Pigmented Purpuric Dermatoses

The pigmented purpuric dermatoses form a group of chronic skin diseases with a characteristic appearance. The aetiology is unknown but they tend to affect the lower limb and there is extravasation of erythrocytes in the skin with marked deposition of haemosiderin.

There are a number of clinical patterns but all with a similar histological appearance. Generally, the treatment and prognosis are similar. The pigmented purpuric dermatoses include:

- Schamberg’s disease (progressive pigmentary dermatosis).
- Eczematoid-like purpura of Doucas and Kapetanakis.
- Majocchi’s disease (purpura annularis telangiectodes).
- Pigmented purpuric lichenoid dermatosis of Gougerot and Blum.
- Lichen aureus.
- Itching purpura.

Many consider itching purpura and eczematoid-like purpura to be variants of Schamberg’s disease.

Pathophysiology

The aetiology is unknown but important contributory factors include venous hypertension, exercise and gravitational dependency. Histology shows a perivascular lymphocytic infiltrate of T cells centred on the superficial small blood vessels of the skin, suggesting that cell-mediated immunity plays a part. There is swelling of endothelial cells and narrowing of the lumen. Extravasation of red blood cells occurs with marked deposition of haemosiderin in macrophages, and a rare granulomatous variant of chronic pigmented dermatosis has been reported.

Epidemiology

The conditions are uncommon. Generally they affect males more than females, except in Majocchi’s disease. There is no racial difference in incidence.

- Schamberg’s disease occurs at any age.
- Itching purpura and the dermatosis of Gougerot and Blum typically affect middle-aged men.
- Majocchi’s disease occurs mostly in children or in young adults.
- Lichen aureus occurs mostly in young adults but rare cases affecting children have been reported.

Both Schamberg’s disease and Majocchi’s disease have rarely been reported in families, suggesting a genetic aetiology or predisposition. It may be autosomal dominant.

Risk factors

Some forms of the pigmented purpuric dermatoses are associated with the ingestion of drugs, including non-steroidal anti-inflammatory agents, sedatives, antihypertensives, drugs with antihistaminic properties and lipid-lowering agents. Cases associated with sildenafil and isotretinoin have been reported. Lichen aureus may be associated with trauma.

Types of pigmented purpuric dermatoses
Schamberg's disease

- Schamberg's disease is the most common form of pigmented purpura and is the most common cause of petechiae in children. It is most commonly seen in the late teens and early 20s, although it has been diagnosed in children aged between 1 and 9 years [9]. It is more common in males than in females, although a report of children included three boys and ten girls.

See separate Schamberg's disease article for further details.

Eczematoid-like purpura of Doucas and Kapetanakis

- This disorder is similar to Schamberg's disease but occurs predominantly in adult males.
- The lesions, which may be itchy, commonly begin on the lower legs and progress to the thighs, trunk and upper limbs.
- It is a chronic relapsing disorder which may remit spontaneously.
- Lichenification may also be present due to chronic scratching of the pruritic lesions.

Majocchi's disease (purpura annularis telangiectodes)

- Majocchi's disease typically presents with annular erythematous plaques and patches which have central areas of atrophy, commonly in a symmetrical distribution on the lower limbs but occasionally on the trunk and upper limbs.
- Girls are more often affected than boys, most often in adolescence or early adulthood.
- Despite a chronic relapsing course which lasts several years, the disorder is benign and self-limiting and treatment is neither effective nor required.
- It is associated with haematological conditions and rarely T-cell cutaneous lymphoma [10].

Pigmented purpuric lichenoid dermatosis of Gougerot and Blum

This condition is extremely similar to Majocchi's disease but presents in adults (usually men) and is not seen in children.

Lichen aureus

- Lichen aureus, as its name suggests, presents with yellowish or red papules or patches which may either itch or be asymptomatic.
- The lesions frequently occur bilaterally on the lower limbs, although can be unilateral and may affect the trunk and upper limbs [11].
- Unlike the other forms of pigmented purpuric eruptions, the lesions of lichen aureus may also occur in a dermatomal distribution, or can follow the distribution of veins or arteries.
- It is thought that it may sometimes be associated with trauma but it does not appear to be associated with the ingestion of drugs.
- Once again, although the lesions may be cleared using oral corticosteroids, the risks of treatment outweigh the benefits, as it follows a benign course.

Differential diagnosis

Despite the distinctive appearance of the lesions, several other conditions must be considered as a differential diagnosis, including:

- T-cell lymphoma.
- Self-induced haematomas.
- Scurvy.
- Benign hypergammaglobulinaemic purpura of Waldenström.
- Drug eruptions.
- Henoch-Schönlein purpura.
- Kawasaki disease.
- Thrombocytopenia.

Investigations

- There is no specific investigation to diagnose the disorder.
- Routine laboratory investigations, such as FBC, clotting studies and ESR, show no abnormality although they may be performed to exclude other disease.
- There are no established guidelines for investigation of patients with pigmented purpuric lesions. Where skin biopsies are performed, histology will reveal evidence of capillaritis of the upper dermal vessels, extravasation of the red blood cells with haemosiderin deposits in the macrophages and an essentially normal epidermis. Biopsy may be helpful to exclude an early T-cell lymphoma.

Management

Non-drug

- Avoid prolonged dependency of the legs.
- If there is venous stasis, compression may be beneficial.
- In view of the appearance and the chronic nature of the disorder, both children and their parents will require reassurance as to the benign nature of the disorder.
- Adolescents and young adults may benefit from counselling because of the unsightly nature of the lesions.
Schamberg’s disease, pigmented purpuric lichenoid dermatosis of Gougerot and Blum and lichen aureus may benefit from treatment with psoralen combined with ultraviolet A (PUVA)\cite{12, 13, 14}.

One study has reported the use of advanced fluorescence technology (a method of delivering an intense beam of fluorescent light) in Schamberg’s disease of the legs, with favourable results\cite{15}.

Difficult or persistent cases of pigmented purpuric eruption may benefit from narrowband UVB therapy\cite{16}. Successful use in children with Schamberg’s disease has been reported\cite{15}.

Drugs

- The pigmented purpuric dermatoses follow a benign but chronic course. Although treatment with oral steroids can be effective in clearing the lesions, these are not generally used, as the risks of treatment far outweigh any potential benefits.
- Antihistamines may offer relief from pruritus but at the cost of sedation.
- Topical steroid creams may bring some symptomatic relief.
- Aminaphtone has been found to be helpful in Schamberg’s disease (not licensed in the UK).
- Pentoxifylline has also been found to be beneficial (also unlicensed use)\cite{3}.
- Treatments tried for lichen aureus include topical pimecrolimus 1% cream and a combination of pentoxifylline and epoprostenol (prostacyclin)\cite{17}.

Prognosis

- All forms of pigmented purpuric dermatoses have a chronic, relapsing and remitting benign course and are not associated with any other physical abnormality.
- Spontaneous remissions may occur.
- Very rare descriptions have occurred of T-cell lymphoma occurring in patients with a diagnosis of Schamberg’s disease\cite{18}.
Further reading & references


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