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CASE REPORT

Purpura annularis telangiectodes of Majocchi: A case report

Yun-Jing Pu, Hong-Jing Jiang, Li Zhang

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Abstract

BACKGROUND

Purpura annularis telangiectodes of Majocchi (PATM), also known as Majocchi, is a rare subclass of pigmented purpuric dermatoses. 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 girl, who has received treated in our department, presented with reddish-brown ring-shaped rash on both lower limbs that had been present for 6 mo. These lesions, red brownish annular or petaloid patches, were mostly found on ankles and lower limber, which do not fade when adding pressure and no feel of infiltration and no atrophy when touching those lesions. Pathological examination showed deposition of hemosiderin in papillary dermis. However, dermoscopy showed the pigmentation in the center as well as the lavender patches on the edge of lesion. The child was thus diagnosed with PATM. After diagnosis, we suggested the patient avoid strenuous exercise. she was given vitamin C tablets for oral and mometasone furoate cream for external use. Follow-up examinations and treatment continue to support the clinical diagnosis to date.

CONCLUSION

This is the first report of investigating PATM using dermoscopy, which can differentiate PATM from other diseases due to its unique microscopic feature under dermoscopy. Although PATM is harmless, it still requires long-term follow-up. Moreover, dermoscopy technique can be applied for observation of multi-site lesions and correlated with histopathology. Thus, we believe this approach could be generalized for future diagnosis of PATM.

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

Core Tip: Purpura annularis telangiectodes of Majocchi (PATM) also known as Majocchi's disease, is a rare subclass of pigmented purpuric dermatoses. The skin lesions are mostly symmetrical ring-shaped reddish-brown macules on the lower limbs. This disease is more commonly found among children and young women and the etiology is unknown. Currently, the diagnosis of PATM mainly depends on clinical and histopathological features. Dermoscopy, a non-invasive detection technique, could be a promising technique for future PATM diagnosis owing to its good correlation with histopathology, and multi-site observation.

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INTRODUCTION

Purpura annularis telangiectodes of Majocchi (PATM), also known as Majocchi disease, is a rare subclass of pigmented purpuric dermatoses (PPD). The etiology of PATM is unknown, but it seems more common among 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. Moreover, histopathological examination is an invasive method, which is not conducive to long-term follow-up. Dermoscopy, a non-invasive detection method, has a good corresponding relationship with histopathology and multi-site observation is more beneficial to diagnose the disease. Herein, we applied 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 6 mo".

History of present illness

In the beginning, the lesions were erythematous, where most of them are annular patches, appear on both insteps and ankles. Subsequently, the lesions evolved to both ankles, with sporadic itches. It recurs 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

Normal.

Physical examination

Physical examination revealed good general condition. Dermatological examination results showed that these lesions are red brownish annular or petaloid patches with various size, 1-3 cm in diameters can be observed on both insteps, ankles and lower limber. And they do not fade when adding pressure. During this process, 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.

Dermoscopy examinations

Dermoscopy showed a large number of reticular or honeycomb pigmentation in the center of the lesion,

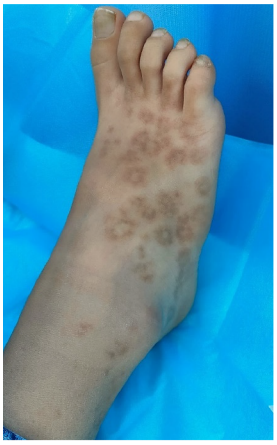

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Figure 1 Macroscopic features of the lesion. The child presented red brownish annular or petaloid patches with various size, 1-3 cm in diameters can be observed on instep.

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 with relevant examinations, the patient was diagnosed as PATM.

TREATMENT

The patient was given orally dipyridamole tablets 25 mg/bid, vitamin C tablets 0.1 g/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 0.1 g/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 data 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[1], and pigmentation is the final form of purplish-red patches. The histopathology results for the early rash of PATM 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

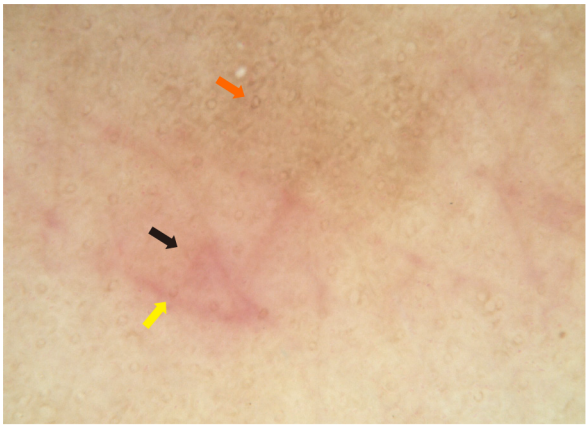

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Figure 2 Dermatoscopic appearance of the lesion. The infiltration method was used (× 50). Dermascopy showed a large number of reticular or honeycomb pigmentation (orange arrow) in the center of the lesion, and lavender patches (black arrow) and a few focally distributed punctate blood vessels (yellow arrow) were seen on the edge of lesion.

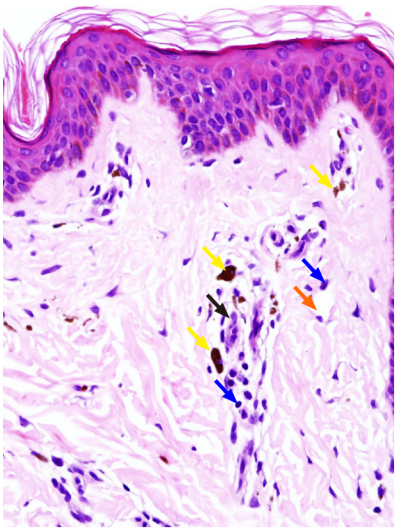

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Figure 3 Histopathological findings. Histopathology showed hyperkeratosis, scattered vacuolar endothelial cells, infiltration of lymphocytes (blue arrows) and histocytes (black arrow) around blood vessels (orange arrow), and deposition of hemosiderin (yellow arrows) in papillary dermis (Hematoxylin eosin staining: Magnification × 400).

clearly observed under dermoscopy.

Differentiating PATM from lichen aureus (LA) and purpuric mycosis fungoides (PMF) could be challenging. LA is another subtype of PPD while PMF is a cutaneous lymphoma with purpuric eruptions as the clinical manifestation[6,7]. PATM and LA can be differentiated by the distribution pattern of pigments and purplish red patches using dermoscopy. 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 unique 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[2,11,12]. Thus, long-term follow-up of PATM is necessary.

CONCLUSION

PATM, a rare subclass of PPD, also known as Majocchi's disease. The skin lesions are mostly symmetrical ring-shaped reddish-brown macules on the lower limbs and more commonly found among 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. According to the histopathology results, the early rash of PATM 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. It is challenging to differentiate PATM from LA and PMF but dermoscopy enables us to visualize the special vascular structure and pigment distribution pattern and distinguish these three diseases. Although PATM is harmless, the disease is prone to relapse and may resemble the early clinical feature of T-cell lymphoma. Thus, long-term follow-up of PATM is crucial. As a non-invasive detection method, Dermoscopy enables us multi-site observation and to correlates the obtained images with histopathology, which could be a promising approach for future PATM.

FOOTNOTES

Author contributions: Pu YJ and Jiang HJ contributed to the work equally; Pu YJ carried out the studies and drafted the manuscript; Jiang HJ and Zhang L participated in its design and helped to draft the manuscript; All authors read and approved the final manuscript.

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