A Case of Cutaneous Polyarteritis Nodosa

Chang Duk Kim, M.D., Byung Chun Kim, M.D., Kyu Suk Lee, M.D.

Department of Dermatology, Keimyung University School of Medicine Taegu, Korea

Cutaneous polyarteritis nodosa(CPAN) is a benign form of rare vasculitis of small and medium-size arteries with a recurrent but benign course without systemic involvement.

We experienced a 61-year-old male who had two months history of multiple deep-purpurish livedo reticularis on both lower legs. Noncutaneous manifestations including malaise, fever, myalgia, and arthritis were absent.

A skin biopsy specimen from the livedo reticularis on the leg showed perivascular and transmural neutrophilic and lymphocytic infiltration of medium-sized arteries in the dermal-subcutaneous junction and fibrinoid necrosis of the vessel walls. The patient was treated with colchicine for 2months and showed markded improvement.

(Ann Dermatol 15(1) 27~30, 2003).

Key Words : Cutaneous polyarteritis nodosa, Livedo reticularis, Colchicine

Polyarteritis nodosa(PAN) is a form of nodular vasculitis that most frequently involves the medium-sized muscular arteries of the kidneys, liver, heart, and gastrointestinal tract[1]. It is accompanied by substantial morbidity and mortality[2]. In the cutaneous form of PAN(CPAN), tender subcutaneous nodules with high fever, arthralgias and myalgias occur without major organ system involvement[3,4]. The histopathological features are those of a nodular arteritis, involving medium-sized arteries in the deep reticular dermis, showing extensive fibrinoid necrosis and vascular destruction.

CASE REPORT

A 61-year-old man visited our department because of multiple deep-purpurish livedo reticularis on both lower legs of 2 months duration. Noncutaneous manifestations including malaise, fever, myalgia, and arthritis were absent and he denied any history of fever, upper respiratory tract infections, jaundice, tuberculosis, and taking any drugs before skin eruption. On past history, he suffered from peripheral neuropathy for 1 year and on physical examination, he had an extensive livedo reticularis with patchy maculopapular, erythematous eruption on the lower legs. Other physical examinations were normal. The result of neurologic conduction velocity was peripheral polyneuropathy. Laboratory findings included an erythrocyte sedimentation rate(ESR) of 28mm/hr. Test results from the following laboratory studies were normal or negative ; complete blood count, urinalysis, serum urine nitrogen, creatinine, electrolyte, liver function test, hepatitis B surface antigen and antibody, antinuclear antibody, and cytoplasmic and perinuclear antineutophil cytoplasmic antibodies.

A skin biopsy specimen from the livedo reticularis on the right leg showed perivascular and transmural neutrophilic and lymphocytic infiltrates of medium-sized arteries in the dermal-subcutaneous junction and fibrinoid necrosis of the vessel walls. Surrounding the vessel was a mixed inflammatory cell infiltrate consisting of neutrophils, eosinophils, lymphocytes, an extravasated erythrocytes. The patient was treated with colchicine, 2.4mg for 2 months and showed marked improvement.

Received April 24, 2002
Accepted for publication September 30, 2002
Reprint request to : Chang Duk Kim, M.D., Department of Dermatology, Keimyung University School of Medicine, Taegu, Korea
Tel. 82-53-250-7624, Fax: 82-53-250-7626
E-mail. kmderma@dsmc.or.kr

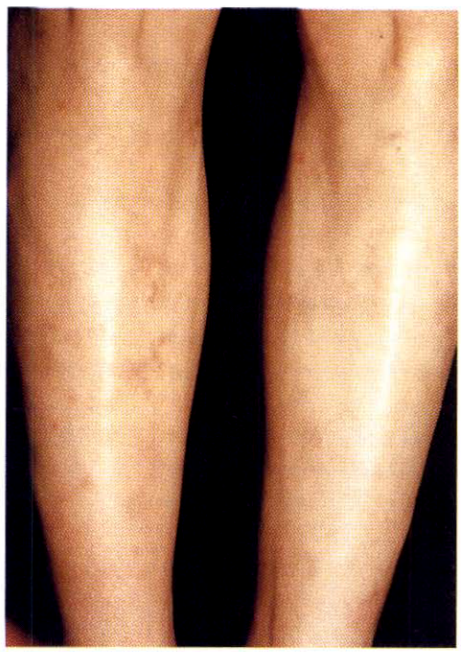

Fig. 1. Cutaneous lesion of livede reticularis on both legs.

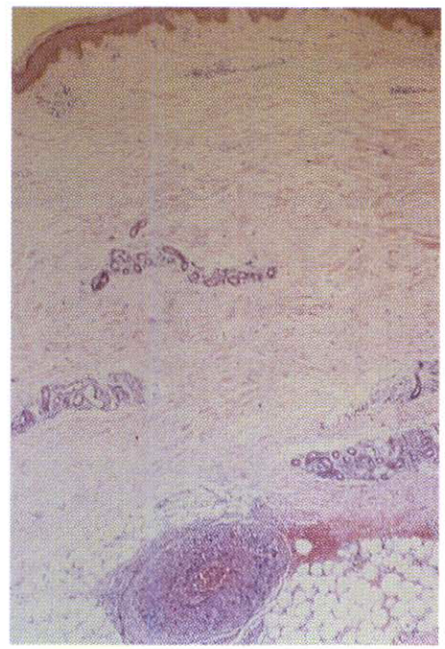

Fig. 2. Panarteritis of small and medium-sized arteries with little perivascular infiltration at dermal-subcutaneous junction (H&E, ×20).

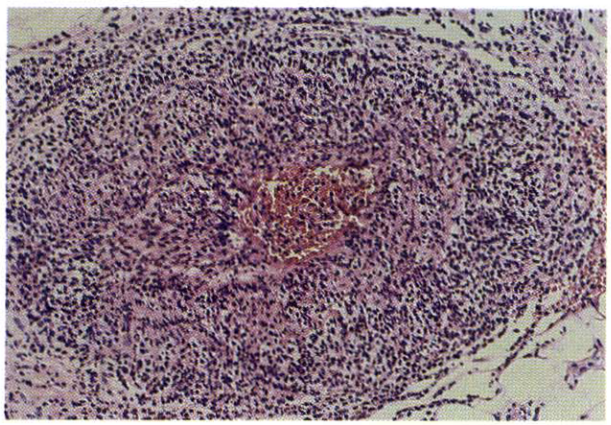

Fig. 3. Transmural and perivascular neutrophilic and lymphocytic infiltration of medium-sized artery with segmental fibrinoid necriosis and thrombotic occlusion of the arterial lumen (H&E, ×100).

DISCUSSION

Borrie and Diaz-Perez described a clinically and histopathologically distinct variant of PAN that they called benign CPAN[3,4]. It is a rare form of vasculitis that appears to be limited primarily to the skin, muscles and joints. In contrast to the systemic form of the disease it is characterized by the absence of visceral lesions and a relapsing but benign course[5]. Clinically, the cutaneous hallmarks of CPAN are tender, erythematous nodules, ranging from 0.5 to 2 cm in diameter, commonly occurring on the lower extremities, although any area of the integument may be affected. Livedo reticularis, especially in a starbust pattern, and skin ulcers are also common clinical features. Fever, arthralgias, nondestructive arthritis, myalgias, localized myositis, and neuropathy frequently occur[4,6]. In Korean literature, Kim et al showed that six of eight patients with a histologic diagnosis of CPAN presented with livedo reticularis on the lower legs[7]. Laboratory tests have usually been unremarkable, except for elevated ESR.

The etiology of CPAN remains unknown, but the presumption of immune-complex mediation in some is based on the demonstration of Ig M and C_3 deposits in lesional biopsy specimens and the detection of circulating immune complexes[8] and it has occurred in association with hepatitis B infection, post-streptococcal infection, tuberculosis, human immunodeficiency virus(HIV) infection, inflammatory bowel disease, falciparum malaria, diphtheria-tetanus immunization, and drugs[9,10].

In histologic aspect, CPAN affects arterioles at

the dermal-subcutaneous junction. Usually, a single vessel is maximally involved and typically a focal, septal panniculitis is present in adjacent tissue. There is a panarteritis with degeneration of the arterial wall, deposition of fibrinoid material, and destruction of the elastic laminae. Neutrophils are present in and around the vessel wall with leukoytoclasis. In contrast, the histopathologic pattern of systemic polyarteritis nodosa often shows small-vessel leukocytoclastic vasculitis of the superficial vascular plexus, and the deep dermis is rarely involved[11,12].

The prognosis of CPAN is favorable in both adults and children. If in the initial evaluation there are no systemic findings, very probably the patient will remain free of generalized disease. The course is benign but prolonged and is characterized by periods of remission disrupted by exacerbations, sometimes preceded by upper respiratory tract infections. Like the initial episode, such relapses show no systemic involvement. Follow-up laboratory evaluations should include urinalysis, erythrocytes sedimentation rate, and hemoglobin, as well as clinical evaluation of nervous, vascular, and muscular systems[4]. Whereas an elevated erythrocyte sedimentation rate is a bad prognostic sign in systemic PAN, in CPAN it is not[13].

Relapses and remissions are frequent either with or without treatment, and the benign course of CPAN dictates that conservative measures should be employed. Moderate doses of corticosteroids(1mg/kg/day of prednisone) may be sufficient to induce remission, while recalcitrant cases can be treated with an immunosuppressant drug, either alone or combined with oral steroids[14,15]. In a case of CPAN associated with Crohn's disease, sulfapyridine was successful and penicillin has been shown to be beneficial in CPAN associated with streptococcal infection[16]. Other treatments which have been reported include nonsteroidal anti-inflammatory drugs, chloroquine, pentoxifylline[10], immunosuppressive drugs, colchicine, dapsone, and intravenous immunoglobulins were also used with efficacy. Colchicine is an alkaloid isolated from the autumn crocus, Colchicum autumnale and produces its anti-inflammatory effects by binding to the intracellular protein tubulin, thereby preventing its polymerization into microtubules and leading to the inhibition of leukocyte migration and phagocytosis. Assicot et al[16] reported three cases of CPAN in children well controlled by colchicine and salicylotherapy. Corticosteroid therapy was used only for invalidating symptoms and the development of neuromuscular signs may warrant the use of general corticosteroid therapy. A benign course and the absence of visceral involvement allow initiating a symptomatic treatment such as colchicine[16,17].

In summary, we have reported a case of CPAN, presented with livedo reticularis, with a good response to colchicine single therapy.

REFERENCES

1. Cupps TR, Fauci AS. The vasculidites. Philadelphia, Pa: WB Saunders Co.26-49, 1981.
2. Cohen RD, Conn DL, Ilstrup DM. Clinical features, prognosis and response to treatment in polyarteritis. Mayo Clin Proc. 55:146-155, 1980.
3. Borrie P: Cutaneous polyarteritis nodosa. Br J Dermatol 87:87-95, 1972.
4. Diaz-Perez JL, Winkelman RK: Cutaneous periarteritis nodosa. Arch Dermatol 110:407-414, 1974.
5. Ozen S, Besbas N, Saatci U, Bakkaloglu A: Diagnostic criteria for polyarteritis nodosa in childhood. J Pediatr 120:206-209, 1992.
6. Moreland LW, Ball GV: Cutaneous polyarteritis nodosa. Am J Med 88:426-429, 1990.
7. Kim YS, Lee CW, Park MH: Clinical features of cutaneous polyarteritis nodosa. Kor J Dermatol 33:225-231, 1995.
8. Diaz-Perez JL, Schroeter AL, Winkelman RK: Cutaneous periarteritis nodosa: immunofluorescence studies. Arch Dermatol 116:56-58, 1980.
9. Albornoz MA, Benedetto AV, Korman M, MCFall S, Toutellotte CD, Myers AR: Relapsing cutaneous polyarteritis nodosa associated with streptococcal infections. Int J Dermatol 37:664-666, 1998.
10. Charoen C, Kunya J: Cutaneous polyarteiritis nodosa: a report of a case associated with melioidosis. Int J Dermatol 37:433-435, 1998.
11. Minkowitz G, Smoller BR, McNutt S: Benign cutaneous polyarteritis nodosa: relationship to systemic polyarteritis nodosa and to heptitis B infection. Arch Dermatol 127:1520-2523, 1991.
12. Dean R, Sunil S: Cutaneous periarteritis nodosa. 29:611-615, 1990.
13. Ginarte M, Pereiro M, Toribio J: Cutaneous polyarteritis nodosa in a child. Ped Dermatol 15:103-107, 1998.
14. Jorizzo JL, White WL, Wise CM, Zanolli MD, Sherertz EF: Low-dose weekly methotrexate for un-

usual neutrophilic vascular reactions: cutaneous polyarteritis nodosa and Behcet's disease. J Am Acad Dermatol 24:973-978, 1991.
15. Rodriguez C, Luelmo J, Boronat M, Malvehy J, FitoA, Carrascosa A: Periarteritis nodosa cutanea en ninos. A proposito de un caso. An Esp Pediatr 38:255-257, 1993.
16. Assicot C, Bourrat E, Prigent F, Moraillon I, Vignon MD, Triller R et al. Cutaneous polyarteritis nodosa in children. Ann Dermatol Venereol 129:207-211, 2002.
17. Maillard H, Szczesniak S, Martin L, Garot D, Machet MC, Machet L et all. Cutaneous polyarteritis nodosa: diagnostic and therapeutic aspects of 9cases. Ann Dermatol Venereol 126:125-129, 1999.