Review Article

# Cutaneous vasculitis

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**Abstract** Vasculitides constitute a heterogeneous group of inflammatory disorders involving walls of blood vessels of integument and viscera. Depending on the size of blood vessels affected and organs involved, different subsets can vary in clinical presentation, histopathology, laboratory markers, treatment and prognosis. The present review focuses on recapitulating the vasculitides involving skin in a tabulated form.

***Key words***
Cutaneous vasculitis, classification, clinical presentations, treatment, diagnosis.

## Introduction

Vasculitis is inflammation of blood vessels with a wide range of clinical and histological presentations. Its severity may range from self-limited to life threatening. The subject has been discussed in varying details as regards its classification, etiology, clinical presentations, histopathology, treatment and prognosis. Size of the vessels affected which is considered the most important parameter as regards various classifications of vasculitis, forms the basis of this tabulated version of vasculitis. Some of the vasculitides which at different periods of time have been included and excluded from true vasculitis because of one reason or the other have also been discussed here.

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**Table 1** [1-44]

| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow up* |
|---|---|---|---|---|---|
| **Predominantly small vessel** | | | | | |
| Cutaneous small vessel vasculitis (CSVV) | ***Cutaneous***<br>Usually a single crop of asymptomatic purpura, papules, vesicles and urticarial lesions on dependent areas.<br>10% are chronic.<br><br>***Systemic***<br>Rare. | ***Histopathology***<br>Endothelial swelling<br>Fibrinoid necrosis of vessel walls<br>Extravasation of RBCs<br>Leucocytoclasia<br><br>***Immunofluorescence***<br>IgM and C3 perivascular deposits in superficial dermal papillary vessels. | ***For diagnosis***<br>Detailed history<br>Thorough examination<br>Skin biopsy<br><br>***For association***<br>Blood and urine examination and culture.<br>HBV, HCV & HIV serology<br>Connective tissue profile<br>Lymph node, liver and bone marrow biopsies | ***Mild cases***<br>NSAIDS<br>Antihistamines<br>Colchicine<br>Dapsone<br><br>***In recurrent or resistant cases***<br>Azathioprine<br>Methotrexate<br>Cyclosporin<br>Cyclophosphamide | 90% have single episode.<br>10% chronic (arthralgia, absence of fever, cryoglobulinemia). |

| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow up* |
|---|---|---|---|---|---|
| Cryoglobuline-mic vasculitis (CV) | ***Cutaneous (15%)***<br>*Common*<br>Purpura<br>Ecchymosis<br>erythematous papules<br>dermal nodules<br>Raynaud's phenomenon<br><br>*Uncommon*<br>Urticaria,<br>Livedo reticularis<br>Bullous lesions<br>ulcerations<br><br>***Systemic***<br>Arthralgias, arthritis and weakness<br>Peripheral neuropathy<br>Nephritis or nephritic syndrome | ***Histopathology***<br>Leukocytoclastic vasculitis<br><br>***Immunofluorescence***<br>Deposition of IgM and IgG complexes in the vessel wall. | ***For diagnosis***<br>Same as for CSVV circulating cryoglobulin levels, & serum Complement profile.<br><br>***For extent of disease***<br>Renal function tests. | ***For HCV-associated disease***.<br>Ribavirin with or without alpha interferon<br><br>***For renal and neurological diseases***.<br>Corticosteroids<br>Colchicines, IVIG, cyclosporin, Melphalan | Treatment of underlying disorder is the key to recovery. However, with most treatment higher cryoglobulin levels may persist even after resolution of symptoms. |

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| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow up* |
|---|---|---|---|---|---|
| Urticarial vasculitis. Normocomplementemic (NUV) and hypocomplementemic (HUV) | ***Cutaneous*** Urticarial lesions lasting more than 24 hours with purpura, Post inflammatory pigmentation and burning. ***Systemic*** Eye symptoms (iritis, uveitis, episcleritis) Angioedema Obstructive airway disease | ***Histopathology*** Leucocytoclastic vasculitis. HUV has more interstitial neutrophils than eosinophils as in NUV. | ***For diagnosis*** Lesions lasting more than 24 hours, pain rather than itch and presence of purpura. Complement levels(C3, C4), ANA. Urinalysis for hematuria. | Antihistamines NSAIDS Corticosteroids Colchicines Hydroxychloroquine Dapsone alone or with pentoxyphylline, Mycophenolate mofetil. | Unpredictable course. Average duration 3 years. |
| Henoch-Schonlein Purpura (HSP) | ***Cutaneous*** Symmetrical purpura ***Systemic*** Arthralgias Colicky abdominal pain Hematuria | ***Histopathology*** Leucocytoclastic vasculitis ***Immunofluorescence*** IgA, C3, and fibrin deposits in dermal blood vessels. | ***For diagnosis*** Same as CSVV Evidence of streptococcal infection Circulating IgA complexes ***For extent of disease*** Stool for occult blood Renal function tests | Corticosteroids Dapsone IVIG Factor VIII Replacement Ranitidine. | Self-limited. Mild relapses in 40% |

**Table 2** [45-54]

| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology Immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow-up* |
|---|---|---|---|---|---|
| **Predominantly medium-sized vessel vasculitis** | | | | | |
| Polyarteritis nodosa (PAN) | ***Cutaneous*** (20-50%)<br>Painful dermal and subcutaneous nodules,<br>"Starburst" pattern of livedo reticularis,<br>Healing with stellate shaped scars.<br>Digital gangrene.<br><br>***Systemic***<br>Weight loss,<br>Arthralgias,<br>Malaise,<br>Abdominal pain,<br>Mononeuritis multiplex,<br>hypertension,<br>Orchitis and<br>Congestive cardiac failure | Necrotizing obliterative arteritis<br>Focal panniculitis<br>Aneurysms may form in weakened arterial walls.<br>Luminal thrombosis | ***For diagnosis***<br>Detailed history<br>Thorough examination<br>Skin biopsy<br><br>***For extent of disease***<br>Muscle and sural nerve biopsies<br>Renal biopsy and angiography<br>p-ANCA in 20% | NSAIDS, aspirin,<br>High dose steroids,<br>IVIG,<br>Methotrexate,<br>Sulfapyridine,<br>Prostaglandins, and<br>Nifedipine.<br><br>Ribavirin with<br>PEX in HBV associated disease. | Outcome favourable, sometimes regressing spontaneously but high recurrence rate with prolonged course. |

**Table 3** [55-60]

| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow-up* |
|---|---|---|---|---|---|
| **Predominantly small and medium sized vessel vasculitis** | | | | | |
| Microscopic polyangiitis (MPA) | ***Cutaneous*** Palpable purpura<br><br>***Systemic*** Constitutional symptoms, Necrotizing GN (79-90%), Pulmonary hemorrhage (12-29%). | ***Histopathology*** Segmental vascular necrosis, Leucocytoclasia.<br><br>***Immunofluorescence*** Few or no immune deposits | Blood picture for leukocytosis, anemia, and ESR<br>C reactive protein<br>Urinalysis for RBCs, cast, and protein.<br>p-ANCA more than c-ANCA,<br>RF.<br>Chest radiography for pulmonary infiltrates.<br>Renal biopsy | ***Limited disease,*** Corticosteroids.<br><br>***Renal or pulmonary disease*** Pulse steroids<br>PEX<br>Cyclophosphamide<br><br>***For other systemic involvements*** Azathioprine<br>IVIG<br>Mycophenolate mofetil. | Frequent relapses.<br><br>*Poor prognostic markers are:* Increasing age<br>Raised cretonne levels,<br>Pulmonary hemorrhage<br>c-ANCA associated disease<br><br>***Follow-up*** ESR<br>Creatinine levels<br>Chest x-rays. |

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| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow-up* |
|---|---|---|---|---|---|
| Wegener's granulomatosis (WG) | ***Cutaneous*** (46-66%).<br>Palpable purpura<br>Oral ulcers<br>Papulonecrotic lesions<br>Subcutaneous nodules<br>Ulcers.<br><br>***Systemic***<br>Upper and lower respiratory tract symptoms (60-80%)<br>Renal disease(18%)<br>Conjunctivitis<br>Scleritis<br>Uveitis | ***Histopathology***<br>Perivascular leucocytoclastic and/or granulomatous infiltrate | Same as for MPA.<br>c-ANCA more than p-ANCA. | ***Limited disease***<br>Corticosteroids.<br><br>Remission induction,<br>Corticosteroids,<br>Cyclophosphamide<br>Methotrexate alone or in combination<br><br>***Remission maintenance***<br>Cyclophosphamide<br>PEX<br>Methotrexate<br>Azathioprine<br>Sulfamethoxazole-trimethprim.<br><br>***Refractory disease***<br>Antithymocyte globulin<br>Anti-CD4 and antiCD52 monoclonal antibodies | Same as for MPA. |

| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow-up* |
|---|---|---|---|---|---|
| Churg-Strauss syndrome (CSS) | ***Cutaneous*** (40-70%)<br>Palpable purpura<br>Subcutaneous nodules<br>Urticaria<br>Livedo reticularis<br>Papulonecrotic lesions.<br><br>***Systemic***<br>Allergic rhinitis,<br>Nasal polyps,<br>Asthma, Pneumonitis,<br>Gastroenteritis,<br>Necrotizing GN,<br>Mononeuritis multiplex,<br>Granulomatous myocarditis | Eosinophilic infiltrate of tissue,<br>Formation of extra vascular granulomas of visceral and cutaneous tissue,<br>Necrotizing vasculitis of arteries and veins. | Same as for MPA<br>Eosinophilia,<br>Raised IgE levels.<br>P-ANCA more than C-ANCA. | Same as for MPA. | Same as for MPA |
| Drug-induced | ***Cutaneous***<br>Acral purpuric plaques and nodules,<br>Digital gangrene.<br><br>***Systemic***<br>Glomerulonephritis<br>Pulmonary hemorrhage. | Lymphocytic vasculitis,<br>Little leucocytoclasia,<br>tissue eosinophilia. | Drug history<br>Eosinophilia<br>Normal complement levels<br>ANCA may be positive. | Withdrawal of offending drug.<br>Corticosteroids,<br>Immunosuppressive agents. | Skin lesions heal by stopping the offending drug<br><br>***Follow-up***<br>Renal function<br>Urinalysis |

| *Classification (Based on vessel size)* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow-up* |
|---|---|---|---|---|---|
| Vasculitis associated with connective tissue diseases (SLE, RA, Sjogren's syndrome) | ***Cutaneous***<br>Palpable purpura, petechiae,<br>Digital infarcts,<br>Ulcers,<br>Nodules,<br>Livedo reticularis,<br>Urticaria or papulonecrotic lesions.<br>Punched out ulcers represent systemic vasculitis.<br><br>***Systemic***<br>GI tract, heart, lungs, or kidneys in RA.<br><br>Central and peripheral nervous system, GIT, lungs, heart, and genitourinary system in SLE.<br><br>CNS, GIT, muscle, kidney, and parotid glands in Sjogren's syndrome. | ***Histopathology***<br>Vascular changes are infrequent in SLE but hyaline changes and fibrinoid degeneration can occur in vascular walls.<br><br>***Immunofluorescence***<br>IgG and C3 deposits at basement membrane zone | Connective tissue profile<br>Similar histopathological and IF studies. | ***For mild cases***<br>Corticosteroids<br>Penicillamine.<br><br>***For severe cases***<br>Cyclophosphamide<br>Azathioprine<br>Chlorambucil<br>MTX<br>PEX. | Recurrences common<br>Response to treatment good<br><br>***Follow-up***<br>Renal function test,<br>Echocardiography,<br>Pulmonary function test |

**Table 4** Other disorders showing LCV [61, 63-65]

| *Other disorders showing LCV* | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow up* |
|---|---|---|---|---|---|
| Erythema elevatum diutinum | ***Cutaneous***<br>Red violaceous, red brown, or yellowish papules over dorsa of hands, knees, buttocks, and Achilles tendon, healing with atrophic scars.<br><br>**Systemic**<br>None | Grenz zone<br>Leucocytoclastic vasculitis<br>Eosinophils in upper and mid-dermis<br>Older lesions show fibrosis and mixed infiltrate<br>Cholesterol deposits in the intra and extracellular tissue | ***For diagnosis***<br>Detailed history<br>Clinical examination.<br>Skin Biopsy<br><br>***For associated ailment***.<br>Autoimmune profile,<br>ASOT, HIV serology,<br>Protein electrophoresis,<br>Bone marrow biopsy | Dapsone<br>Nicotinamide,<br>High potency topical or intralesional steroids<br>Other therapies used in CSVV | New crops may appear for 5-35 years |
| Granuloma faciale | ***Cutaneous***<br>Soft brown red nodules or plaques on face<br><br>***Systemic***<br>None | ***Histopathology***<br>Grenz zone,<br>Leucocytoclastic vasculitis,<br>Neutrophilic and eosinophilic infiltrate<br><br>***Immunofluorescence***<br>Deposition of IgG in and around dermal vasculature | Clinically typical lesion.<br>Typical histopathology | Intralesional steroids, with or without cryosurgery.<br>Clofazamine<br>Dapsone<br>Surgery<br>Laser | Recurrences are common<br>Resistant to treatment |

**Table 5** Neutrophilic vascular reactions [62, 66-69]

| | *Clinical features* | *Histopathology immunofluorescence* | *Work up* | *Treatment options* | *Prognosis and follow up* |
|---|---|---|---|---|---|
| Pyoderma gangrenosum (PG) | Papular or pustular lesion with violaceous undermined edges, later ulcerating and healing with cribriform scar. | Central necrosis and ulceration of epidermis and dermis surrounded by an intense neutrophilic infiltrate<br>No leucocytoclasia | Detailed history<br>Physical examination,<br>Skin biopsy for histopathology and culture for bacteria, mycobacteria, fungi and occasionally viruses<br>Syphilis serology | ***Mild cases***<br>Topical or intralesional steroids<br>Hydrophilic occlusive dressings<br>Tacrolimus etc.<br><br>***Severe cases***<br>Oral steroids<br>Clofazamine<br>Dapsone<br>Azathioprine<br>Cyclosporin<br>Cyclophosphamide<br>Methotrexate or Tacrolimus | Recurrences may occur but are unpredictable. |
| Sweet's syndrome | Constitutional symptoms<br>Painful erythematous papules, plaques, nodules,<br>Psuedovesicular, ustular lesions | Dense neutrophilic infiltrate in upper dermis,<br>Neutrophil karyorrhexis,<br>Leucocytoclasia.<br><br>***Negative IF*** | ***For diagnosis***<br>Fever, typical skin lesions &<br>typical histopathology<br>Neutrophilia<br><br>***For associations***<br>Evidence of malignancy, respiratory or GI infections, connective tissue profile | Excellent response to:<br>Systemic steroids,<br>Potassium iodide and Colchicine.<br><br>Somewhat lesser response to:<br>Dapsone<br>Clofazamine<br>Cyclosporin<br>Indomethacin<br>Etretinate<br>Alpha interferon. | Same as PG |

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| *Neutrophilic vascular reactions* | *Clinical features* | *Histopathology immunofluorescence* | *Workup* | *Treatment options* | *Prognosis and follow up* |
|---|---|---|---|---|---|
| Nodular vasculitis | Tender, dusky, often suppurative nodules or plaques on posterolateral legs of obese women with venous stasis | Leucocytoclastic vasculitis of subcutaneous tissue leading to necrotic reaction in subcutaneous tissue and deep dermis | ***For diagnosis***<br>Same as for CSVV<br>Deep skin biopsy.<br><br>***For etiology***<br>Mantoux test<br>Chest radiography | ***Tuberculous patients***<br>Antituberculosis treatment for a minimum of 9 months.<br>Tuberculosis not present<br>Supportive measures<br>Systemic steroids<br>Potassium iodide or any treatment mentioned for CSVV | Same as PG |
| Bowel associated dermatitis arthritis syndrome | Constitutional symptoms followed by crops of macular, papular or pustular lesions, and oral ulcers | Same as Sweet's syndrome | Same as for CSVV<br>Barium studies and endoscopy | ***For bowel pathology***<br>Surgical correction<br><br>***For others***<br>Tetracycline<br>Metronidazole<br>Erythromycin | Same as PG |

ANCA, antineutrophilic cytoplasmic antibodies; CSSV, cutaneous small vessel vasculitis; CV, cryoglobulinemic vasculitis; HSP, Henoch-Schonlein purpura; HUV, hypocomplementemic urticarial vasculitis; IF, immunofluorescence. IVIG, intravenous immunoglobulin; MPA, microscopic polyangiitis; MTX, methotrexate; NUV, normocomplementemic vasculitis; PEX, plasma exchange; PG, pyoderma gangrenosum; WG, Wegener's granulomatosis

## References


1. Fiorentino DF. Cutaneous vasculitis. *J Am Acad Derm*atol 2003; **48**: 311-40.
2. Stone JH, Nousari HC. "Essential" cutaneous vasculitis: What every rheumatologist should know about vasculitis of skin. *Curr Opin Rheumatol* 2001; **13**: 23-34.
3. Stone JH, Calabrese LH, Hoffman GS *et al.* Vasculitis. A collection of pearls and myths. *Rheum Dis Clin North Am* 2001; **27**: 677-728.
4. Harper L. Small vessel vasculitides. *Medicine* 2002; **30**: 15-20.
5. Carline OS Savage. Medium vessel vasculitides. *Medicine* 2002; **30**: 21-5.
6. Jennette JC, Falk RJ. Small vessel vasculitis. *N Engl J Med* 1997; **337**: 1512-23.
7. Jennette CJ, Milling DM, Falk RJ. Vasculitis affecting the skin. A review. *Arch Dermatol* 1994; **130**: 899-906.
8. Stegeman CA, Kallenberg CG. Clinical aspects of primary vasculitis. *Springer Semin Immunopathol* 2001; **23**: 231-51.
9. Sais G, Vidaller A, Jucgla A *et al.* Prognostic factors in leukocytoclastic vasculitis: a clinicopathologic study of 160 patients. [see comments]. *Arch Dermatol* 1998; **134**: 309-15.
10. Scott DG, Watts RA. Classification and epidemiology of systemic vasculitis. [see comments]. *Br J Rheumatol* 1994; **33**: 897-9.
11. Ekenstam E, Callen JP. Cutaneous leukocytoclastic vasculitis. Clinical and laboratory features of 82 patients seen in private practice. *Arch Dermatol* 1984; **120**: 484-9.
12. Jessop SJ. Cutaneous leucocytoclastic vasculitis: a clinical and aetiological study. *Br J Rheumatol* 1995; **34**: 942-5.
13. Gyselbrecht L, DeKeyser F, Ongenae K *et al.* Etiological factors and underlying conditions in patients with leucocytoclastic vasculitis. *Clin Exp Rheumatol* 1996; **14**: 665-8.
14. Blanco R, Martinez-Taboada VM, Rodriguez-Valverde V, Garcia-Fuentes M. Cutaneous vasculitis in children and adults. Associated diseases and etiologic factors in 303 patients. *Medicine* (Baltimore) 1998; **77**: 403-18.
15. Swerlick RA, Lawley TJ. Cutaneous vasculitis: its relationship to systemic disease. *Med Clin North Am* 1989; **73**: 1221-35.
16. Vena GA, Cassano N. Immunosuppressive therapy in cutaneous vasculitis. *Clin Dermatol* 1999; **17**: 633-40.
17. Atzori L, Ferreli C, Biggio P. Less common treatment in cutaneous vasculitis. *Clin Dermatol* 1999; **17**: 641-7.
18. Callen JP. Colchicine is effective in controlling chronic cutaneous leukocytoclastic vasculitis. *J Am Acad Dermatol* 1985; **13**: 193-200.
19. Wells GC. Allergic vasculitis (tri-symptom of Gougerot) treated with dapsone. *Proc R Soc Med* 1969; **62**: 665-6.
20. Allen NB, Bressler PB. Diagnosis and treatment of the systemic and cutaneous necrotizing vasculitis syndromes. *Med Clin North Am* 1997; **81**: 243-59.
21. Boehm I, Bauer R. Low-dose methotrexate controls a severe form of polyarteritis nodosa. *Arch Dermatol* 2000; **136**: 167-9.
22. Heurkens AH, Westedt ML, Breedveld FC. Prednisone plus azathioprine treatment in patients with rheumatoid arthritis complicated by vasculitis. *Arch Intern Med* 1991; **151**: 2249-54.
23. Ferri C, Greco F, Longombardo G *et al.* Antibodies to hepatitis C virus in patients with mixed cryoglobulinemia. *Arthritis Rheum* 1991; **34**: 1606-10.
24. Lamprecht P, Gause A, Gross WL. Cryoglobulinemic vasculitis. *Arthritis Rheum* 1999; **42**: 2507-16.
25. Agnello V, Chung RT, Kaplan LM. A role for hepatitis C virus infection in type II cryoglobulinemia. [see comments]. *N Eng J Med* 1992; **327**: 1490-5.
26. Ramos-Casals M, Trejo O, Garcia-Carrasco M *et al.* Mixed cryoglobulinemia: new concepts. *Lupus* 2000; **9**: 83-91.
27. Pascual M, Perrin L, Giostra E, Schifferli JA. Hepatitis C virus in patients with cryoglobulinemia type II. *J Infect Dis* 1990; **162**: 569-70.
28. Cohen SJ, Pittelkow MR, Su WP. Cutaneous manifestations of cryoglobulinemia: clinical and histopathologic study of seventy-two

patients. *J Am Acad Dermatol* 1991; **25**: 21-7.
29. Durand JM, Cacoub P, Lunel-Fabiani F *et al*. Ribavirin in hepatitis C related cryoglobulinemia. *J Rheumatol* 1998; **25**: 1115-7.
30. Monti G, Saccardo F, Rinaldi G *et al.* Colchicine in the treatment of mixed cryoglobulinemia. *Clin Exp Rheumatol* 1995; **13** (Suppl 13): S197-9.
31. Ballare M, Bobbio F, Poggi S *et al.* A pilot study on the effectiveness of cyclosporine in type II mixed cryoglobulinemia. *Clin Exp Rheumatol* 1995; **13** (Suppl 13): S201-3.
32. Kuhl V, Vogt T, Anghelescu I. Intravenous immunoglobulin and prednisolone treatment of cryoglobulinemic polyneuropathy. *Nervenarzt* 2001; **72**: 445-8.
33. Wisnieski JJ. Urticarial vasculitis. *Curr Opin Rheumatol* 2000; **12**: 24-31.
34. Mehregan DR, Hall MJ, Gibson LE. Urticarial vasculitis: a histopathologic and clinical review of 72 cases. *J Am Acad Dermatol* 1992; **26**: 441-8.
35. Davis MD, Daoud MS, Kirby B *et al.* Clinicopathologic correlation of hypocomplementemic and normocomplementemic urticarial vasculitis. *J Am Acad Dermatol* 1998; **38**: 899-905.
36. Wisnieski JJ, Baer AN, Christensen J *et al*. Hypocomplementemic urticarial vasculitis syndrome. Clinical and serologic findings in 18 patients. *Medicine* 1995; **74**: 24-41.
37. Nurnberg W, Grabbe J, Czarnetzki BM. Urticarial vasculitis syndrome effectively treated with dapsone and pentoxifylline. *Acta Derm Venereol* 1995; **75**: 54-6.
38. Saulsbury FT. Henoch-Schonlein purpura in children. Report of 100 patients and review of the literature. *Medicine* (Baltimore) 1999; **78**: 395-409.
39. Rosenblum ND, Winter HS. Steroid effects on the course of abdominal pain in children with Henoch-Schonlein purpura. *Pediatrics* 1987; **79**: 1018-21.
40. Rostoker G, Desvaux-Belghiti D, Pilatte Y *et al.* High-dose immunoglobulin therapy for severe IgA nephropathy and Henoch-Schonlein purpura. *Ann Intern Med* 1994; **120**: 476-84.
41. Lamireau T, Rebouissoux L, Hehunstre JP. Intravenous immunoglobulin therapy for severe digestive manifestations of Henoch-Schonlein purpura. *Acta Paediatr* 2001; **90**: 1081-2.
42. Fukui H, Kamitsuji H, Nagao T *et al.* Clinical evaluation of a pasteurized factor XIII concentrate administration in Henoch-Schonlein purpura. *Japanese Pediatric Group Thromb Res* 1989; **56**: 667-75.
43. Narin N, Akcoral A, Aslin MI, Elmastas H. Ranitidine administration in Henoch-Schonlein vasculitis. *Acta Paediatr Jpn* 1995; **37**: 37-9.
44. Lhote F, Cohen P, Guillevin L. Polyarteritis nodosa, microscopic polyangiitis and Churg-Strauss syndrome. *Lupus* 1998; **7**: 238-58.
45. Guillevin L, Lhote F, Cohen P *et al*. Polyarteritis nodosa related to hepatitis B virus. A prospective study with long-term observation of 41 patients. *Medicine* (Baltimore) 1995; **74**: 238-53.
46. Guillevin L, Lhote F. Polyarteritis nodosa and microscopic polyangiitis. *Clin Exp Immunol* 1995; **101**: 22-3.
47. Diaz-Perez JL, Winkelmann RK. Cutaneous periarteritis nodosa. *Arch Dermatol* 1974; **110**: 407-14.
48. Guillevin L, Lhote F, Leon A *et al.* Treatment of polyarteritis nodosa related to hepatitis B with short-term therapy with anti-viral agents and plasma exchanges. A prospective trial in 33 patients. *J Rheumatol* 1993; **30**: 289-98.
49. Calderon MJ, Landa N, Aguirre A, Diaz-Perez JL. Successful treatment of cutaneous PANS with pentoxifylline. *Br J Dermatol* 1993; **128**: 706-8.
50. Gedalia A, Sorensen R. Intravenous immunoglobulin in childhood cutaneous polyarteritis nodosa. *Clin Exp Rheumatol* 1998; **16**: 767.
51. Schartz NE, Alaoui S, Vignon-Pennamen MD *et al.* Successful treatment in two cases of steroid-dependent cutaneous polyarteritis nodosa with low-dose methotrexate. *Dermatology* 2001; **203**: 336-8.
52. Bajema IM, Hagen EC. Evolving concepts about the role of antineutrophil cytoplasm autoantibodies in systemic

vasculitides. *Curr Opin Rheumatol* 1999; **11**: 34-40.
53. Niles J, Bottinger E, Saurina G *et al*. The syndrome of lung hemorrhage and nephritis is usually an ANCA-associated condition. *Arch Intern Med* 1996; **156**: 440.
54. Guillevin L, Durand-Gasselin B, Cevallos R *et al*. Microscopic polyangiitis: clinical and laboratory findings in eighty-five patients. *Arthritis Rheum* 1999; **42**: 421-30.
55. Guillevin L, Lhote F. Treatment of polyarteritis nodosa and microscopic polyangiitis. *Arthritis Rheum* 1998; **41**: 2100-5.
56. Frances C, Du LT, Piette JC *et al*. Wegener's granulomatosis. Dermatological manifestations in 75 cases with clinicopathologic correlation. *Arch Dermatol* 1994; **130**: 861-7.
57. Davis MD, Daoud MS, McEvoy MT, Su WP. Cutaneous manifestations of Churg-Strauss syndrome: a clinicopathologic correlation. *J Am Acad Dermatol* 1997; **37**: 199-203.
58. Stone J, Tun W, Hellman D. Treatment of non-life-threatening Wegener's granulomatosis with methotrexate and daily prednisone as the initial therapy of choice. *J Rheumatol* 1999; **26**: 1134.
59. Patten S, Tomecki K. Wegener's granulomatosis: cutaneous and oral mucosal disease. *J Am Acad Dermatol* 1993; **28**: 710-8.
60. Langford CA. Treatment of polyarteritis nodosa, microscopic polyangiitis, and Churg-Strauss syndrome: where do we stand? [letter; comment] *Arthritis Rheum* 2001; **44**: 508-12.
61. Barham KL, Jorizzo JL, Grattan B, Cox NH. Vasculitis and neutrophilic vascular reactions. In: Burns T, Breathnach S, Cox N, Griffiths C, eds. *Rook's Textbook of Dermatology*. Oxford: Blackwell Science; 2004. p. 49.16-49.17.
62. Barham KL, Jorizzo JL, Grattan B, Cox NH. Vasculitis and neutrophilic vascular reactions. In: Burns T, Breathnach S, Cox N, Griffiths C, eds. *Rook's Textbook of Dermatology*. Oxford: Blackwell Science; 2004. p. 49.32-49.45.
63. Yiannias JA, el-Azhary RA, Gibson LE. Erythema elevatum diutinum: a clinical and histopathological study of 13 patients. *J Am Acad Dermatol* 1992; **26**: 38-44.
64. Van de Kerkhof PC. On the efficacy of dapsone in granuloma faciale. *Acta Derm Venereol* 1994; **74**: 61-2.
65. Lever WF, Schaumburg-Lever G, eds. *Histopathology of Skin*. New York: Lippincot, 1990.
66. Powell PC, Su WPD. Pyoderma gangrenosum: classification and management. *J Am Acad Dermatol* 1996; **34**: 395-409.
67. Chow RKP, Ho VC. Treatment of pyoderma gangrenosum. *J Am Acad Dermatol* 1996; **34**: 1047-60.
68. Van den Driesch P. Sweet's syndrome: Acute febrile neutrophilic dermatosis. *J Am Acad Dermatol* 1994; **31**: 535-56.
69. Malone JC, Slone SP, Wills-Frank LA *et al*. Vascular inflammation (vasculitis) in Sweet's syndrome. *Arch Dermatol* 2002; **138**: 345-9.

## Erratum

In the article "Histopathological spectrum of cutaneous leishmaniasis in North West Frontier Province" published in the October-December, 2004 issue of JPAD (*J Pak Assoc Dermatol* 2004; **14**: 209-14), the name of one of authors was misprinted as **Shagufta Nazir**. Her correct name is **Shagufta Nasir**.