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Purpura annularis telangiectodes of Majocchi: a case report

PATM:a case report

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Abstract

BACKGROUND

Purpura annularis telangiectodes of Majocchi (PATM), also known as Majocchi, is a rare subclass of pigmented purpuric dermatoses (PPD). The etiology of PATM is unknown, but it seems more common in children and young women. The skin lesions are mostly symmetrical ring-shaped reddish-brown macules on the lower limbs.

CASE SUMMARY

A 9-year-old female child. Because of the "repeated reddish-brown ring-shaped rash on both lower limbs for half a year", the patient was treated in our department. These lesions were red brownish annular or petaloid patches with various size, 1-3 centimeters in diameters can be observed on both insteps, ankles and lower limber. And they do not fade when adding pressure, light brownish pigmentation can be observed central of these macules (Figure 1). No feel of infiltration and no atrophy when touching those lesions. The patient had no specific personal and family history. The laboratory findings were normal. Dermoscopy showed a large number of reticular or honeycomb pigmentation in the center of the lesion, and lavender patches and a few focally distributed punctate blood vessels were seen on the edge of lesion (Figure 2). Pathological examination showed scattered vacuolar endothelial cells, infiltration of

lymphocytes and histocytes around blood vessels, and deposition of hemosiderin in papillary dermis (Figure 3). The child was finally diagnosed with PATM. After diagnosis, the patient was given orally vitamin C tablets 0.1g/bid, and topical mometasone furoate cream for external use. We suggested that the girl avoid prolonged stand, as well as strenuous exercises. The skin lesions subsided after 2 wk of treatment. In December 2022, the patient's disease recurred again after intense exercise, and the lesions gradually subsided one month after external medication. Follow-up is ongoing.

CONCLUSION

Although PATM is harmless, cutaneous T-cell lymphoma needs to be ruled out in some cases. Thus, long-term follow-up of PATM is necessary. Dermoscopy, as a non-invasive detection method, has a good corresponding relationship with histopathology, and multi-site observation is more beneficial to the follow-up of children.

Key Words: Pigmented purpuric dermatoses; Majocchi's disease; Dermatoscope; Histology; Case report

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Core Tip: PATM also known as Majocchi's disease, is a rare subclass of PPD. The skin lesions are mostly symmetrical ring-shaped reddish-brown macules on the lower limbs more common in children and young women. The etiology is unknown. The diagnosis of PATM mainly depends on clinical and histopathological features. Dermoscopy, as a non-invasive detection method, has a good corresponding relationship with histopathology, and multi-site observation is more beneficial to the follow-up of children.

INTRODUCTION

PATM, also known as Majocchi, is a rare subclass of PPD. The etiology of PATM is unknown, but it seems more common in children and young women [1]. The skin lesions are mostly symmetrical ring-shaped reddish-brown macules on the lower limbs [2,3]. Thus, some researchers believe that gravity and venous hypertension may be the inducing factors of this disease [4]. The diagnosis of PATM usually depends mainly on the clinical findings and histopathological features. However, different sampling sites or time may affect the pathological diagnosis. And histopathological examination is an invasive method, which is not conducive to long-term follow-up. Dermoscopy, as a non-invasive detection method, has a good corresponding relationship with histopathology, and multi-site observation is more beneficial to observe the disease. Here, we applicated the dermoscopy to observe a girl who suffered from PATM.

CASE PRESENTATION

Chief complaints

A 9-year-old girl admitted to Kunming Children's Hospital, Kunming City, Yunnan Province, China in November 2021 due to the "repeated reddish-brown ring-shaped rash on both lower limbs for half a year".

History of present illness

In the beginning, the lesions were erythematous, where most of them are annular patches, appear in both insteps and ankles. Subsequently, the lesions evolved to both ankles, with sporadic itches. It can recur after topical glucocorticoids. They spread to both legs, with occasional itching. No clinic symptoms of hematuresis, hematocheiza, joint pain or hypodynamia observed during this period of time.

History of past illness

The patient had no history of systemic symptoms, allergies and no specific history of past illness.

Personal and family history

The patient had no specific personal and family history.

Physical examination

Physical examination revealed good general condition. The vital signs were stable, and no other abnormality was found. Dermatological examination showed These lesions were red brownish annular or petaloid patches with various size, 1-3 centimeters in diameters can be observed on both insteps, ankles and lower limber. And they do not fade when adding pressure, light brownish pigmentation can be observed central of these macules. No feel of infiltration and no atrophy when touching those lesions. Such lesions on instep are shown in Figure 1.

Laboratory examinations

Blood routine, urine routine, liver function, kidney function, antinuclear antibody, coagulation function and erythrocyte sedimentation rate tests were normal. These laboratory findings did not point to any specific diagnosis.

Imaging examinations

none

Dermoscopy examinations

Dermoscopy showed a large number of reticular or honeycomb pigmentation in the center of the lesion, and lavender patches and a few focally distributed punctate blood vessels were seen on the edge of lesion (Figure 2).

Pathological examinations

Pathological examination showed scattered vacuolar endothelial cells, infiltration of lymphocytes and histocytes around blood vessels, and deposition of hemosiderin in papillary dermis (Figure 3).

FINAL DIAGNOSIS

Combining clinical and histopathological features, the child was diagnosed with PATM.

TREATMENT

The patient was given orally dipyridamole tablets 25mg/bid, vitamin C tablets 0.1g/bid, and topical mometasone furoate cream and mucopolysaccharide polysulfonate cream bid for external use.

OUTCOME AND FOLLOW-UP

The patient was given orally vitamin C tablets 0.1g/bid, and topical mometasone furoate cream for external use. We suggested that the girl avoid prolonged stand, as well as strenuous exercises. The skin lesions subsided after 2 wk of treatment. In December 2022, the patient's disease recurred again after intense exercise, and the lesions gradually subsided one month after external medication. Follow-up is ongoing.

DISCUSSION

In this case, dermoscopy showed a large number of reticular or honeycomb-shaped pigmentations in the center of the lesion, some lavender patches, and a few focally distributed punctate blood vessels at the edge of the lesion. These structures are often observed in lichen sclerosus and pigmented purpuric dermatosis [5]. The purplish red patches under dermoscopy correspond to red blood cell overflow in the histopathological picture, while the pigmentation corresponds to hemosiderin deposition. It is well known that overflowing red blood cells are engulfed to form hemosiderin. Pigmentation is the final form of purplish-red patches. The early rash of PATM, on histopathology shows swollen vascular endothelial cells in the upper dermis

and dermal papilla, with a large number of lymphocytes and histiocytes around the lumen, extravasation of red blood cells, and occasional neutrophil infiltration. However, the inflammatory infiltration of old lesions is not as obvious as in the early stages, with reduction in extravasation of red blood cells, and deposition of hemosiderin. Although the pathological manifestations of this patient were consistent with those of old lesions, the active margin could be clearly observed under dermoscopy.

PATM often needs to be differentiated from lichen aureus（LA） and purpuric mycosis fungoides (PMF). LA is another subtype of PPD. PMF is a cutaneous lymphoma with purpuric eruptions as the clinical manifestation [6, 7]. The distribution pattern of pigments and purplish red patches under dermoscopy in PATM and LA are not similar. PATM shows a ring-like distribution, while LA shows a diffuse distribution [8]. Neither of them has a special vessel structure under dermoscopy, but PMF has its own characteristic vessels such as spermatozoa-like vessels [9]. Therefore, the differences between PATM, LA and PMF on dermoscopy are clear. In addition, we could distinguish the three diseases.

Due to the detection of epidermotropism or monoclonality in inflammatory infiltrates, other hypotheses believed that PPDs represent a type of T lymphocyte, occult and metaepithelial change [10]. There are even some described cases of progression to mycosis fungoides [11, 12]. To sum up, although PATM is harmless, cutaneous T-cell lymphoma needs to be ruled out in some cases [2]. Thus, long-term follow-up of PATM is necessary. Dermoscopy, as a non-invasive detection method, has a good corresponding relationship with histopathology, and multi-site observation is more beneficial in the follow-up of children.

CONCLUSION

PATM also known as Majocchi's disease, is a rare subclass of PPD. The skin lesions are mostly symmetrical ring-shaped reddish-brown macules on the lower limbs more common in children and young women. The purplish red patches under dermoscopy correspond to red blood cell overflow in the histopathological picture,

while the pigmentation corresponds to hemosiderin deposition. Overflowing red blood cells are engulfed to form hemosiderin. Pigmentation is the final form of purplish-red patches. The early rash of PATM, on histopathology shows swollen vascular endothelial cells in the upper dermis and dermal papilla, with a large number of lymphocytes and histiocytes around the lumen, extravasation of red blood cells, and occasional neutrophil infiltration. However, the inflammatory infiltration of old lesions is not as obvious as in the early stages, with reduction in extravasation of red blood cells, and deposition of hemosiderin. Although the pathological manifestations of this patient were consistent with those of old lesions, the active margin could be clearly observed under dermoscopy. PATM often needs to be differentiated from LA and PMF. The differences between PATM, LA and PMF on dermoscopy are clear. We could distinguish the three diseases. Although PATM is harmless, cutaneous T-cell lymphoma needs to be ruled out in some cases. Thus, long-term follow-up of PATM is necessary. Dermoscopy, as a non-invasive detection method, has a good corresponding relationship with histopathology, and multi-site observation is more beneficial to the follow-up of children.

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