



A PEEP INTO PPP/SCHAMBERG DISEASE

Swarnagowri BN

Department of Pathology, Dr.B.R.Ambedkar Medical College, Kadugondanahalli, Bengaluru, Karnataka 560045.

Corresponding Author:- **Swarnagowri B.N.**

E-mail: swarnagowri@ymail.com

Article Info

Received 15/12/2014

Revised 27/12/2014

Accepted 02/01/2015

Key words: Purpura, pigmentation disorders.

ABSTRACT

Schamberg's disease or progressive pigmented purpuric dermatitis is a chronic discoloration of the skin which usually affects the lower limbs and spreads. This disease is more common in males and may be seen in any age. Schamberg's disease is caused by fragile and inflamed superficial blood vessels which allow red blood cells to leak into the skin. The red blood cells in the skin break down to release their iron, the hemosiderin of which gives rise to brownish color. We had a case of Schamberg's purpura, which was confirmed by biopsy.

INTRODUCTION

Schamberg Disease, (also known as "Progressive pigmentary dermatosis of Schamberg" [1], "Purpura pigmentosa progressiva" (PPP), and "Schamberg's purpura"^[1]) is a chronic discoloration of the skin found in people of all ages, usually affecting the legs. It slowly spreads throughout the body, and is most common in males [2]. It is named after Jay Frank Schamberg, who described it in 1901, in a 15 year-old boy. The condition is rare. One study, in a UK hospital-based dermatology practice serving a population of 300,000, identified only ten cases of purpuric pigmented dermatosis (of which Schamberg's disease is only one type) during a 10-month period [3]. The hemosiderin released from red cell breakdown gives rise to brownish irregular patches. These may last for several days to months.

Case report

We had a case of multiple purplish/rustic macular eruptions, in a 32 yr.old male on both the lower limbs from the Dept. of Dermatology. The haematological, including the platelet count and biochemical parameters were within normal limits. The skin biopsy from the representative area was sent for histopathological examination.

The histopathology sections with H&E staining showed the epidermis and dermis. The epidermis showed mild focal acanthosis with basket weave orthokeratosis.

The dermis revealed focal papillary oedema with few red cells exuded out from capillaries [fig.1] and hemosiderin pigments [fig.2]. Sparse lymphocytic infiltration was noted focally.

DISCUSSION

Some of the causes of Schamberg's disease are absorbent blood vessel, viral infection or allergic reaction and reaction to unknown causal agent [4]. Schamberg's Disease is superficial and does not cause pain. Pigmented purpuric reactions have five disease types: [5]

- 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 preponderance in young females and manifests as annular erythematous plaques and patches.
- Eczematoid-like purpura of Doucas and Kapetanakis - occurs in men, with bilateral intensely itchy lesions on legs.
- Lichen aureus - a localized persistent form of pigmented purpuric dermatitis [6]



There is no cure for Schamberg's disease; however, the itching can be controlled by a cortisone cream, and Colchicine treatment has been successfully used to prevent recurrence of the symptoms.^[3] Aberrant cell-mediated immunity is observed with perivascular infiltrate having specific types of CD cells only. Associated with certain medications - thiamine, aspirin, chlorthalidone and paracetamol. It has also been reported with bezafibrate [7]. There are no symptoms apart from itching and patients note 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. There is clinical and histological overlap between these and they may actually represent variable presentations of the same disease process [8].

There has been a case report of four family members with Schamberg's disease suggesting a possible genetic link [6,9]. Schamberg's disease usually runs a chronic course with frequent exacerbations and remissions. The rash may be present for many years with slow extension. Lesions may occasionally disappear spontaneously. Other causes of purpura:

- Vasculitis, e.g. leukocytoclastic vasculitis.

- T-cell lymphoma (especially if presenting in young males) [1].
- Drug eruption.
- Trauma.
- Self-induced purpura.
- Mycosis fungoides [10].

Differential diagnosis includes other associated diseases [2,5], Diabetes mellitus, Rheumatoid arthritis, Systemic lupus erythematosus, Thyroid abnormalities, Hepatic disease, Porphyria, Malignancies, Primary benign hypergammaglobulinaemic purpura of Waldenström [11] Dyslipidemia

Investigations include

- Blood tests - including platelets and clotting - are usually normal.
- Autoantibody screening 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 [4].

Examination of the skin using a dermatoscope may be helpful [7].

Figure 1. Red cells exuded out from capillaries

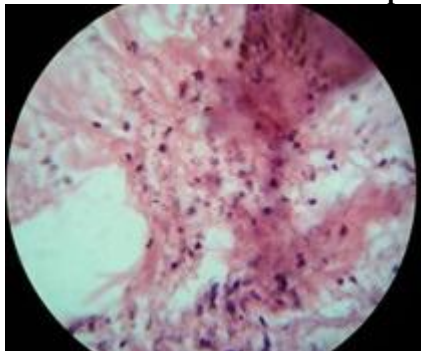
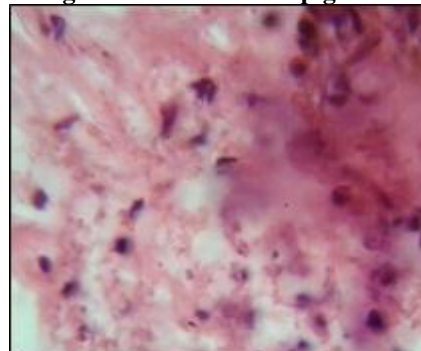


Figure 2. Hemosiderin pigments



CONCLUSION

Schamberg's disease usually runs a chronic course with exacerbations and remissions. The rash may be

present for a long period with slow extension. Lesions may occasionally disappear slowly.

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