CASE 9

A 21-year-old Thai female from Nakornsrithammarat

Present history

The patient presented with rash on her right arm and thigh for 5 months. Initially, the lesion began with slightly itchy, dull red-colored macules on the forearm and thigh for 3 weeks. It gradually extended distally to the lower arm and turned into brownish macule and patches.

Past history:

She is otherwise healthy.

Physical examination

Vital signs: normal
HEENT: not pale, no icteric sclerae
Heart & lung: WNL
Skin: Discrete brownish macules and non-branchable sclerotic patches in linear distribution along right arm and right thigh.

Histopathology (S05-12110)

Superficial and deep perivascular and interstitial infiltrate of lymphocyte admixed with some plasma cells

Foci of dermal sclerosis in the reticular dermis.

Diagnosis: Linear morphea
Presenter: Thanya Suwansathit
Consultant: Somyot Charuwichitratana

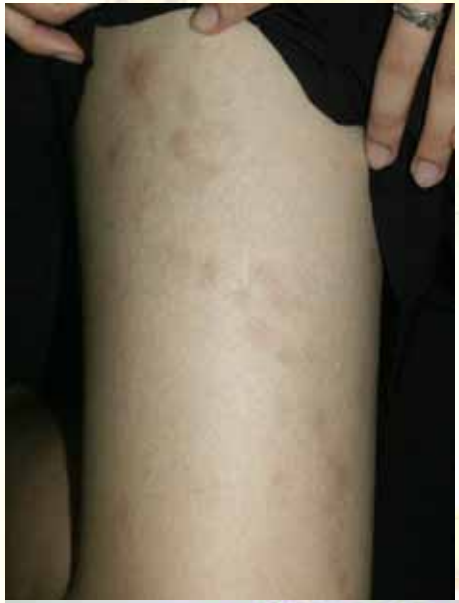

Fig. 9.1

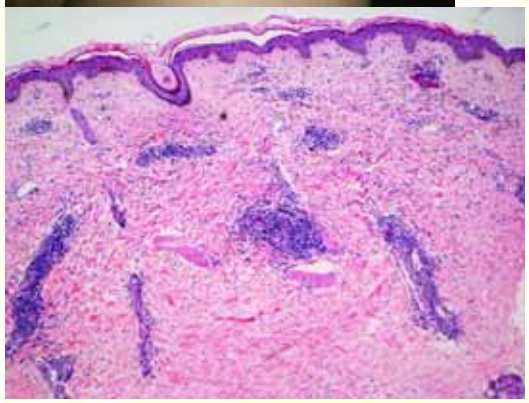

Fig. 9.2

Discussion:

Linear morphea is an uncommon form of localized scleroderma, and three subtypes exist. The classic form occurs as a linear, sclerotic band that extends the length of an extremity. *En coup de sabre*, or facial linear morphea, typically presents as a single linear plaque on the forehead and frontal scalp. *Parry-Romberg syndrome* is considered to be a severe variant of linear morphea, manifesting as progressive facial hemiatrophy with associated epilepsy, exophthalmos, and alopecia.

Linear morphea constitutes approximately 20% of all cases of localized scleroderma. It occurs most often during the first two decades of life. About 50% of patients diagnosed with linear scleroderma are yonger than 20 years. Females are three times more likely to have the disease than males. Initial lesions are dull red or violaceous macules, which then progress to hard, atrophic, bound-down, and ivory-colored plaques. An important distinction between linear morphea and more typical morphea is that the former often affects the underlying fascia, muscle and tendons, in addition to the skin. Therefore, limb involvement may result in muscle weakness and shortening, joint contractures, and frozen joints. Calcinosis within linear morphea lesions also can develop.

The pathogenesis of localized scleroderma in general is unknown. Many consider localized scleroderma to be an immunologic disorder, since it has been described in patients with a variety of autoimmune disease including SLE, DLE, Hashimoto's thyroiditis, vitiligo and often accompanied by the presence of autoantibodies. Linear morphea has two distinct immunologic features that support an autoimmune etiology. Compared to other forms of localized scleroderma, linear morphea is more frequently associated with high ANA titers. Also, anti-single stranded DNA antibodies are particularly common in this condition. Another theory implicates *Borrelia burgdorferi*, the tick-borne spirochete that causes Lyme disease, as the cause of localized scleroderma. Evidence for this has been found in Europe and Japan, but studies done in North America have had contradictory findings. Other proposed etiologies include focal vascular abnormalities, derangement of fibroblast activation, and deficiency of collagen-degrading matrix metalloproteinases.

Left untreated, linear morphea typically undergoes gradual progression. Spontaneous improvement has been reported, mainly in patients with the classic subtype. Reported treatments for linear morphea include topical, intralesional, and oral cortico-steroids; oral penicillin; penicillamine; methotrexate; and phototherapy with PUVA or UVA-1. Topical calcipotriene 0.005% oilment may be an effective treatment for localized scleroderma, but double-blind placebo controlles studies are needed to be confirmed. Distressing complications of linear sclero-derma include muscle atrophy, joint disability resulting from extension of the lesions over joints. Therefore, physical therapy is vital in cases of linear morphea with joint involvement.

References:

1. Fleischmajor R. Scleroderma. In: Freedberg IM, Eisen AZ, Woof K, Austen KF, Goldsmith LA, Karz SI, eds. Fitzpatrick's Drematology in General Medicine Volume II. 6th ed. McGraw-Hill; 2003. p.1710-11.
2. Sehgal VN, Srivastava G, Aggarwal AK, Choudhary M, Bajaj P. Localized scleroderma/morphea. Int J Dermatol 2002 Aug;41(8):467-75.
3. Hunzelmann N, Scharffetter Kochanek K, Hager C, Krieg T. Management of localized scleroderma. Semin Cutan Med Surg 1998;17(1):34-40.
4. Tuffanelli DL. Localized scleroderma. Semin Cutan Med Surg 1998;17(1):27-33.
5. Dervis E, Acbay O, Barut G, Karaoglu A, Ersoy L. Association of vitiligo, morphea, and Hashimoto's thyroiditis. Int J Dermatol 2004;43(3):236-7.
6. Sapadin AN, Fleischmajer R. Treatment of scleroderma. 2002 Jan;138(1):99-105.
7. Dutz J Treatment options for localized scleroderma. Skin Therapy Lett 2000;5(2):3-5.
8. Cunningham BB, MD, Landells IDR, Langmen C, Sailer DE, Paller. Topical calcipotriene for morphea/linear scleroderma. J Am Acad Dermatol 1998;39:211-5
9. Majeed M, Al-Mayouf SM, Al-Sabban E, Bahabri S. Coexistent linear scleroderma and juvenile systemic lupus erythematosus. Pediatr Dermatol 2000;17(6):456-9.
10. Weide B, Walz T, Garbe C. Is morphoea caused by Borrelia burgdorferi? A review.Br J Dermatol 2000 Apr;142(4):636-44.