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Generalized Schamberg Disease in a Child

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Dear Editor:

Schamberg disease is a type of pigmented purpuric dermatosis that is characterized by red-brown purpuric macules that are known as 'cayenne pepper' spots. It usually develops insidiously on one or both lower legs and onset occurs on average in the fifth decade of life. Herein, we report a generalized form of Schamberg disease that occurred in a child. A 12-year-old boy presented with a 2-month history of an asymptomatic widespread rash. There was no specific history such as recent drug intake. He had been treated previously with topical steroids, but there had been no significant change in his condition. Physical examination revealed purpuric and telangiectatic macules and patches scattered all over his trunk and extremities (Fig. 1). Laboratory findings, including clotting profile, were within normal limits. Histological examination of a skin biopsy revealed extravasated red blood cells and enlarged vessels with perivascular lymphocytic infiltrates in the upper dermis (Fig. 2). Based on these clinical and histopathological findings, the patient was diagnosed with Schamberg disease. After treatment with oral prednisolone (10 mg) and pentoxifylline (400 mg) daily for 4 weeks, the skin lesions improved, leaving hyperpigmentation.

The onset age in the present case was interesting as most cases occur in adulthood. However, some authors have suggested that Schamberg disease is relatively common in children, sometimes occurring before puberty[1]. Torrelo et al[1]. reviewed 13 cases of Schamberg disease that occurred before 10 years of age and found that it predominantly occurred in girls, consistently involved the lower limbs, and was a chronic persistent disease. A case of a 1-year-old infant with Schamberg disease has also been reported[2].

An interesting feature of the present case was that the eruptions were generalized. The lesions in Schamberg disease usually develop on one or both lower legs, and they sometimes spread and involve the trunk or upper extremities. Three cases of generalized Schamberg disease have been reported, all of which occurred in children[3,4]. In these three cases, the generalized lesions responded well to psoralen plus ultraviolet A or narrow band ultraviolet B therapy and treatment resulted in clearing of the lesions after either the tenth, twelfth, or twentieth treatment session. The present case improved after treatment with an oral corticosteroid and pentoxifylline. These results indicate the benign nature of generalized Schamberg disease in childhood. Corticosteroids are well known anti-inflammatory agents that have shown positive out-

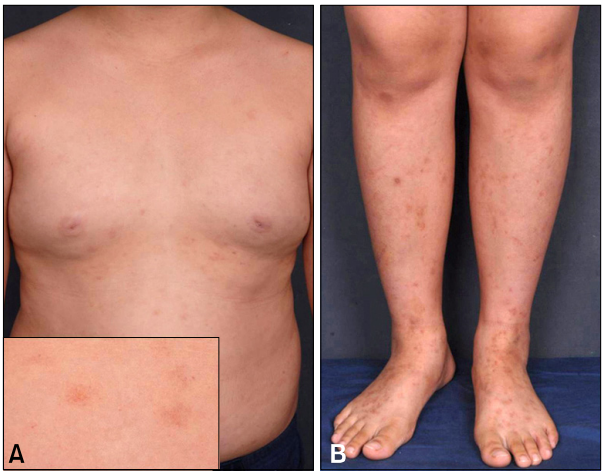

Fig. 1. Physical examination revealed purpuric and telangiectatic macules and patches scattered all over his trunk (A; inset: a close-up new) and extremities (B).

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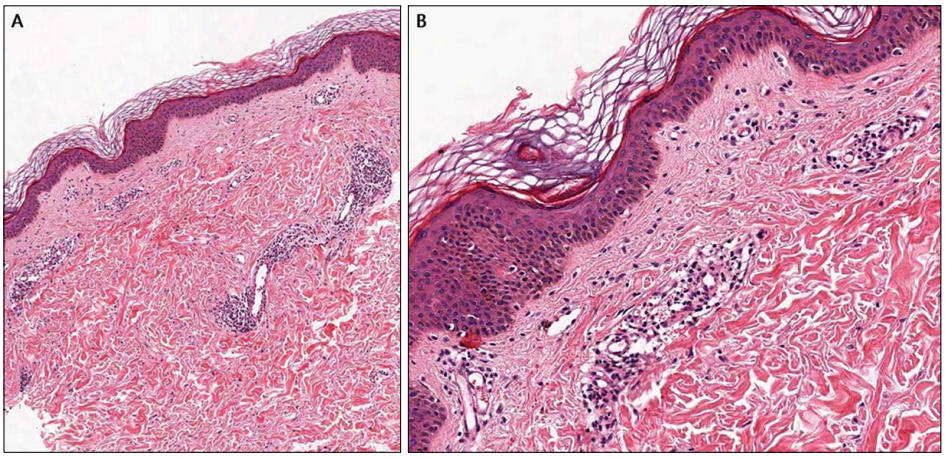

Fig. 2. Skin biopsy revealed extravasated red blood cells and enlarged vessels with perivascular lymphocytic infiltrates in the upper dermis (A: H&E, ×40; B: H&E, ×200).

comes in several cases of Schamberg disease. The administration of pentoxifylline can have several effects, including decreased fibrinogen concentration, and reduced platelet adhesiveness. In addition, it inhibits T-cell adherence to endothelial cells and keratinocytes.

Childhood Schamberg disease needs to be distinguished from other dermatoses such as drug hypersensitivity reactions, Henoch-Schonlein purpura, and thrombocytopenia associated with viral infection. Dermoscopic examination can be used for the diagnosis of pigmented purpuric dermatosis[5], as findings will show purpuric globules over a purpuric and orange-brown background.

In conclusion, we reported a case of generalized Schamberg disease occurring in a child. Accurate diagnosis of pigmented purpuric eruptions in childhood is needed considering the benign nature of this condition.

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