Thyroid Dysfunction and Linear Pigmented Purpuric Dermatosis in A 15-Year-Old Girl

YAHYA Aishatu M
North West University, Kano, Nigeria.
Corresponding Author: Aishatu Yahya M
Email address: aishayahya35@yahoo.com; ***Phone number:*** +2348035054507

ABSTRACT

Pigmented purpuric dermatoses (PPD) are a group of skin disorders characterized by petechial haemorrhages resulting from capillaritis and are benign and self-limiting. They usually present as remitting and relapsing, non-palpable, flat purpura distributed bilaterally on the legs of the elderly. Linear presentation is rare and occurs mainly among children. The aetiology of PPD is not known, but some factors, such as drugs, physical activity, and capillary fragility, appear as trigger agents.

Pigmented purpuric dermatoses can also be associated with systemic diseases like diabetes mellitus, thyroid dysfunction, systemic lupus erythematosus, rheumatoid arthritis, liver diseases, solid neoplasms, mycosis fungoides, etc. We present the case of a 15-year-old girl with PPD associated with thyroid dysfunction.

Conclusion: Pigmented purpuric dermatoses with linear presentation are rare and usually occur in children with a unilateral distribution. A high index of suspicion aids diagnosis, and with atypical presentations, other diseases like thyroid disorder, vasculitis, collagen vascular diseases, and coagulopathies should be screened for.

Keywords: Pigmented purpuric dermatosis, Thyroid dysfunction, Children

Dysfonctionnement thyroïdien et dermatose purpurique pigmentée linéaire chez une fille de 15 ans

Résumé

Les dermatoses purpuriques pigmentées (DPP) sont un groupe de troubles cutanés caractérisés par des hémorragies pétéchiales résultant d'une capillarite et sont bénignes et auto-limitantes. Elles se présentent généralement sous la forme d'un purpura plat, non palpable, rémittent et récidivant, distribué bilatéralement sur les jambes des personnes âgées. La forme linéaire est rare et survient principalement chez les enfants. L'étiologie de la DPP n'est pas connue, mais certains facteurs, tels que les médicaments, l'activité physique et la fragilité capillaire, semblent être des agents déclencheurs.

Les dermatoses purpuriques pigmentées peuvent également être associées à des maladies systémiques comme le diabète sucré, un dysfonctionnement thyroïdien, le lupus érythémateux disséminé, la polyarthrite rhumatoïde, les maladies hépatiques, les néoplasmes solides, le mycosis fongoïde, etc. Nous présentons le cas d'une fille de 15 ans atteinte de PPD associée à un dysfonctionnement thyroïdien.

Conclusion : Les dermatoses purpuriques pigmentées à présentation linéaire sont rares et surviennent généralement chez les enfants avec une distribution unilatérale. Un indice de suspicion élevé facilite le diagnostic, et en cas de présentation atypique, d'autres maladies comme un trouble thyroïdien, une vascularite, des maladies vasculaires du collagène et des coagulopathies doivent être recherchées.

Mots-clés : dermatose purpurique pigmentée, dysfonctionnement thyroïdien, enfants

Introduction

Pigmented purpuric dermatosis (PPD) describes a group of different sub-types of benign, chronic purpuric skin eruptions.[1] They are characterized by red or purple macules, patches, and petechiae with extravasation of erythrocytes and deposition of hemosiderin in the dermis of the affected skin, leading to red-brown or golden-brown discolouration as the hemosiderin is absorbed.[1] The lesions occur mainly on the lower extremities and

sometimes on the arms and are often asymptomatic but can occasionally present with mild itching.[1]

The aetiology of PPD is unknown, but some factors such as drugs, physical activity, and capillary fragility appear to be trigger agents.[2] Cell-mediated immunity is also implicated in the pathogenesis and may contribute to vascular fragility.[2] Perivascular infiltration of lymphocytes and dendritic cells, which interact with vascular endothelial cells, can affect the permeability of the micro-vasculature.[2] PPD can also be associated with systemic diseases like diabetes mellitus, thyroid dysfunction, systemic lupus erythematosus, rheumatoid arthritis, liver diseases, solid neoplasms, mycosis fungoides, etc.[3] PPD are not associated with coagulopathies or thrombocytopenia.[3]

There are many clinical variants, but Schamberg disease and lichen aureus are the most common types in children.[1] Other clinical variants include Purpura Annularis, Telangiectodes of Majocchi, Pigmented purpuric lichenoid dermatitis of Gougerot and Blum, Eczematis-like Purpura of Doucas and Kapetanakis, and Granulomatous pigmented purpura.[4]

Linear-pigmented purpura is uncommon and occurs mainly in children and adolescents.[1]It is described as quadrantic capillaropathy or unilateral linear capillaritis.[1] The lesion is similar to lichen aureus and Schamberg disease but is unilaterally distributed and linear.[1] Other dermatoses with linear, blaschkoid, or dermatomal-like distribution must be differentiated from linear PPD. [1] These include lichen aures (segmental variant), unilateral nevoid telangiectasia, angioma serpenginosum, linear epidermal nevi, linear morphea, lichen striatus, or linear lichen planus.[1]

Purpuric dermatosis occurs in all races and is more common in men except for Majocchi disease, which is more common in women.[1] They predominantly affect adults, although cases have been reported in children.[1]

Schamberg disease is the most common type.[1] It affects all ages but commonly occurs in middle-aged to older men and is uncommon in children. It is usually localized to lower limbs but can occur on the thighs, buttocks, trunk, and arms.[5] It is a chronic-persistent flexural dermatosis that can sometimes extend to the proximal areas. Most patients are asymptomatic, but some have mild itching. [5] Clinically, it appears as pinpoint petechiae with a 'cayenne pepper' – like appearance, as yellow-orange-brown patches with an oval to irregular outline.

Lichen aureus, also known as lichen purpuricus is a more localized variant of PPD.[6] The lesions are persistent and typically small in number and solitary.[6] It is characterized by the sudden appearance of small yellow-orange papules with lichenoid appearance and a tendency to coalesce into plaques measuring about 1 to 20cm with associated millimetric purpuric lesions.[6] It primarily affects the lower extremities but can occur on any body part.[6] They are usually asymptomatic. In children and adolescents, the zosteriform and segmental variants that follow the lines of Blaschko or have a saphenous course have been described.[7]

Unilateral naevoid telangiectasia (UNT) is a rare condition that can be congenital or acquired.[8] The rare congenital form is autosomal dominant, is more common in males, and occurs shortly after the neonatal period.[8] The acquired form occurs exclusively in young female patients with physiologic conditions.[8]It is characterized by multiple, unilateral linear arranged blanching telangiectases in a dermatomal distribution (mostly involving the third and fourth cervical dermatomes) or Blaschkoid pattern. It has a predilection for the face, neck, shoulder, arm region, and thorax. [8] A pale ring, an anaemic halo, may be observed surrounding the telangiectases.[8] The reason for the strict unilateral distribution of the disease is not apparent. However, it is due to a localized increase in estrogen receptors caused by unmasking chromosomal mosaicism at times of relative estrogen excess.[9] Histologically, multiple dilated, thin-walled vessels are lined by plump endothelial cells in the papillary and upper reticular dermis, and there are no signs of neo-genesis.[9] Unilateral naevoid telangiectasia typically persists, but rarely, the acquired cases resolve spontaneously if the causative factor is removed.[8]

Angioma serpiginosum is a rare dermatologic condition characterized by pinpoint violaceous to coppery-red punctate maculopapular eruptions that cluster together in linear, serpiginous (snake-like) or gyrate (ring) patterns on an erythematous background.[10] It affects the lower limbs mainly and is

more frequently seen in females.[10] The exact pathophysiology is unknown, but the condition may evolve from the proliferation of endothelial cells and the formation of new capillaries, not only the dilation of pre-existing capillaries.[11]

Linear epidermal nevi have features of hyperkeratosis, acanthosis, and papillomatosis on histology and are often present in younger age groups.[12]

Linear morphea and lichen striatus are seen mostly among children, and skin biopsy shows evidence of sclerosis in linear morphea and peri-eccrine lymphocytic inflammation in lichen striatus.[13,14]

Linear lichen planus, unlike linear PPD, has significant pruritus, a deep purple colour, and hyperpigmentation on clinical examination.[13] Histopathology reveals a lichenoid band of inflammation in papillary dermis with epidermal hyperplasia and hypergranulosis.[14]

Here, we report a 15-year-old girl with unilateral linear PPD associated with thyroid dysfunction.

Case Report

A 15-year-old girl was referred from the endocrinology clinic, where she is managed for thyroid dysfunction with a three-month history of unilateral purpuric rash on the chest, back, and extremities. The rash first appeared on the chest and later on the hands, arms, and back. There was no history of trauma, and rashes appeared before she commenced on propranolol and carbimazole. The lesions appeared after the symptoms of thyrotoxicosis.

On examination, unilateral brown linear lesions were observed on her chest, back, and left arm. Other clinical features of hyperthyroidism observed include palpitations, tremors, weight loss, warm skin, increased appetite, bulging of the eyes, and body weakness.

Full blood count, ESR, renal and liver function tests, thyroid function tests, coagulation profiles, and viral serology for hepatitis B and C were done.

Thyroid function test showed elevated T3 - 8.2pg/ml (2.50-4.64pg/ml), T4 - 2.9 (0.95-1.52ng /ml), and low TSH 0.12 μu /ml (0.32-3.0). Other tests were essentially normal. A skin biopsy taken from the lesion site showed a superficial mild perivascular lymphocytic infiltrate with extravasation of erythrocytes and no sign of vasculitis. Diagnosis of unilateral linear PPD associated with hyperthyroidism was made, and the child was counselled and placed on antihistamines, topical corticosteroids, and vitamin C and commenced on anti-thyroid medications. Her comorbidity of hyperthyroidism and financial constraints limited the use of other alternative methods of treatment. The lesions did not significantly improve after about four months of treatment. Therefore, the patient was counselled on the benign nature of the disease.

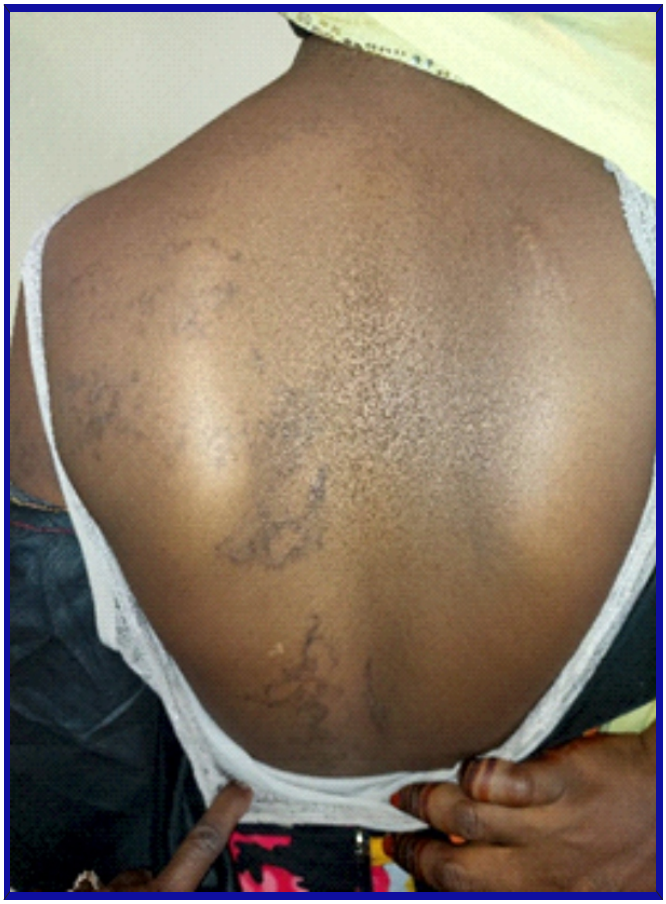

Figure 1: Linear brown lesions on the back of the patient

Discussion

Unilateral linear capillaritis (ULC) is a rare, unique variety of pigmented purpuric dermatosis (PPD) characterized by linear or segmentally distributed pigmented purpuric macules, mainly on the lower extremities. Atypical presentations on other sites can occur.[4] They are usually asymptomatic. The majority of cases of PPD are idiopathic. Still, some are associated with systemic diseases like diabetes

mellitus, thyroid dysfunction, systemic lupus erythematosus, rheumatoid arthritis, liver diseases, solid neoplasms, mycosis fungoides, etc.[2] Our patient presented with a lesion which clinically characterized PPD. The patient also has an associated systemic disease, which is thyroid disorder. Most cases of PPD are asymptomatic but can present with mild pruritis.[1] Our patient presented with mild pruritus. Despite the different clinical features of all types of PPD, they have the same histopathologic features: dilated blood vessels, endothelial cell swelling, hemosiderin deposits, red cell extravasation, and perivascular lymphocytic macrophage infiltration.[5] Skin biopsy taken from the site of the lesion of our patient showed superficial mild perivascular lymphocytic infiltrate with extravasation of erythrocytes with no sign of vasculitis, which is a feature of PPD.

The doctors requested that Full blood count and peripheral blood smear exclude thrombocytopenia and coagulation studies to exclude other causes of purpura. There were no abnormalities from the above investigation on our patient. Standard guidelines for the treatment of PPD are not available.[15] They are usually persistent and resistant to treatment, as observed in our patients.[15] Watchful waiting is done for asymptomatic cases.[1] But when symptomatic or with diffuse distribution, treatment is required.[15]

Treatment modalities include topical corticosteroids, phototherapy, vitamin C, rutoside, and laser therapy in the pruritic forms or those with evident erythema.[7] Since our patient was symptomatic, we administered antihistamines, topical corticosteroids, and vitamin C. Financial constraints and her comorbidity of thyroid dysfunction prevented us from giving other medications.

Piyush Kumar et al.[16] reported a 6-year-old girl who presented with unilateral linear PPD. Her lesions were on the medial aspect of the left leg. She was asymptomatic and without features of any underlying systemic disease, as is in the case of our patient. Histology confirmed the diagnosis, and dermoscopy showed red dots and clods due to extravasation of blood. No therapy was given in their case, but the patient was counselled.

Similarly, Gabriela P et al. [17] reported a 7-year-old girl who presented with a 6-month history of a symptomatic lesion on her leg with no remarkable medical history. Physical examination showed red-brown macules extending in linear distribution on her left leg, which did not follow dermatome or blaschko lines. Histology confirmed the diagnosis of linear PPD. She was treated with mometasone furoate 0.1% cream. After 15 months of treatment, the hyperpigmentation was still evident.

Conclusion: Linear PPD is rare and may be associated with systemic disease. A high index of suspicion is required for diagnosis, and response to treatment is poor.

REFERENCES

1. Sardann K, Sarkar R, Sehgal V. Pigmented dermatoses: An overview. *Int. J. Dermatol.*1999;41:827-9.
2. Von den Driesch P, Simon M. Cellular adhesion antigen modulation in purpura pigmentosa chronica. *J Am Acad Dermatol.*1994;30:193-200.
3. Newton R, Raimer S. Pigmented purpuric eruptions. *Dermatol Clin*. 1985;3(1):165-9.):165-9.
4. Spigariolo C, Giacalone S, Nazzaro G. Pigmented purpuric dermatoses: a complete narrative review. *J Clin Med* 2021; 10(11):2283.
5. Bonnet U., Sella C, Isbruch K. Recurrent Purpura due to Alcohol-related Schambergs disease and its association with serum immunoglobulins: A longitudinal observation of a heavy drinker. *J Med* Case Rep.2016; 10:301.
6. Ratnam KVSu, WPDPeters MS. Purpura simplex (inflammatory purpura without vasculitis): a clinicopathological study of 174 cases. *J AM Acad Dermatol* 1991; 25(4)642-47.
7. Price ML Jones, EWCalnan CD, MacDonald DM. Lichen aureus: a localized, persistent form of pigmented purpuric dermatitis. *Br J Dermatol* 1985;112(3)307-14.
8. Hynes L, Shenefelt P. Unilateral nevoid telangiectasia: Occurrence in two patients with hepatitis C. *J Am Acad Dermatol*. 1997; 36:819.

9. Sadick N, Urmacher C. Estrogen and progesterone receptors: Their role in post sclerotherapy angiogenesis telangiectatic matting. *Dermatol Surg* 1999: 25:539-43.
10. Bishara M, Jiaravuthisan M, Weinstein M. A 13-year-old presenting with recurrent angioma serpiginosum. *J Cutan Med Surg* 2018;22: (5):511-13.
11. Dhanta A, Taneja G, Hazarika N, Joshi P. Clinicopathological and dermoscopic features in Blashkoid angioma serpiginosum. *Dermatol Pract Concept*. 2021;11(1): e2021107.
12. Kim J, Kim Y. Zosteriform unilateral linear capillaritis. *Ann Dermatol* 2021; 33: 298-300.
13. Hoesly P, Cappel M, Sluzevich J. Persistent dermatomal eruption on a leg. *Dermatol online J* 2019;25.
14. Rodriguez-Garijo N, Tomas-Velazquez A., Estenaga A, Antonanzas J, Idoate MA, Espana A. A case of unilateral linear capillaritis. *Pediatr Dermatol* 2020;37:750-51.
15. Pallas M, Del Mazo RC, Biosca VL. Pigmented Purpuric Dermatosis: a review of literature. (accessed 4th February 2021) https://pubmed.ncbi.nml.nih.gov/31983388)
16. Piyush Kumar, Chirag Desa, Anupam Das. Unilateral Linear capillaritis. *Indian Dermatol Online J* 2021; 12:486-87.
17. Gabriela P, Elena R, Laura S, Jose-Manuel S, Alejandra P, Hugo .V. Unusual pigmented purpuric dermatosis with linear distribution and granulomatous inflammation. *Indian J Dermatol* 2021; 66:224