CASE 9

Patient: A 25 year old woman from Bangkok

Chief Complaint: Painful eruption on both hands

Present Illness: She presented to our clinic with painful purplish eruption on both hands for one week duration. She is currently a second year science student who has to work mainly in the laboratory. Prior to this illness, she has been working in the lab where she describe the environment that is cool, approximately below 10 degree Celsius and even sometimes below freezing point temperature, at least minus 4 degree Celsius. Furthermore, she has also reported frequent exposure to the fridge and freezer including regular contacts with some cold vaporizing chemical agents like dry ice, acetone and dichlormethane. The lesions first appeared as tender red to orange bumps and some of which progressed into larger thin plaques with dramatic color change into purple. They have been proceeding over a week with increasing pain on touching. She also experienced slight intermittent numbness at tip of fingers but no alteration in color mentioned.

Review of Systems: unremarkable

Past History: no known underlying disease

Family History: non contributory

Dermatological Examination (Figure 9.1-2)**:**

- Right hand, lateral aspect of 2^{nd} to 4^{th} fingers: multiple discrete red to orange papules without an overlying epidermal change, mild tenderness on palpation.
- Left hand, dorsal aspect, over 2^{nd} to 4^{th} knuckle area: a well defined markedly purplish thin dermal plaque with no epidermal changes, slightly tender.

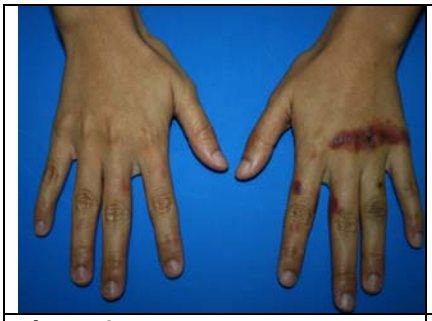

Figure 9.1

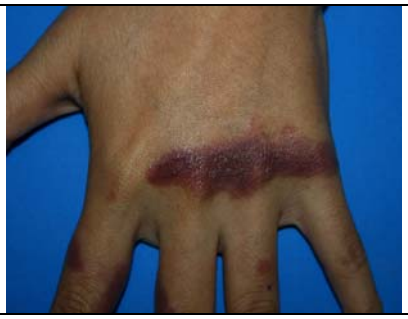

Figure 9.2

Histopathology (S10-9715) (Figure 9.3-4):

- vacuolar alteration of basal cell layer and occasional necrotic keratinocytes in the epidermis
- superficial and deep perivascular and periadnexal infiltrate of lymphocytes and extravasated erythrocytes
- interface infiltration of lymphocytes and some nuclear dust along the dermoepidermal junction

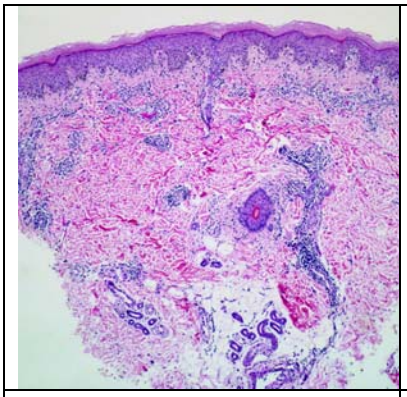

Figure 9.3, H&E 40x

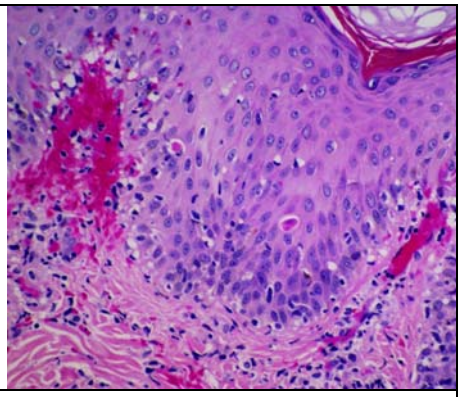

Figure 9.4, H&E 200x

Investigations: Normal CBC, ESR and negative ANA, Anti Ro and La

Diagnosis: Idiopathic perniosis or Chilblains

Presenters: Wikanda Panmanee

Marinya Pongpudpunth

Consultant: Kumutnart Chanprapaph

Discussions:

Chilblains or perniosis are inflammatory cutaneous lesions that occur in association with cold exposure and are characteristically localized to acral skin, thighs and buttock. It is relatively common in cold and humid climate countries of Northwestern Europe and North America.[1-3] Chilblains is an Anglo-Saxon term meaning chill and blegen (sore), where as perniosis is a Latin word that is synonymous.[4]

While the clinical aspect suggest the diagnosis in majority of cases, the chilblain can be difficult to distinguish from acral lesions of lupus erythematosus (LE), especially in the case of the so-called chilblain lupus.

Perniosis occurs most commonly among young women between the ages of 15-30 years but may occur among older individuals or children and persons with low body mass index.[5] Perniosis develops among susceptible individuals who are continuously exposed to nonfreezing cold. Absolute temperature is less important than the cooling of non-adapted tissue.[1] Characteristically, the lesions begin in the fall or winter and disappear in the spring or early summer. Acute perniosis may develop 12-24 hours after exposure to the cold. The lesions are characterized by single or multiple erythematous, purplish, edematous papules or plaques, accompanied by intense pain, itching or burning sensation. In severe cases, blister, pustules, and ulceration may occur. They tend to affect the toes and dorsum of proximal phalanges; however, nose, ears and face are also reported.[6]

A variety of conditions have been described in association with perniosis. Among adults, perniosis has been reported secondary to systemic lupus erythematosus, lupus anticoagulant, anticardiolipin, and antiphospholipid antibodies, chronic myeloid leukemia, anorexia nervosa, metastases from carcinoma of the breast, and reaction to medication; sulindac.[7,8] These may present as acral perniotic lesion that mimic perniosis.

The histopathologic diagnosis of chilblains has been variably reported. The clear distinction between idiopathic versus secondary perniosis is still controversial. The typical changes of idiopathic perniosis were deep dermal infiltrate with lymphocytic exocytois localized to acrosyringia with or without accompanying interface dermatitis.[7] In addition, the epidermal necrosis[9], epidermal spongiosis[10], dermal edema[10] are also reported. The presence of lymphocytic vasculitis was emphasized by Ackerman et al and was mentioned in the cases described by Herman et al.[11] Some studies suggested that the vasculitis was a more frequent finding in cases of secondary perniosis, where thrombosis affected the reticular dermal blood vessels, whereas vascular injury in cases of idiopathic perniosis tended to be confined to the superficial dermal capillaries.[7]

When evaluating a patient with perniosis, it is important to exclude an underlying diagnosis and the additional testing is often necessary especially the screening for connective tissue disease.[5] Nevertheless, positive antinuclear antibody (ANA) without other clinical or laboratory abnormalities that warranted the diagnosis of SLE or any other specific collagen vascular disease was also reported in idiopathic perniosis.[7]

Spontaneous remission is common when the weather is warmer, and relapse is frequent during the following

winters. The main treatment is the protective measures with rest, warmth, thick and appropriate clothing. Nifedipine, calcium channel blockers is a drug of choices in severe idiopathic perniosis which is postulated to promote faster healing and prevent recurrence, albeit prone to undesirable side effects.[12-14] Other medications; pentoxyfylline, oral and topical steroids are also documented.[15,16] In the case of persistant or chronic perniosis, the long-term follow-up of these patients is warranted to exclude associated connective tissue disease.[17]

In our patient, the history of constant exposure to frigid environment, altogether with the conventional clinical picture, common histologic findings and normal laboratory studies, these are consistent with idiopathic perniosis. The avoidance of cold was recommended and high potency topical steroid was described. The improvement was gradually seen on 2 weeks follow-up with some residual hyper and hypo-pigmentation and no new lesions observed. In conclusion, this case is unique because it occurred in tropical country and most likely to be occupational related perniosis which has never been previously mentioned.

References

1. Wolff K, Goldsmith LA, Katz SI, Gilchrest BA, Pallor AS, Leffell DJ. Fitzpatrick's dermatology in general medicine. 7th ed. Mcgraw-hill medical;2007;849-50.
2. Ackerman AB, Chongchitnant N, Sanchez J, Guo Y. Histologic diagnosis of inflammatory skin diseases: an algorithmic method based on pattern analysis. 2nd ed. Baltimore:Williams&Wilkins;1997;606-7.
3. Jacob JR, Weisman MH, Rosenblatt SI, Bookstein JJ. Chronic pernio: a historical perspective of cold-induced vascular disease. Arch Intern Med.1986; 146: 1589-92.
4. Spraycar M, ed. Stedman's medical dictionary. 26th ed. Baltimore, MD: Williams & Wilkins; 1995.
5. Simon TD, Soeb JB, Hollister JR. Pernio in Pediatrics. Pediatrics 2005;116; 472-475.

6. Fritz RL, Perrin DH. Cold exposure injuries: prevention and treatment. Clin Sports Med. 1989; 8:111-28.
7. Crowson AN, Magro CM. Idiopathic perniosis and its mimics: a clinical and histological study of 38 cases. Hum Pathol 1997;28:478-84.
8. Reinertsen JL. Unusual pernio-like reaction to sulindac. Arthritis Rheum. Sep 1981;24:1215-18.
9. Cribier B, Djeridi N, Peltre B, Grosshans E. A histologic and immunohistochemical study of chilblains. J Am Acad Dermatol 2001;45:924-29.
10. Boada A, Bielsa I, Fernandez- Figueras MT. Perniosis: clinical and histopathological analysis. Am J Dermatopathol 2010;32:19-23.
11. Herman EW, Kezis JS, Silvers DN. A distinctive variant of pernio: clinical and histopathologic study of nine cases. Arch Dermatol 1981;117:26-8.
12. Rustin MH, Newton JA, Smith NP, Dowd PM. The treatment of chilblains with nifedipine: the results of a pilot study, a double-blind placebo-controlled randomized study and a long-term open trial. Br J Dermatol 1989;120: 267-75.
13. Almahameed A, Printo DS. Pernio (chilblains). Curr Treat Options Cadiovasc Med. 2008;10: 128-35.
14. Patra AK, Das AL, Ranadasan P. Diltiazem vs. nifedipine in chilblains: a clinical trial. Indian J Dermatol Venereol 2003;69: 209-11.
15. Noaimi AA, Fadheel BM. Treatment of perniosis with oral pentoxyfylline in comparison with oral prednisolone plus topical clobetasol ointment in Iraqi patients. Saudi Med 2008;29:1762-64
16. Ganor S. Corticosteroid therapy for pernio. J Am Acad Dermatol 1983; 8:136.
17. Viguier M, Pinquier L, Cavelier-Balloy B et al. Clinical and histopathologic features and immunologic variables in patients with sever chilblains. A study of the relationship to lupus erythematous. Medicine (Baltimore) 2001;80:180-88.