

Alexander KC Leung^{1*}, Amy AM Leung² and Benjamin Barankin³

¹Clinical Professor of Pediatrics, The University of Calgary, The Alberta Children's Hospital, Calgary, Alberta, Canada

²Medical Student, Faculty of Medicine and Dentistry, University of Alberta Edmonton, Alberta, Canada

³Dermatologist & Medical Director, Toronto Dermatology Centre, Toronto, Ontario, Canada

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***Corresponding author:** Dr Alexander KC Leung, Clinical Professor of Pediatrics, The University of Calgary, Pediatric Consultant, The Alberta Children's Hospital, #200, 233-16th Avenue NW, Calgary, Alberta, Canada, T2M 0H5, Tel: (403) 230 3300; Fax: (403) 230-3322; E-mail: aleung@ucalgary.ca

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Introduction

Schamberg disease, also known as Schamberg purpura or progressive pigmentary purpura, is characterized by orange-brown or reddish-brown macules/patches with overlaying purpuric spots simulating "cayenne pepper" [1,2]. The condition was first described in 1901 by Jay Frank Schamberg who reported a 15-year-old boy with a 5-year history of "diffuse, reddish-brown, non-elevated, irregular oval patches with borders consisting of pin-head size, reddish-brown, scarcely elevated puncta or cayenne-pepper spots" [3]. The condition now bears his name. To our knowledge, Schamberg disease has not been reported in Chinese patients. The purpose of this communication is to alert the medical profession to the existence of this condition in Chinese patients so that the diagnosis would not be overlooked.

Case Report

A 54-year-old Chinese woman presented with a 2-month history of an asymptomatic, erythematous eruption on her legs. The lesions began on her ankles and spread proximally to the legs and knees. The patient noted steady accumulation of new lesions and their coalescence into wider areas of discoloration. There was no history of recent fever, upper respiratory tract infection, and trauma or drug intake. The patient was diagnosed with an adenocarcinoma in the ascending colon 3 years ago and had a right hemicolectomy performed. The post-operative course was uneventful. She had no known allergies. Family history was negative of similar skin findings or any bleeding disorder.

Physical examination revealed purpuric macules and patches of varying sizes on the ankles, legs, and knees (Figure 1). The lesions were nonblanchable and nonpalpable. Close inspection of the lesions revealed pinpoint petechiae resembling grains of cayenne pepper superimposed on the reddish-brown macules/patches. The rest of the physical examination was unremarkable.

Case Report

Schamberg Disease in a 54-Year-Old Chinese Woman

Abstract

Schamberg disease, also known as Schamberg purpura or progressive pigmentary purpura, is characterized by orange-brown or reddish-brown macules/patches with overlaying purpuric spots simulating "cayenne pepper". We report a 54-year-old Chinese woman with Schamberg disease presenting with purpuric macules and patches of varying sizes on the ankles, legs, and knees. The lesions were nonblanchable and nonpalpable and asymptomatic. Close inspection of the lesions revealed pinpoint petechiae resembling grains of cayenne pepper superimposed on the reddish-brown macules/patches. To our knowledge, Schamberg disease has not been reported in the scientific literature in Chinese patients.

Laboratory investigations revealed a hemoglobin of 149 g/L, a white blood cell count of $6.2 \times 10^9/L$ with a normal differential count, and a platelet count $219 \times 10^9/L$. The prothrombin time international normalized ratio (INR) was 0.9. A clinical diagnosis of Schamberg disease was made based on the typical history and physical findings. No skin biopsy was deemed necessary. The patient was treated with topical desoximetasone 0.25% cream once a day, and advised to take Vitamin C 500 mg po bid. Follow-up at two weeks revealed a significant decrease in the number and extent of the lesions. Follow-up at one month showed total resolution of the lesions, by which time desoximetasone 0.25% cream was discontinued.

Discussion

Typically, patients with Schamberg disease present with symmetrical reddish-brown macules/patches with superimposed speckled cayenne pepper-like petechiae [1,2]. It is believed that the condition results from a T cell-mediated immune reaction with the interaction of intercellular cell adhesion molecule 1 [1,4]. This results in capillary damage with extravasation of erythrocytes and consequent deposition of hemosiderin in the dermis, giving rise to



Figure 1: Purpuric rash extending from ankles to knees coalescing into reddish-brown macules and patches.



the distinct reddish-brown discoloration. The condition is usually asymptomatic; rarely, patients may complain of mild pruritus [2]. The most common site of predilection is the legs, followed by the thighs and buttocks [2,5]. Onset occurs on average in the fifth decade of life [5]. There is a male predominance [6].

Schamberg disease is generally idiopathic. Some cases may be attributed to medications, venous hypertension, gravitational dependency, capillary fragility, contact allergy to wool or clothing dyes, trauma, or exercise [1,6]. Schamberg disease may also be familial [7].

The differential diagnosis include, among others, purpura annularis telangiectodes of Majocchi, pigmented purpuric lichenoid dermatitis of Gougerot and Blum, lichen aureus, eczematid-like purpura of Doucas and Kapetanakis, Henoch Schönlein purpura, stasis dermatitis, contact dermatitis, drug sensitivity purpura, traumatic purpura, thrombocytopenic purpura, collagen vascular disease, paraproteinemia, hyperglobulinemic purpura, and cryoglobulinemia.

Currently, there are insufficient data for conclusions on racial or ethnic predilections. Suffice to say, Schamberg disease has been reported in many races, such as Caucasian, Korean, and Japanese [1,2,5,8]. We herewith report a Chinese patient with Schamberg

disease that, to our knowledge, has not been previously reported in an English-language PubMed literature search. One should bear in mind that this disease may also affect Chinese patients and that the disease should not be overlooked in that racial group.

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