaire (non mélanocytaire) des mimiques mélanocytaires.

Mots clés: Dermoscopie, Urticaire pigmentaire, Dermatoscopie, Mastocytose cutanée

Introduction

Urticaria pigmentosa (UP) is the most common subtype of cutaneous mastocytosis (CM). Cutaneous mastocytosis (CM) is a benign proliferation of mast cells in the skin characterized by the release of mast cell mediators such as histamine, leukotriene, prostaglandins, and others.[1] Its clinical presentation varies depending on the subtype. Other subtypes include diffuse cutaneous mastocytosis (erythroderma) and skin mastocytoma.[1,2] In UP, which is the most common pattern seen in children, lesions are characteristically seen on the trunk and

consist of red-brown macules, papules, and plaques that have a widespread distribution but are focused on the trunk. They are absent on the palms, soles, face, and scalp.[3] Lesions of UP, although non-melanocytic, may be confused with melanocytic lesions such as melanocytic nevi and melanoma.

Dermoscopy is a useful tool for screening melanocytic and non-melanocytic lesions obviating the need for unnecessary biopsies.[4] Where the dermoscopic diagnosis suggests a benign lesion like a nevus, a further biopsy may be unnecessary.

There have been no reports of the dermoscopic features of UP in black African skin in our environment. We present a case of a 2-year-old child who presented with several hyperpigmented lesions resembling multiple congenital nevi but which turned out to be UP

Case Synopsis

A 2-year-old child presented with multiple hyperpigmented lesions 3 months after birth. These lesions were initially noticed on her neck as reddish patches but later darkened. They were itchy but not painful. Similar lesions later developed on her arms, legs, trunk, back, and face with the affectation of the palms and soles. There was no family history of similar lesions or associated systemic features such as flushing, wheezing, or diarrhea.

Skin examination of the child showed multiple light brown macules and plaques on the affected areas (Figures 1 and 2). Darier's sign was negative. Systemic examination showed no hepatosplenomegaly or lymphadenopathy. An initial clinical diagnosis was multiple congenital nevi to rule out urticaria pigmentosa presenting on atypical sites. Dermoscopy was done using Dermlite DL 4 3 Gen, USA and photos were captured using iPhone 6S; this showed a dark brown pigment network on a reddish background (Figure 3). A punch biopsy was done which revealed infiltration of the papillary dermis by inflammatory cells which have abundant eosinophilic cytoplasm, distinct cell boundaries, and oval to spindle nuclei (mast cells). Few adnexal structures are seen as well as edema of the papillary dermis. Full blood count results were normal.

She was placed on loratadine syrup 5 mg daily which improved the pruritus. Caregivers were counselled on the diagnosis and the importance of avoiding triggers such as spicy food, opioids, friction, and radioactive contrasts.

Discussion

Urticaria pigmentosa, although a variant of CM, also has other subvariants which include the nodular type and telangiectasia macularis eruptiva perstans (TMEP).[2] The onset of lesions can start from birth and persist to adolescence.[5] Pediatric mastocytosis had a prevalence of 2.5 cases per 10,000 children in Denmark[6] while in Mexico, Kiszwekski et al reported 1:500.[6] In Nigeria, Yusuf reported a case of UP in a 6-month-old child.[7] Ben-Amitai and Metzker found that 11% of 117 patients with UP had a positive family history.[8]

Some studies showed a male preponderance.[6,8] Urticaria pigmentosa is believed to be caused by an activating mutation in proto-oncogene c-KIT which codes the stem cell factor (SCF) receptor, which is considered to be the crucial event in the excessive, SCF-independent mast cell proliferation.[9]

It is characterized by monomorphic macules, papules, plaques, or nodules. Occasionally, it can present with blisters.[9] It typically affects the trunk, thighs, and soles, sparing the palms, soles, and scalp. These skin lesions may be itchy; and systemic complaints such as abdominal pain, diarrhea, palpitations, and syncope can occur as a result of the physiological effects of secreted mast cell mediators. Stroking the lesion elicits Darier's sign, which manifests as a wheal-like edematous erythema.[10] Darier's sign, on the other hand, is negative in some cases of UP.[11]

The diagnosis of UP is made clinically and confirmed by skin biopsy. Mast cell infiltration around blood arteries and within skin appendages may be observed in a microscopic examination. Dermoscopy serves as an important tool to differentiate between the subtypes of cutaneous mastocytosis. In UP, it reveals the existence of a pigmented network pattern with a reddish background. This pigmented network can be attributed to mast cell growth factor-induced enhanced melanogenesis and hyperpigmentation of the epidermal basal layer. [12]

Our dermoscopic findings showed a thick dark brown pigment network on a reddish background which was similar to Chauhan, Bhardway, and Shirazi.[12] Some studies reported the presence of a light brown reticular network.[11,13] Interestingly, a Spanish study noted an additional finding of a light brown blot pattern in about half of the patients with UP.[14] Although pigment network pattern is also observed in melanocytic lesions, the distinguishing feature is the background reddish hue which is absent in melanocytic lesions. This reddish hue is suggested to occur as a result of dermal mast cell degranulation.[12] In addition to urticaria pigmentosa, other non-melanocytic lesions including dermatofibroma, seborrheic keratoses, accessory nipple, and Kaposi sarcoma, also exhibit pigment networks and are common mimics of melanocytic lesions like nevus and melanoma.[14] For the dermoscopic findings of other subtypes of UP such as TMEP, reticular telangiectasia is seen along with brown reticular lines with a reddish background while in mastocytoma, a yellow-orange blot pattern is seen.[13,14]

In Conclusion, we report this case to demonstrate the usefulness of dermoscopy in diagnosing UP and in differentiating it from other melanocytic lesions. Dermoscopy is beneficial in distinguishing between UP (non-melanocytic) and other melanocytic mimics.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the caregiver of the patient has given her consent for her images and other clinical information to be reported in the journal. The caregiver understands that her name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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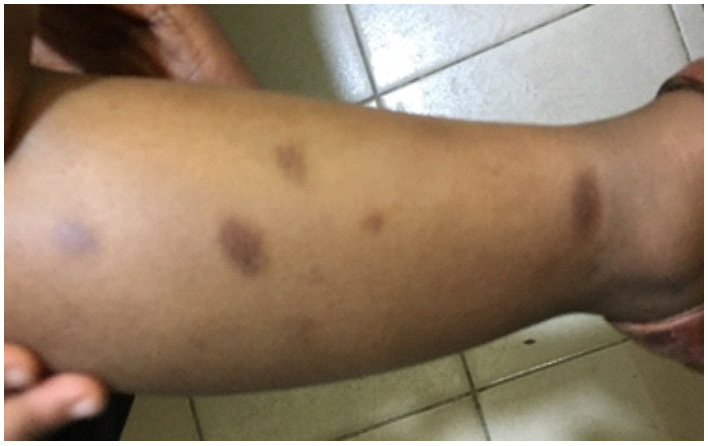

Figure 1 – Brownish plaques on the left leg

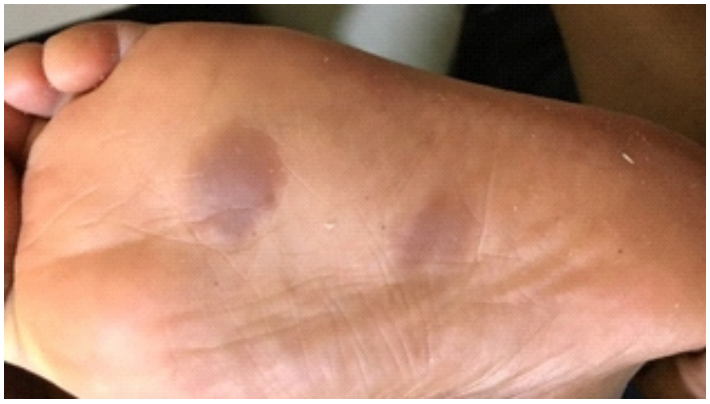

Figure 2 – Light brown plaques on the left sole

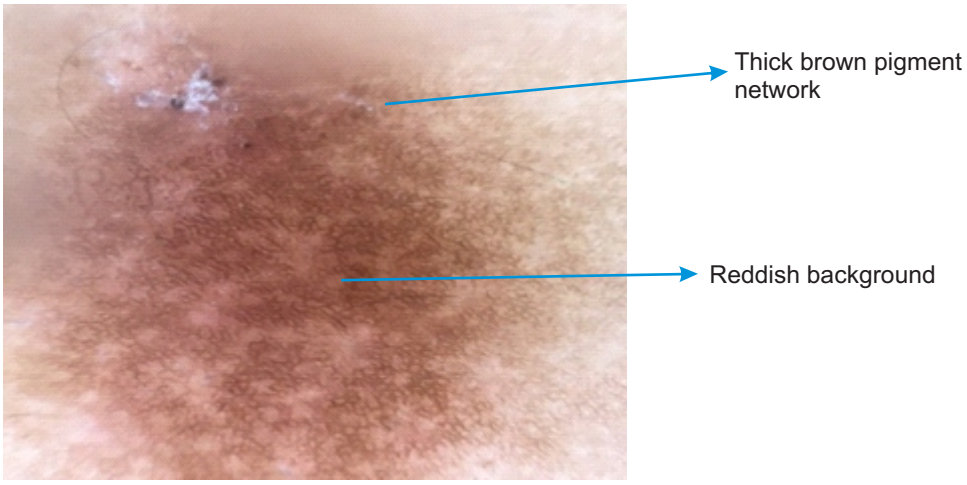

Figure 3 – Dermoscopy of the macule of the left leg revealed a pigment network with a reddish hue. (Derm Lite 3L Gen, USA); polarized mode, 10x magnification