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Short Article

Schamberg's Disease: A Comprehensive Review

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Abstract

Schamberg's disease, also known as progressive pigmented vasculitis, is a rare, chronic skin disorder characterized by petechiae and brown spots, mainly on the lower extremities. In this review article, we provide a detailed overview of Schamberg's disease. We review its causes, pathogenesis, and histopathological features. We also describe the clinical presentation, diagnostic methods, and treatment options. We emphasize the crucial role of a multidisciplinary approach in the treatment of this condition. We also emphasize the need for further research to better understand the mechanisms underlying Schamberg's disease. This could lead to the development of more targeted therapies in the future.

Keywords: Schamberg’s disease, etiology, diagnosis and treatment

Introduction

Schamberg's disease is a chronic, benign skin disorder first described by Jay Frank Schamberg in 1901 [1]. It is the most common type of cutaneous pigmentary vasculitis (PPD), a group of disorders characterized by petechiae, purpura, and hyperpigmentation [2]. Schamberg's disease primarily affects the lower extremities. It is more common in men than women, with a peak incidence in the fourth and fifth decades of life [3]. Although the exact cause remains unknown, various factors, such as venous hypertension, capillary fragility, and cellular immunity, have been implicated in its etiology [4]. This review aims to provide a comprehensive overview of Schamberg's disease. We will focus on its etiology, pathogenesis, histopathological features, clinical presentation, diagnostics, and treatment options.

Etiology and pathogenesis

The causes of Schamberg's disease are not fully understood, but scientists point to several factors that may contribute to its development. One of them is venous hypertension - increased pressure in the veins can damage the capillaries and lead to the extravasation of red blood cells, which results in the formation of petechiae and purpura [5]. The fragility of the capillaries, associated with abnormalities in the structure or functioning of their walls, may also predispose to the development of Schamberg's disease [6]. The involvement of cellular immunity in the pathogenesis of Schamberg's disease is also suspected. This is evidenced by the presence of activated T lymphocytes in the dermis, which suggests that the immune response involving these cells may play a role in the development of the disease [7]. It has been observed that some drugs, such as paracetamol, aspirin or thiamine, can trigger or exacerbate the symptoms of Schamberg's disease [8]. Interestingly, viral infections, such as hepatitis B and C, have also been associated with the development of this disease [9].

The mechanism of Schamberg's disease involves the extravasation of red blood cells from superficial capillaries of the skin, leading to the deposition of hemosiderin and the development of hyperpigmentation [10]. The exact mechanism of this process is not yet fully understood, but it is thought to involve a combination of vascular damage, inflammation, and impaired immune response. Capillary damage may be caused by factors such as venous hypertension or vascular fragility, leading to the leakage of red blood cells into the dermis. Inflammation, involving activated T lymphocytes and other immune cells, may contribute to the persistence and progression of the lesions. The extravasated red blood cells lyse, releasing hemosiderin. This is then taken up by macrophages (siderophages) and deposited in the dermis, causing the characteristic hyperpigmentation seen in Schamberg's disease.

Histopathological Features

Histopathological examination of skin biopsies from patients with Schamberg's disease reveals several characteristic features. One of the most prominent is a perivascular lymphocytic infiltrate, with a predominance of T lymphocytes around superficial blood vessels of the dermis [11]. This infiltrate suggests that an immune-mediated process is involved in the development of the lesions. Another key feature is the presence of extravasated erythrocytes in the dermis, indicating damage and leakage from capillaries [12].

Extravasation of red blood cells is a hallmark of Schamberg's disease and is responsible for the clinically observed petechiae and purpura. Hemosiderin deposition is also a characteristic finding in Schamberg's disease. This iron storage complex is located in the dermis and contributes to the clinically visible hyperpigmentation [13]. The presence of hemosiderin results from the breakdown of extravasated erythrocytes and subsequent uptake by macrophages. The dermis of patients with Schamberg's disease often contains siderophages, which are macrophages containing hemosiderin [14].

These cells play a key role in the removal of extravasated erythrocytes and the deposition of hemosiderin in the dermis. It is worth noting that Schamberg's disease does not show true vasculitis, unlike other purpuric conditions [15]. The absence of vasculitis helps distinguish Schamberg's disease from conditions such as leukocytoclastic vasculitis or Henoch-Schönlein purpura, which may present with a similar clinical picture.

Clinical Presentation and Diagnosis

Schamberg's disease typically presents with asymptomatic, irregular, reddish-brown macules and patches on the lower extremities, particularly the ankles and calves [16]. The lesions are usually bilateral and symmetrical, although unilateral presentation has been reported in rare cases. The macules and patches may vary in size, from a few millimeters to several centimeters in diameter. Over time, the lesions may coalesce to form larger patches and plaques, giving the skin a mottled appearance resembling cayenne pepper [17].

A characteristic feature of Schamberg's disease is the presence of petechiae within or around the hyperpigmented areas. These petechiae, which are small, nonfading red or purple spots that do not fade with pressure, represent extravasated red blood cells in the dermis. They may be more visible in the early stages of the disease and tend to fade as the discoloration becomes more pronounced. The onset of Schamberg's disease is usually insidious, with lesions developing gradually over weeks or months. In most cases, the lesions tend to persist and progress slowly over time, although spontaneous resolution has been reported in some cases. The disease is usually asymptomatic, but some patients may experience mild itching or a burning sensation in the affected areas. The diagnosis of Schamberg's disease is based primarily on the clinical presentation, as the appearance and distribution of the lesions are often characteristic [18].

However, histopathological examination of skin biopsies can be valuable in confirming the diagnosis and excluding other conditions. The histopathological features of Schamberg's disease, as discussed earlier, include perivascular lymphocytic infiltrate, extravasated erythrocytes, hemosiderin deposits, and the presence of siderophages. When considering the differential diagnosis of Schamberg's disease, other pigmented vasculitides of the skin (PPD) should be considered. These include Majocchi disease (telangiectasia annular purpura), which presents with annular or arcuate lesions and prominent telangiectasias, and Gougerot-Blum disease (lichenoid pigmentary vasculitis), characterized by lichenoid papules and plaques [19].

Additionally, several other conditions may mimic the appearance of Schamberg's disease and should be ruled out. Vasculitis, particularly leukocytoclastic vasculitis, can present with purpuric lesions on the lower extremities but is usually associated with systemic symptoms and a more acute onset [20]. Stasis dermatitis, which occurs due to venous insufficiency, may also present with hyperpigmentation and hemosiderin deposition on the lower legs but is often accompanied by edema, eczematous changes, and pruritus. Drug-induced eruptions, especially those caused by medications such as nonsteroidal anti-inflammatory drugs (NSAIDs) or

antibiotics, can sometimes resemble Schamberg's disease but tend to have a more rapid onset and may resolve upon discontinuation of the offending drug.

Treatment Options

Because Schamberg's disease is a mild and asymptomatic condition, treatment is often not necessary. However, for patients who wish to improve the appearance of their skin or are experiencing discomfort, several treatment options are available. Topical corticosteroids, particularly low- or medium-strength ones, can help reduce inflammation and improve the appearance of lesions [21]. These medications work by suppressing the immune response and reducing capillary permeability, thereby minimizing red blood cell extravasation and subsequent discoloration. Topical vitamin C has also been suggested as a potential treatment for Schamberg's disease. The antioxidant and anti-inflammatory properties of vitamin C may help reduce the appearance of lesions by scavenging free radicals and reducing oxidative stress [22].

Additionally, vitamin C plays a role in collagen synthesis, which may help strengthen capillary walls and prevent further damage. PUVA therapy (psoralen plus ultraviolet A radiation) has been shown to be effective in some cases of Schamberg's disease, although its mechanism of action is not fully understood [23]. PUVA therapy involves the administration of psoralen, a photosensitizing agent, followed by exposure to ultraviolet A radiation. This combination has been shown to have immunomodulatory and anti-inflammatory effects, which may contribute to the improvement of the lesions. Laser therapy, including pulsed dye laser and Q-switched laser, has also been used to treat the discoloration associated with Schamberg's disease [24].

These laser methods target hemosiderin deposits in the dermis, breaking them down and allowing the body's immune system to remove them. Laser therapy may be particularly useful for patients with persistent or refractory lesions. In patients with venous insufficiency, compression stockings may help reduce venous hypertension and prevent disease progression [25]. By applying graduated pressure to the lower extremities, compression stockings promote venous return and reduce blood stasis in the legs, which may help minimize capillary damage and the development of new lesions.

Conclusion

Schamberg's disease is a chronic, benign cutaneous disorder characterized by petechiae, purpura, and hyperpigmentation, primarily affecting the lower extremities. Although the exact etiology remains unknown, various factors, including venous hypertension, capillary fragility, and cell-mediated immunity, have been implicated in its pathogenesis. Histopathological examination reveals perivascular lymphocytic infiltrate, extravasated erythrocytes, hemosiderin deposition, and siderophages. The diagnosis is based on clinical findings and supported by histopathology. Treatment options include topical corticosteroids, vitamin C, PUVA therapy, laser therapy, and compression stockings, although the efficacy of these interventions varies. Further research is needed to better understand the underlying mechanisms of Schamberg's disease and develop targeted therapies to improve patient outcomes.

Disclosure:

Author's contribution:

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