Schamberg's disease (Benign dermatoses with pupura)

What is Schamberg's disease?

Synonyms: progressive pigmented purpuric dermatitis, Gougerot-Blum capillaritis, itching purpura, pigmented purpuric eruption

Schamberg's disease is the most common type of the pigmented purpuric dermatoses (PPDs). It is chronic, benign, cutaneous eruptions characterised by petechiae, purpura and increased skin pigmentation (brown, red, or yellow patchy) [1] . The PPDs are grouped into:

- Progressive pigmentary purpura or Schamberg's disease.
- Pigmented purpuric lichenoid dermatitis of Gougerot and Blum - red/brown papules and plaques in men - which responds to psoralen combined with ultraviolet A (PUVA) treatment.
- Purpura annularis telangiectodes - rare, with a preponderance in young females and manifesting as annular erythematous plaques and patches.
- Eczematoid-like purpura of Doucas and Kapetanakis - occurring in men, with bilateral intensely itchy lesions on legs.
- Lichen aureus - a localised persistent form of pigmented purpuric dermatitis.
- Itching purpura of Lowenthal (disseminated pruriginous angiodermatitis) - rare (like Schamberg's disease) but accompanied by itching.

There is clinical and histological overlap between these and they may actually represent variable presentations of the same disease process.

Who is affected by Schamberg disease? (Epidemiology)

Schamberg's disease is rare and there are few epidemiological studies in the literature.

Schamberg disease may affect all ages but commonly occurs in middle-aged to older men and less frequently in children [2] .

One study of patients attending an Indian outpatient clinic over an 18-month period found that there were 100 cases of pigmented purpuric dermatosis in a total of 55,323 patients (0.18%). 95 of these patients had Schamberg's disease. The male-to-female ratio was 3.8:1. The age range was from 11-66 years with a mean of 34 years [3] .

There has been a case report of four family members with Schamberg's disease, suggesting a possible genetic link [4] .

Schamberg's disease causes (aetiology) [5] [6] [7]

The underlying cause is not known. However, the following have been postulated:

- Recent viral infection.
- Hypersensitivity to an unknown causal agent.
- Aberrant cell-mediated immunity (perivascular infiltrate has specific types of CD cells only).
- Associated with certain medications - thiamine, aspirin, chlordiazepoxide and paracetamol. It has also been reported with bezafibrate and amlodipine [8] .

Schamberg's disease symptoms [7]

There are no symptoms of Schamberg's disease apart from itching and patients noting their skin looks blotchy. For some this is enough to cause psychological distress. However, some patients have reported pains in their limbs - which may be coincidental.

Lesions are most commonly on the lower limbs bilaterally but can occur anywhere or be unilateral. A case involving the genitals has been reported.

The lesions consist of:

- Asymmetrical brown/orange patches.
- Non-blanchable purpura.
- Petechiae called 'cayenne pepper spots' (develop at the edge of old lesions).

Patterns can vary - eg, annular, linear. There may also be associated lichenification, scaling and pruritic marks.

Differential diagnosis[6] [9]

Other causes of pigmented purpura:

- Vasculitis - eg, leukocytoclastic vasculitis.
- T-cell lymphoma (especially if presenting in young males).
- Drug eruption.
- Trauma.
- Self-induced purpura.
- Mycosis fungoides[10] .
- Primary benign hypergammaglobulinaemic purpura of Waldenström[11] .

Investigations

- Blood tests - including platelets and clotting - are usually normal.
- Autoantibody screen and hepatitis serology should be performed.
- Skin biopsy - histology reveals a capillaritis of dermal vessels. Other changes that may be seen include perivascular inflammatory infiltrate, endothelial hypertrophy with extravasation of blood cells and haemosiderin-laden macrophages[12] .

- Examination of the skin, using a dermatoscope, may be helpful although there are limited reports in the literature [13] .

Associated diseases [5] [6]

- Diabetes mellitus.
- Rheumatoid arthritis.
- Systemic lupus erythematosus.
- Thyroid abnormalities.
- Hepatic disease, including hepatitis B.
- Porphyria.
- Malignancies.
- Dyslipidaemias.

Schamberg's disease treatment and management [5] [7]

Any suspected precipitants should be withdrawn.

- Pruritis - treat with mild topical corticosteroid or antihistamines.
- Good results have been obtained with narrow-band ultraviolet light.
- One study reported the successful use of aminaphtone, a drug normally used in other venous conditions such as chronic venous congestion of the lower limbs [14] .
- Superimposed infection - will need antibiotics.
- Systemic steroids provide some benefits but these are outweighed by the risks of systemic side-effects.
- Advanced fluorescent technology has produced some promising cosmetic results [15] .

Other tried treatments include vitamin C supplements, laser therapy and wearing support hosiery to prevent venous stasis. There is no evidence of definite benefit of the former two. Immunosuppressants have also been used [6] . PUVA treatment has been used successfully [2] .

Prognosis

Schamberg's disease usually runs a chronic course with frequent exacerbations and remissions. The rash may be present for many years with slow extension. Pigmented purpura may occasionally disappear spontaneously.

The development of T-cell lymphoma in patients with Schamberg's disease has been reported [16] .

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