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Review Article

Schamberg Disease

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Abstract *- In this clinical case, we attend to a patient with pigmented purpuric dermatosis, diagnosed with Schamberg disease. It is the most frequent type of all pigmented purpuric dermatosis. It is a benign, idiopathic, harmless skin condition, but its importance is mainly due to these characteristics. Good general practitioners should be able at least to consider a differential diagnosis whenever a patient shows a pigmented purpura. We update the most relevant information about Schamberg disease (concept, epidemiology, aetiology, pathophysiology, clinical features, diagnosis, investigations, associated diseases, treatment and management, prognosis and complications) with references where the reader can easily find more specific information.*

Keywords *- Schamberg disease, Pigmented purpura, Pigmented dermatosis.*

1. Clinical case report. Schamberg disease

A 31-year-old male attends our consult. He has no relevant past medical history, no known drug allergies, and is not on any chronic medication or supplement.

He has a five-week skin rash, asymptomatic in both distal legs, unrelated to exercise or any known issue. Approximately one year ago, he had a similar rash in the same places that subsided with a corticosteroid cream. This time the rash is not getting better. No other signs or symptoms

On physical examination, we can observe petechial lesions in both distal legs, mainly on the right side (See Figures 1, 2 and 3).

With the clinical suspicion of Schamberg disease, he is referred to a private consultant dermatologist.

In the discharge letter, Schamberg disease is confirmed by the dermatologist; no biopsy was needed.

2. Other synonyms

Progressive pigmented purpuric dermatitis, capillaritis, itching purpura, pigmented purpuric eruption.

3. What is Schamberg disease?

Schamberg disease (SD) is the most frequent type of pigmented purpuric dermatoses. It is a benign, idiopathic, harmless skin condition characterised by reddish-brown patches caused by leaky capillaries. [1]

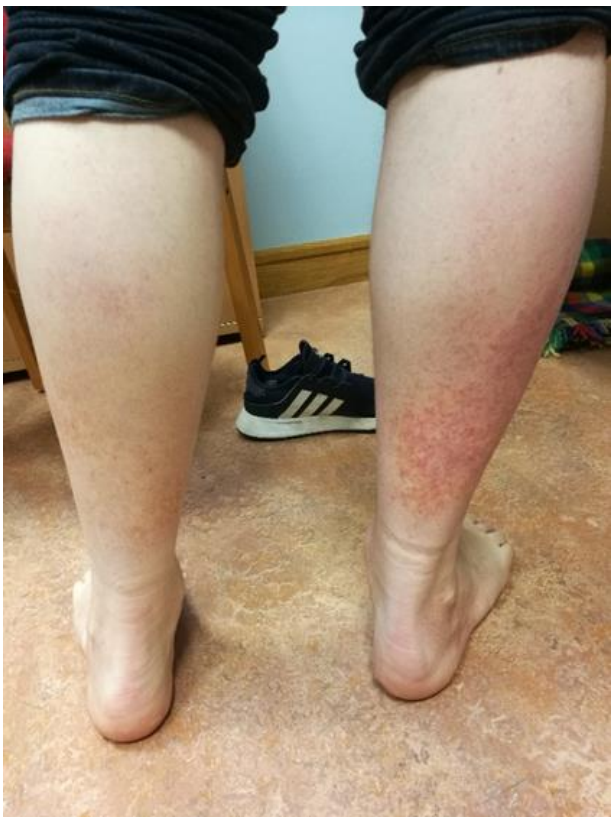

Fig. 1

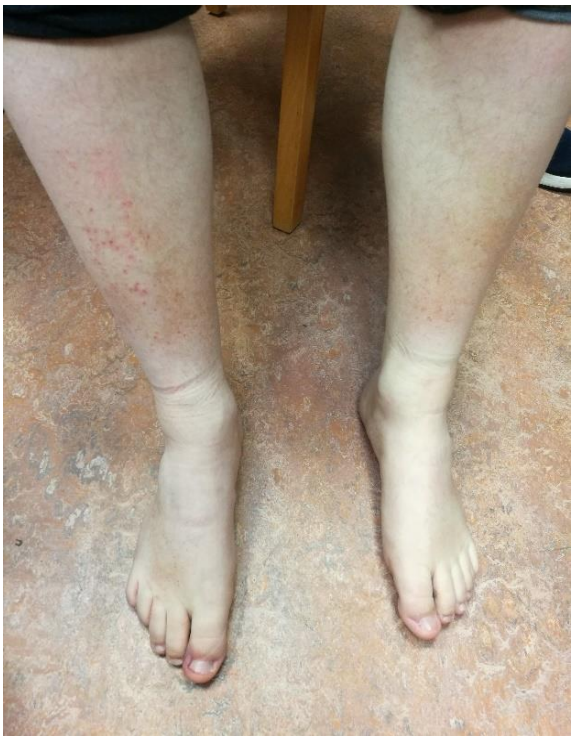

Fig. 2

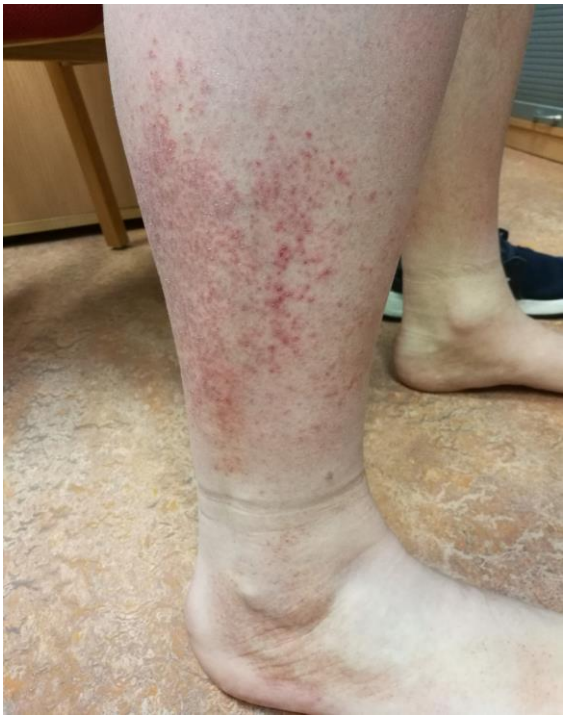

Fig. 3

4. Epidemiology

SD is not a very frequent disease, with few epidemiological studies published. SD may affect all ages but more frequently in middle-aged to older men and less frequently in children. [2]

An Indian outpatient clinic study of 55,323 patients diagnosed 100 cases of pigmented purpuric dermatosis (0.18%). Of these 100 patients, 95 had SD. It is more frequent in males (4:1). The age range is from 11 to 66 years, with a mean of 34 years. [3]

A report of four family members with SD had been published, thus suggesting a possible genetic link (at least, in some cases). [4]

5. Aetiology

SD is considered idiopathic, but the underlying cause of capillary inflammation is unknown. Aetiology looks multifactorial, and potential contributors can be genetic causes, alcohol intake, gravitational dependency, viral infection, hepatitis B antigenemia, dysfunctional immune system, drugs (aspirin, paracetamol, thiamine, amlodipine, glipizide, nitroglycerin, chlordiazepoxide, bezafibrate and sildenafil). [1, 5].

6. Pathophysiology

The main pathophysiological mechanism of SD is the extravasation of erythrocytes, and the deposition of hemosiderin in macrophages with a T-cell lymphocytic infiltrate perivascularly. [5]

The capillary permeability increases due to oedema and endothelium damage. Among other factors that can worsen this process it had been mentioned: venous stasis, exhausting exercise, gravitational dependency, capillary fragility, infections…

7. Histopathology

SD shows perivascular T-cell lymphocytic infiltrate around superficial small blood vessels. This infiltrate leads to endothelial cell swelling and narrowing of the lumen. [5]

8. Clinical features

SD is characterized by petechiae, purpura, and increased skin pigmentation (brown, red, or yellow patchy). On physical examination, we can see non-blanching purpura, asymmetrical brown/orange patches and a type of petechiae called “cayenne pepper spots” (developing at the edge of older lesions). The patterns can vary (annular, linear…) and can be associated with lichenification, scaling and pruritic marks. [5, 6]

It is more frequently located bilaterally on the lower limbs and not rarely in the thighs, buttocks, trunk, or upper extremities. It can be anywhere or even unilateral. It has been reported one case involving the genitals.

SD is usually asymptomatic but can cause itchiness and blotchy skin appearance. It can produce psychological distress in some patients. When a patient complains of limb pains, they are usually coincidental. [1]

9. Diagnosis

SD diagnosis is usually made on clinical features, observing and identifying the classic morphology of the lesions. Dermatoscopy can help, as the petechiae can be seen more clearly. [1]

The biopsy is not mandatory, but it may be required to rule out the differential diagnosis, mainly mycosis fungoides. [5] It should be requested, when needed, by the dermatologist.

Differential diagnosis should be made from other causes of pigmented purpura: exercise-induced capillaritis, drug eruption, local trauma, self-induced purpura, vasculitis (leukocytoclastic), T-cell lymphoma (especially if presenting in young males), lichen aureus, Majocchi purpura, mycosis fungoides. [1, 5]

10. Investigations

SD is not associated with any laboratory abnormality. Nevertheless, we have to request some blood tests to exclude other causes of purpura: Full blood count and coagulation screen. Other recommended blood tests include autoantibody screen (antinuclear antibodies, rheumatoid factor) and hepatitis B / hepatitis C serology.

When the diagnosis is unclear, dermatologists can perform a skin biopsy. [1, 5, 6]

11. Associated diseases

SD had been associated, among others, with diabetes mellitus, rheumatoid arthritis, systemic lupus erythematosus, porphyria, hepatitis B, malignancies, thyroid abnormalities…. [5, 7]

12. Treatment and management

As SD is idiopathic, there is no established aetiologic therapy. Any suspected precipitants should be withdrawn. [5, 6]

As SD is a mild condition and usually asymptomatic, most patients do not require any treatment. There is no curative treatment. [8]

The pruritus can be managed with mild topical corticosteroid or oral antihistamines. [8]

Precocious treatment on superimposed infection, with proper antibiotics.

When the lower leg is affected, wearing graduated compression elastic support hosiery to prevent venous stasis could help. [6]

Despite oral corticosteroids could provide some benefits, they should be used with much caution because, in SD, the benefits are outweighed by their systemic adverse effects.

Phototherapy can clear SD, but it does not prevent exacerbations.

Other uncommon treatments, only for use by specialists, may involve narrow-band ultraviolet light, advanced fluorescent technology, aminaphtone, immunosuppressants and PUVA treatment. [1, 5, 6]

Multiple other therapies had been employed in SD, but with extremely little evidence: vitamin C, bioflavonoids, griseofulvin, cyclosporine, methotrexate, vascular lasers…[5]

13. Prognosis

We must reassure our patient that SD is a benign disease that usually runs a chronic course with exacerbations and remissions. The rash and residual pigmentation may be present for many years with slow extension or recurring. Occasionally it may also disappear spontaneously within a few weeks. [1, 5]

It was reported that about 60% of patients cleared their lesions completely, whilst 40% remained stable or developed worsening symptoms. [9]

14. Complications

Infrequently, it can get worse, with recurrent chronic skin lesions, occasionally resistant to treatment.

The development of T-cell lymphoma in patients with SD is extremely uncommon, but it has been reported. [10]

It is also very rare to develop mycosis fungoides after an initial diagnosis of SD. Nevertheless, we should suspect mycosis fungoides whenever the course lasts longer than one year. [5, 11]

Some patients, usually young females, can develop psychological trauma and emotional stress secondary to the longstanding pigmentation in the skin, leading to anxiety and depression.

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