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Original Article

Schamberg's Disease: The clinical and histopathological findings in favour of Stasis Vasculitis

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Abstract

Objective Schamberg's disease is the most common type of pigmented purpuric dermatosis, characterized by a benign course with reddish-brown patches or macules with overlying purpuric spots resembling a cayenne pepper. It is not uncommon condition, but the available information regarding its epidemiological, clinical, and histopathological features are very limited. The objective of the study is to evaluate the clinical and histopathological features of Schamberg's disease trying to have more insight into its etiopathogenesis.

Methods A descriptive cross sectional study was conducted in the Center of Dermatology and Venereology, Medical City, Baghdad, Iraq, during the period from October 2018 to March 2020. Fifty patients with clinical features of Schamberg's disease were included. The demographic features were recorded. Full history and dermatological examination were carried out.

Results 50 patients were included with mean age of 47.22±11.8 years. 76% were males and 24% were females. The lesions were symmetrical and consisted of macules (44%), petechiae and macules (32%). Histopathological examination showed normal stratum corneum in 11 (100%), acanthosis in 18% , epidermal atrophy in 72%, spongiosis in 45%, rete ridges effacement in 45%, basal layer vacuolization in 27%. Many pathological changes in superficial dermis were observed like RBC extravasation, mostly around dilated and congested blood vessels was noted in 81%, variable perivascular lymphocytic infiltrates were seen in all 11 cases; 36% mild infiltrates, 36% moderate infiltrate and 27% extensive with fibriniod alterations in some of the sections. These changes were suggestive of lymphocytic vasculitis. Perls stained sections revealed dermal iron deposition (Hemosiderin) with in siderophages mainly in the upper dermis in 82% that was mild in 11%, moderate in 33% and extensive in 55% of the patients.

Conclusion Schamberg's disease is a chronic, benign, asymptomatic disease affecting lower limbs in a symmetrical fashion with characteristic clinical and histopathological features that suggest lymphocytic vasculitis rather than capillaritis of dermis, leading into hemosiderin deposition as an outcome. These features with poor circulation do suggest that disease is a form of vasculitis that could be named stasis vasculitis as the main etiopathological finding.

Key words
Pigmented purpuric dermatosis, Schamberg disease, Perl stain, hemosiderin.

Introduction

Schamberg's disease is a chronic disorder that most frequently affects the lower limbs and spreads slowly, but may occur anywhere on the body, including the hands, arms, torso and even the neck. The lesions may vary and break out in large numbers.[1] They consist of irregular patches of orange or brown pigmentation with characteristic "cayenne pepper" spots appearing

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within yellowish background. There are usually no symptoms, though there may be some slight itching, however, there is no pain. The eruption may persist for a long period.[2] In India, it is found that the prevalence to be 0.18% of the total patient admitted to the dermatology and venereology clinic over a period of 1.5 years. Overall, it is not common in United States and in United Kingdom in outpatient dermatology clinic that cover a community of 300 thousand population only ten patients were diagnosed with capillaritis and half of them diagnosed with lichen aureus.[2]

This disease is mostly affected males and may occur at any age but most commonly in the 3rd or 4th decade. This condition is observed universally and has nothing to do with race or ethnic background.[1] Schamberg disease can be diagnosed clinically by trained dermatologist, however, primary care physician or untrained eye may misdiagnose it as bleeding disorder, thus it may require reviewing medical history and investigations in addition to the thorough clinical examination.[2] If there is any suspicion in the diagnosis of Schamberg disease, complete blood count (CBC) is ordered to ensure that the skin lesions are not caused by any other conditions and to rule out bleeding disorders that cause purpura. The CBC results are usually normal.[3] Although most cases of Pigmented Purporic Dermatosis (PPD) can readily be diagnosed clinically. A skin biopsy sometimes is needed to ensure the diagnosis PPD showed numerous findings including perivascular lymphocytic infiltration, red blood cell extravasation, hemosiderin deposition, endothelial cell swelling, spongiosis, lymphocyte exocytosis, and lichenoid lymphocytic infiltration.[4] No comprehensive study has been performed in Iraqi population; thus, this study aims to describe the clinical and histopathological characteristics of the disease in order to reach new insight about the pathogenesis of the disease.[5]

Methods

This is a descriptive, cross sectional study that was carried out in the Center of Dermatology at Baghdad Teaching Hospital, during the period from October 2018 to March 2020. Fifty patients with Schamberg disease were included in this study through a convenient sample and excluded any patient with vasculitis, and patients who have known hematological disorder. The diagnosis was established on history and clinical features with or without histopathological examination. Information is collected from each patient by a questionnaire that included socio-demographic characteristics, past medical history, clinical characteristics of disease, associated systemic diseases and hematological investigations were advised in every case. While Skin biopsy was performed for eleven patients only and the variables, including cell count, a scoring system of less than 3 (scant), 3–5 (mild), 5–10 (moderate), and more than 10 (severe)cells per high power field (HPF) was used.

The categorical variables were presented as frequencies and percentages. The continuous variables were presented as mean and standard deviation.

An approval was taken from the scientific committee of the Scientific Council of Dermatology and Venereology- Arab Board for Health Specializations. A verbal consent was taken from each patient before the examination.

Results

The total sample was fifty patients. 76% were males and 24% were females with a mean age of 47.22±11.8 years. Twenty percent of patients with Schamberg's disease were housewives, followed by retirees (16%) (**Table 1**).

Table 1 Demographic features of patients with Schamberg's disease.

Demographic Criteria	*No*	*%*
Age		
Mean± SD	(47.22±11) years	
Range	23-85 years	
Age (years)		
10-20	0	0
21-30	3	6%
31-40	10	20%
41-50	19	38%
>50	18	36%
Sex		
Male	38	76%
Female	12	24%
Male: Female	3.2:1	
Occupation		
Housewife	10	20%
Retirees	8	16%
Freelancer	7	14%
Serviceman	6	12%
Teacher	6	12%
Engineer	3	6%
Butcher	2	4%
Health worker	2	4%
Officer	1	2%
Pharmacists	1	2%
Barber	1	2%
Professor	1	2%
Military	1	2%
Driver	1	2%
Past surgical history		
Positive	2	4%
Past medical history		
Positive	21	42%

The disease is distributed bilaterally in all cases (100%). The rash is mostly located in the legs and feet, 60% and 36% respectively (**Figure 1**), three exceptional cases were noted where the lesions extended to the thighs, soles (**Figure 2**) and dorsa of the hands respectively. As for the most common type of primary skin lesions were macules (44%) while the lesion color ranged from reddish brown to brown (36% and 28% respectively) with a mean duration of 1.86±2.6 years (**Table 2**).

Biopsy was performed for 11 cases and both H&E and Perls stains were used that revealed the following findings: normal stratum corneum in all 11 patients (100%), acanthosis (18%), epidermal atrophy (72%), spongiosis (45%) and basal layer vacuolization (27%). Features of vasculitis were seen in 3 patients (27%) where dilatation and rupture of vessels wall with free RBC extravasation surrounded by cuffing of lymphocytes (**Figure 3**). RBC extravasation was found mostly around dilated and congested blood vessels in 9 patients (81%) (**Figure 4**). Variable perivascular lymphocytic infiltrates were seen in all 11 cases, mild infiltrates in 4 patients (36%), moderate infiltrates in 4 patients (36%) and extensive infiltrates in 3 patients

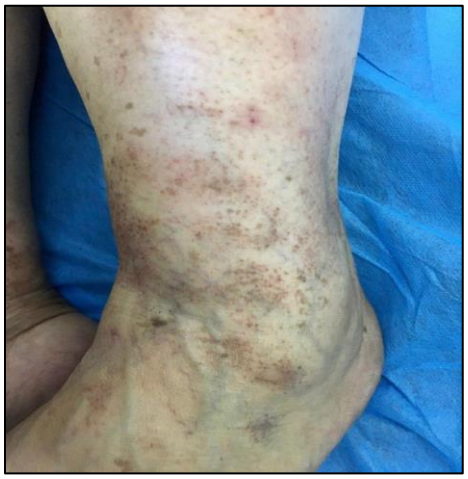

Figure 1 55 year old female with Schamberg's disease showing reddish-brown macules with overlying petechiae on the dorsum of the foot & leg.

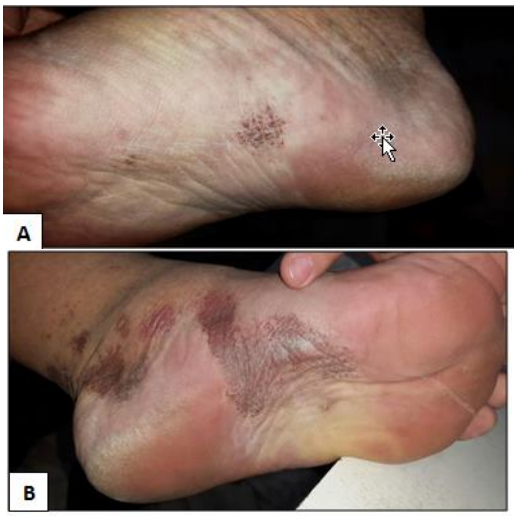

Figure 2 42 year old male with Schamberg's disease; affecting the sole and presented with petechiae overlying reddish-brown plaque.

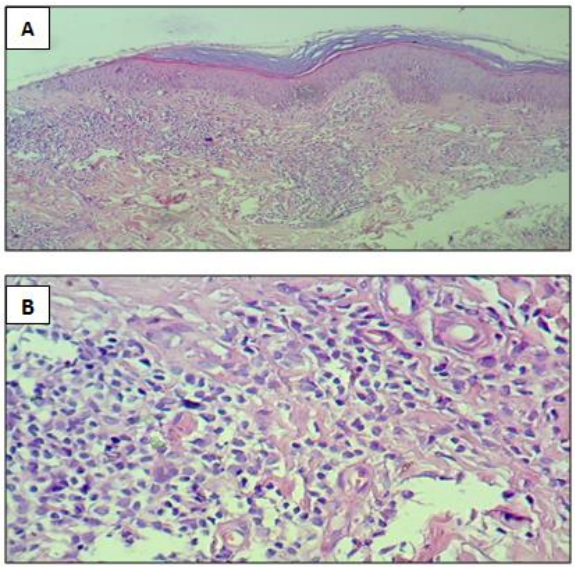

Figure 3 Hematoxylin and Eosin stained sections showing marked perivascular infiltration (A) and dilatation & congestion of blood vessels surrounded by cuffing of lymphocytes (B).(A-x100-B-x400).

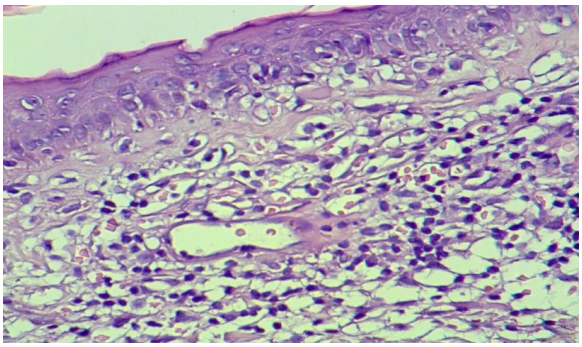

Figure 4 Hematoxylin and Eosin stained section showing perivascular lymphocytic infiltration & extravasated RBC.(x400).

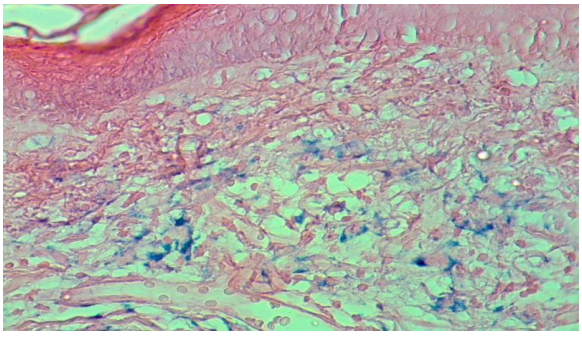

Figure 5 Perl Prussian stained section showing blue hemosiderin deposition in the dermis (x400).

(27%) with fibrinoid changes in some sections. These pathological findings were consistent with lymphocytic vasculitis. (**Table 3**). Perls stained

Table 2 Clinical criteria of patients with Schamberg's disease.

Clinical Criteria	*No*	*%*
Distribution of lesion		
Leg	30	60%
Feet with one case with sole involvement	18	36%
Thigh	1	2%
Dorsum of hand	1	2%
Aggravating factors		
Varicose vein	4	8%
Drugs		
Aspirin	3	6%
Paracetamol	2	4%
Beta-blocker	2	4%
Insulin	2	4%
Diazepam	1	2%
ACE inhibitors	1	2%
Glucophage	1	2%
Anti-hypertensive	1	2%
History of Prolonged standing	4	8%
Primary skin lesion		
Macules	22	44%
Macules & petechiae	16	32%
Patches	10	20%
Plaques & macules	1	2%
Plaques & petechiae	1	2%
Color		
Reddish -brown	18	36%
Brown	14	28%
Yellowish- Brown	12	24%
Purplish –brown	6	12%
Duration		
<1yr	32	64%
≥1yr	18	36%
Symptoms		
Asymptomatic	36	72%
Itching	14	28%
Associated disorders		
Diabetes mellitus	10	20%
Hypertension	9	18%
Cerebro- Vascular Accident	1	2%
Chronic Renal Failure	1	2%

sections revealed increased dermal iron deposition (Hemosiderin) within siderophages mainly in the upper dermis in 9 patients (82%) (**Figure 5**). Cases were classified by counting the number of siderophages in 10 HPFs then dividing it by 10 to get number of siderophages per single HPF. Among 9 patients, 1 patient (11%) was classified as mild, 3 patients (33%) as moderate and 5 patients (55%) as extensive (**Table 3**).

Table 3 Histopathological features of patients with Schambergs disease.

Feature	*Patients No (%)*
Normal St. corneum	11 (100%)
Spongiosis	5 (45%)
Epidermal atrophy	8 (72%)
Acanthosis	2 (18%)
Rete ridges effacement	5 (45%)
Basal layer vacuolization	3 (27%)
RBC extravasation	9 (81%)
Lymphocytic vasculitis	3 (27%)
Perivascular lymphocytic infiltrates of dermis Mild = 4 (36%), Moderate = 4 (36%), Severe= 3 (27%)	11 (100%);
Bullous lesion	1 (9%)
Hemosiderin deposition (Siderophages/ HPF)	
Negative	2 (18%)
Positive	9 (82%)
Mild = 1 (11%)	
Moderate = 3 (33%)	
Severe = 5 (55%)	
Total	11 (100%)

Discussion

Schamberg's disease is included into a disease group known as "purpura pigmentosa progressiva". These diseases are benign with similar clinical and histopathological characteristics. In the present study, Schamberg's disease is most commonly occurs at 4th and 5th decades of life with a mean age of 47.22±11 years. However, it may occur in a younger age.[6] It is mostly seen in males (76%) than females (24%). These findings are in concordance with a study by Gupta *et al*. and with an Indian study by Sharma *et al*.[7]

The most accepted mechanism involves factors that may increase intravascular pressure which leads to constant aneurysmal dilation of the capillaries, making their walls fragile and prone to rupture in the papillary dermis. Some of these predisposing factors are occupations with long periods of standing, our result found the housewives (20%) carry the commonest incidence, followed by retirees (16%), This finding is also consistent with Gupta *et al*. study which reveals that housewives constituted the largest group comprising (45%) of the patients, followed by businessmen (13.30%).[7] Drugs are provocative factors which have been reported to cause Schamberg's disease include glipizide,[8] acetaminophen,[9] aspirin, medroxyprogesterone acetate,[10] pseudoephedrine[11] and vitamin B1 (thiamine).[12] In this study, the most common reported drugs were aspirin, acetaminophen, insulin, B-blockers, as well as diazepam, ACE inhibitors, metformin, and anti-hypertensives. Although varicosity among females and males is a common medical problem, but it is reported by only (8%) of patients enrolled in this current study. However, in another study by Gönül *et al*, varicosity was reported in 20.8%. Hence, this may be attributed to the incidental nature of varicosity rather than a causative factor. Another predisposing factor is the presence of systemic co-morbid diseases as of all patients, 38% had a systemic disease. Diabetes mellitus being the most common one (20%), followed by hypertension (18%). This finding is variable in different studies and a close result to our finding was found by Gönül *et al* study who detected co-morbidities in the majority of their patients; nearly 71%, and diabetes is the most common (23.5%).[13]

The present work shows that most cases were asymptomatic (72%) apart from (28%) who have a mild itching. This is similar to a study by Sharma and Gupta *et al*.[14] who observed itching in 30% of cases and also in sardana *et al*. study, where most of the Schamberg's disease patients were asymptomatic and distributed bilaterally.[4] The rash in the present study is mainly located in the legs (60%) in a symmetrical fashion, and then in dorsa of feet (36%) while in one exceptional case, the sole was involved and in the other patient, the hands dorsa were inflicted. These findings agreed with the studies done by

Sharma and Gupta,[14] and Kim *et al*.[15] who also observed upper extremity involvement in addition to the lower extremity in 13.2% of cases. The morphology of the lesions in this study were mostly macules (44%), while macules with petechiae were present in 32%. Gupta *et al*.[7] mentioned a wide variation with most of the patients having more than one type of lesion and associated presence of macules and purpura was the most evident morphology (33.3%). Taketuchi *et al*. also described variable presentations of the rash with petechial, macular, brown, red or yellow patchy pigmentation seen. The rare unilateral presentation was detected in only two cases.[16] In the present study, the lymphocytic infiltrate is well markedly seen and documented in all cases (100%), 36% are mild cases, 36% moderate and 27% extensive. The feature of vasculitis is seen in the present study where dilatation and rupture of vessels wall surrounded by cuffing of lymphocytes and free RBC, these features support of actual lymphocytic vasculitis rather than capillaritis as has been suggested by previous studies.[17] Hemosiderin levels were seen within siderophages especially in the superficial part of the dermis and was classified as mild cases (11%), moderate cases (33%), and severe in (55%) cases. This agrees with a study by Sardana *et al*. which showed that RBCs extravasation with significant deposition of hemosiderin detected by iron stain in macrophages especially at the superficial part of dermis.[4] The present work demonstrated that the disease process is a chronic one as no patient mentioned a remission during his clinical course of the disease although spontaneous improvement after few months is said to be usual. This appears to be in concordance with other studies. However, recurrences have been found to occur. The patients were given treatment according to their symptoms and cosmetic issue. In spite of the straight way to diagnosis, the disease identity remains a mystery and a therapeutic challenging issue.[4] Upon the results of the present study that is supported by previous literatures and in relation with clinical and histopathological findings, we can conclude that the etiopathogenesis of Schamberg's disease could elucidated in form of vasculitis of legs in a patient with poor venous circulation. This type of vasculitis could be named as stasis vasculitis.

References

1. Cho J. H., Lee, J. D., Kang, H., *et al*. The clinical manifestations and etiologic factors of patients with pigmented purpuric dermatoses. Korean J Dermatol. 2005;**43(1)**: 45-52.
2. Kim D., Seo S., Ahn H., *et al*; Characteristics and Clinical Manifestations of Pigmented Purpuric Dermatosis. *Ann Dermatol*. 2015;**27(4)**:404-10.
3. Murota H, Katayama I. Lichen aureus responding to topical tacrolimus treatment. *J Dermatol*. 2011;**38(8)**:823-5.
4. Sardana K., Sarkar R., Sehgal V. Pigmented purpuric dermatoses: An overview. *Int J Dermatol*. 2004;**43(7)**:482–8.
5. Jacob M., Wright R., Mazur L., *et al*. Pigmented purpuric dermatosis. *J Paediatr Neonatal Dis*. 2018;**3(1)**:107.
6. Tristani P, Meadows K, Vanderhooft S. Pigmented purpuric eruptions of childhood: A series of cases and review of literature. *Pediatr Dermatol*. 2001;**18**:299-304.
7. Gupta G., Mushtaq S., Dogra D *et al*. A cross-sectional study of clinico-etiological profile and associated comorbidities in Indian patients of pigmented purpuric dermatoses. *Indian J Dermatol*. 2020;**65(3)**:187.
8. Adams B., Gadenne A. Glipizide-induced pigmented purpuric dermatosis. J Am Acad Dermatol. 1999;**41(5)**:827-9.
9. Kwon S., Lee C. Figurate purpuric eruptions on the trunk: acetaminophen-induced rashes. *J Dermatol*. 1998;**25(11)**:756-8.
10. Tsao H., Lerner L. Pigmented purpuric eruption associated with injection medroxyprogesterone acetate. *J Am Acad Dermatol*. 2000;**43(2)**:308-10.
11. Díaz Jara, Tornero P, Barrio MD, *et al*. Pigmented purpuric dermatosis due to pseudoephedrine. *Contact Derm*. 2002;**46(5)**:300-1.

12. Yung A, Victoria G. Pigmented purpuric dermatosis (capillaritis) induced by bezafibrate. *J Am Acad Dermatol*. 2005;**53(1)**:168-9.
13. Gönül M, Çakmak S, Özcan N, *et al*. Clinical and laboratory findings of pigmented purpuric dermatoses. *Ann Dermatol*. 2014;**26(5)**:610-4.
14. Sharma L, Gupta S. Clinicoepidemiological study of pigmented purpuric dermatoses. *Indian Dermatol Online J*. 2012;**3(1)**:17.
15. Kim D, Seo H, Ahn H, *et al*. Characteristics and clinical manifestations of pigmented purpuric dermatosis. Ann Dermatol. 2015;**27(4)**:404-10.
16. Takeuchi Y., Chinen T., Ichikawa Y., *et al*. Two cases of unilateral pigmented purpuric dermatosis. *J Dermatol*. 2001;**28(9)**:493-8.
17. Schroeder D T. Pigmented Purpuric dermatoses. In: Wolf K, Goldsmith LA, Katz SI, Gilchrest BA, Paller AS, Leffel DA, editors. Fitzpatrick's Dermatology in General Medicine. 7thed. McGraw-Hill; 2008: 1633-1636.