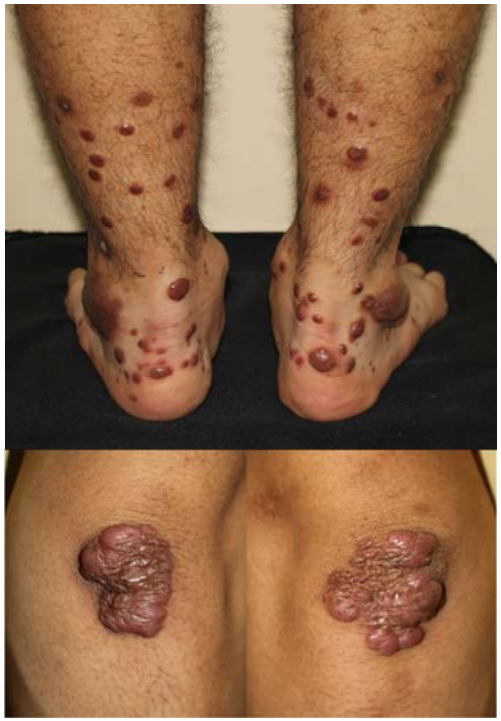
Case 7
A 21-year-old Thai man from Bangkok

Chief complaint: Multiple papules and nodules on bilateral elbows and legs for 4 months


(Fig. 7.1)

Present illness: He developed multiple asymptomatic erythematous papules and nodules on bilateral lower legs for four months. Two months later similar lesions appeared on bilateral extensor surfaces of his elbows. All of the lesions have rapid progression in size. He has no fever or arthritis.

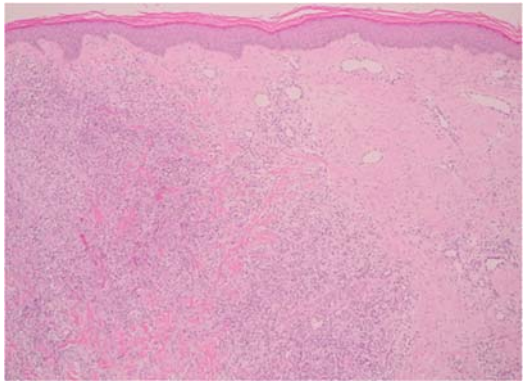
Past history: No underlying diseases

Personal history: Homosexual man with history of unsafe sex

Physical examination:
Vital signs: T 37.2˚C, BP 133/79 mmHg, P 95 beats/min, RR 20/min
HEENT: No pale conjunctivae, anicteric sclera, no oral thrush
Lymph nodes: Not palpable
GI: No hepatosplenomegaly
Neuro: No stiffness of neck

Dermatologic examination: (Fig. 7.1)
Multiple discrete erythematous to violaceous papules and nodules, some coalesce to form plaques on bilateral lower legs and extensor surfaces of the elbows

Histopathology: (S16-038263, Left elbow) (Fig. 7.2)


(Fig. 7.2)

- Dense nodular to diffuse inflammatory cell infiltrate of mainly neutrophils, nuclear dusts admixed with some lymphocytes and eosinophils
- Perivascular and dermal fibrosis are noted

Laboratory investigations:

- CBC: Hct 42%, WBC 7,830 cells/µL (N 65%, L 26%, Mono 5%, Eo 3%, Baso 1%), Platelets 370,000 cells/µL
- Anti HIV: Positive
- CD4 T cell 7% (220 cells/µL)
- Serum protein electrophoresis: Normal
- G6PD screening: Deficiency

Diagnosis: Erythema elevatum diutinum with 1st diagnosis of HIV infection

Treatment:

- Antiretroviral drugs
- Oral colchicine 0.6 mg three times a day
- 0.05% clobetasol propionate apply on lesions twice daily

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Discussion:

Erythema elevatum diutinum (EED) is a rare, chronic skin disease in the spectrum of cutaneous leukocytoclastic vasculitis.[1,2] The incidence of EED is unknown.

EED was first described in elderly men in 1888 by Hutchinson.[3] and in young women with a history of rheumatic disease in 1889 by Bury.[4] Then in 1894, Radcliff-Crocker and Williams concluded that the two types should be described as one entity and so gave the name EED.[5]

Clinically, EED is characterized by symmetrical, erythematous to violaceous papules and nodules and then may coalesce to form larger nodules or plaques. The most common sites of involvement are the extensors surfaces of the hands, fingers, elbows, knees, legs, and Achilles tendon

The lesions are often asymptomatic; nevertheless, pain or burning sensation may occur.[1] There is no significant mortality associated with EED. Most patients are well although arthralgia and constitutional symptoms may be present. [5]

EED can occur at any age with a slight male predominance and no racial predilection, but most commonly affects patients in fourth and sixth decade, and it appears to present earlier in those with human immunodeficiency virus.[2]

The etiology of EED is not entirely clear, however, the prevailing and traditional theories are based on immune complex deposition within vessel walls, complement fixation, inflammation, and subsequent vascular destruction.[6]

The histologic findings of EED, early lesion begin with leukocytoclastic vasculitis with polymorphonuclear cell infiltration and fibrin deposition in superficial and mid-deep dermis.[4] Polymorphonuclear cells, macrophages, histiocytes and occasionally eosinophils may surround blood vessels in early stages.[2] Late stage fibrosis has been reported and this is most likely a consequence of chronic dermal injury.[7] Blood vessel dilatation with continual damage and hypertrophic endothelial cells may protrude into vessel lumen which may then stimulate granulation. In this case, lesions may be mistaken for Kaposi's sarcoma or bacillary angiomatosis.[8] Cholesterol crystals may develop in long-standing cases secondary

to erythrocyte extravasation.[9] The chronic form with more complex histology separates EED from other vasculidities. PMN leukocytes are always present, and their absence should prompt consideration of alternative diagnoses.[10]

EED has been reported in association with a wide range of disease including bacterial infection (commonly streptococcal), autoimmune disease, lymphoma, IgA monoclonal gammopathy which was found to be the most commonly related heamatological disorder. The recurrent or flare up of EED occur in association with these disease activities.[11] Recently, there have been several reports of EED in association with HIV[12,15] and in some cases, EED was reported as the first manifestation for diagnosing HIV infection.[13] Nodular lesions which are rare variants in general population most often develop in HIV patient and can easily misdiagnosed as bacillary angiomatosis or Kaposi sarcoma.[14] Physicians should have awareness of the presence of EED in those infected with HIV.

As EED is often associated with a variety of underlying disease, the physician may need to search for underlying disease and treatment of underlying disease is of clinical benefit.

For the treatment of EED, limited treatment options are available for EED, with most case using dapsone. Treatment courses are long and relapse is common. From review article dapsone monotherapy was effective in 80% of cases.[11] Dramatic response is usually seen within 48 hours of initiation of therapy with near complete resolution within weeks to month.[16] Dapsone also be the treatment of choice in HIV related EED with 85% of cases show good response.[11] But in nodular lesion dapsone may be ineffective due to fibrosis of lesion, adding in other agents such as intralesional steroid, colchicine, sulphonamide may be benefit. Surgical excision is another option.[15]

In subject whom dapsone is not appropriate, sulphate antibiotics, colchicine would be the choice.[5,17] However, a few cases have reported the successful use of such agents.

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