Case 19

Linear depression on the forehead

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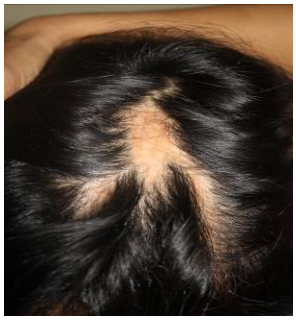
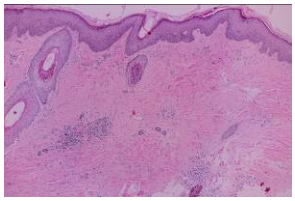
Patient: A 10 years old Thai girl from Rayong

Chief complaint: Scarring alopecia on the scalp for 3 years

Present illness: Three year previously, the patient gradually developed asymptomatic localized alopecia on the scalp. One year later, she developed a brownish atrophic patch on the forehead. There was no sign of inflammation before.

Past history: No underlying disease

Skin examination: Patches of scarring alopecia on the scalp with a brownish linear atrophic patch on forehead

Histopathology: (S14-9951, forehead)

There is dense inflammatory cell infiltrate of mostly lymphocytes in the superficial and deep dermis in association with a hint of homogenized collagen bundles.

Diagnosis: **Linear morphea (En coup de sabre)**

Investigation:

CBC: Hct 37.9%,Hb 12.4 g/dl, WBC 15,400 (N54%, L40%, Mo5%), Platelet 347,000/mm^3
ESR 23 mm/hr
LFT: AST 22 U/L, ALT 38 U/L, ALP 277 U/L
Thyroid function test: Normal
ANA: Negative
RF: Negative

Treatment:

- Prednisolone 0.5 mg/kg/day
- Methotrexate 7.5 mg weekly with folic acid 5 mg/day

Discussion:

En coup de sabre, as known as linear morphea or linear scleroderma is often observed in childhood or youth and is probably the most common variant of morphea in these age groups, affecting between 40% and 70% of the children studied. Presentation is typically with a single unilateral lesion having a linear distribution, most commonly on the limbs, face, or scalp. When the lesions are located on the scalp, they produce linear plaques of alopecia that are frequently atrophic and slightly depressed, and the indurated skin is smooth, shiny, and ivory-colored, though in some cases the plaques may be pigmented. The unilateral nature of these lesions, their preference for the parietal region, and their tendency to deform the bone, giving rise to depressed lesions. Lesions can sometimes extend onto or exclusively affect the cheek, nose, or upper lip. At these sites, there is often only a mild linear pigmentation on the skin surface, but deeper involvement can produce deformities and asymmetry of facial structures and alterations of dental implantation. When the sclerotic disorder affects a whole side of the face, it is called progressive facial hemiatrophy or Parry-Romberg syndrome.[1] En coup de sabre can present with refractory partial seizures even before the characteristic skin lesions develop. Other neurologic complications associated with the disease include neuromyotonia, mimics of hemiplegic migraine, dystonia, trigeminal neuralgia, and brain cavernomas.[2]

Neuroimaging findings are more frequently ispilateral to the skin lesions, but contralateral involvement has been described. Neurologic symptoms should not be used as a predictor for MRI abnormalities because neurologic lesions have been discovered in asymptomatic patients. Moreover, symptomatic patients were sometimes proven to have normal radiologic exams. Sometimes, there is a smooth depression in the frontoparietal skull, with thinning of the diploic space, but no significant cerebral abnormality. Even though the brain is often normal on MRI in en coup de sabre, subtle cortical atrophy, white matter lesions and focal subcortical calcification have been described on CT and MRI. SPECT imaging (single photon mission computed tomography) has been shown to be more sensitive than MRI in detecting subtle changes in cerebral blood flow, with diminished blood flow in the frontalortemporal lobes in patients with en coup de sabre. Cerebral angiograms and magnetic resonance angiograms studies showed vascular involvement suggestive of vasculitis.[3,7]

Histopathologic findings of morphea can vary depending on the stage and depth of the lesion, but do not differ based on subtype. In the earliest phase, lesions of morphea can be highly inflammatory with a perivascular, interstitial, syringotropic and perineural array of lymphocytes and plasma cells. In later lesions, there is a lack of dermis. Folliculosebaceous units are typically absent and atrophic eccrine glands appear to be positioned higher in the dermis because of replacement of the subcutis by pale sclerotic, homogenized collagen bundles.[4-6]

Laboratory investigations are nonspecific. The diagnosis of linear morphea en coup de sabre is clinical and based on characteristic cutaneous and soft tissue findings. Currently no diagnostic laboratory tests exist. Nonetheless, 37–50% of the patients may present a positive ANA test (homogenous or speckled patterns), as well as anti-single-stranded-DNA antibodies. Antinucleosome antibodies, soluble interleukine-2 receptor, and recently, antiagalactosyl immunoglobulin G antibodies have been reported in linear scleroderma. In some patients, autoantibody may be present even before the disease manifestation and patients with Scl-70, anticentromere, Ro/La, or U1RNP antibodies should be

followed closely, as systemic disease might ensue.[7] Skin biopsy will confirm the diagnosis and should be obtained early to avoid costly, invasive, and high-risk workups.[9]

Management must be based on the extent and the severity of the disease, and focus primarily on the risk of deformities and movement limitation. The recently published LoSCAT (Localized Scleroderma Cutaneous Assessment Tool) is a promising measurement tool as it appears to be able to differentiate between activity and damage. Using the LoSCAT, the physician can quantify damage to the skin and extracutaneous tissues in a patient with localized scleroderma by evaluating a series of symptoms and clinical signs of activity (skin hardening, erythema) and damage (atrophy and alterations of pigmentation) together with the results of a number of laboratory parameters. The drugs most widely accepted as useful in the treatment of this disease are methotrexate and systemic glucocorticoids almost always administered in combination. The methotrexate dosages varied between 0.3 and 0.4 mg/kg per week in children and between 15 and 25 mg per week in adults. Glucocorticoids are usually administered as high dose intravenous boluses of methylprednisolone, sometimes followed by a tapering regimen of oral prednisolone. It is generally agreed that the administration of high-dose boluses of glucocorticoids achieves the desired anti-inflammatory and immunomodulator effect with a lower risk of the side effects than can develop with long-term administration of these drugs. Adults are usually administered 1 gm/day of methylprednisolone on 3 consecutive days per month for a maximum of 6 months, whereas the dose in children is of 30 mg/kg/d of methylprednisolone (maximum dose, 500 mg/d) as intravenous boluses on 3 consecutive days, repeated each week or each month. Monotherapy with oral glucocorticoids at dosages between 0.5 and 1 mg/kg/day has been shown to be effective in some studies, but there is probably a higher risk of relapse after stopping treatment.

Broadband UV-A therapy with or without psoralen (bath, cream, or oral), UV-A1 (intermediate or low doses, 40 sessions at 3-5 sessions per week), and narrowband UV-B, have been used in the treatment of localized morphea. The mechanism underlying the beneficial effects of phototherapy in morphea is unknown. The majority of studies of phototherapy in morphea have used UV-A1. This form of UV radiation can induce apoptosis of Langerhans cells and T lymphocytes, reduce collagen synthesis, increase collagenase secretion, and alter the local concentrations of cytokines such as interleukin 6, transforming growth factor β, and interferon γ, which also affect collagen and glycosaminoglycan production, fibroblast growth, and the concentration of metalloproteinases in the matrix. Phototherapy is as effective in patients with darker skin phototypes (phototype IV or higher). The use of other immunosuppressants for the treatment of morphea has been proposed based on their application in systemic scleroderma. Mycophenolate mofetil, an immunosuppressant that is usually well tolerated, was found to be effective in children with morphea that had not responded to treatment with a combination of glucocorticoids and methotrexate. Physiotherapy is an option worth considering in patients in whom morphea has led to joint contractures and limitations of limb movement, although once again the true usefulness of such therapy has not been demonstrated by any studies.[1] In terms of the cosmetic correction of atrophic skin lesions, hyaluronic acid offers many advantages. Hyaluronic acid filler may be safely and successfully used as monotherapy for temporary cosmetic improvement of en coup de sabre lesions. The benefit is

most prominent in well-selected patients who may experience atrophy but in whom the prominent feature is not tethering to underlying structures.[8] If the lesion is narrow, it can be resected and directly sutured. However, in the case of wide lesion, simple resection and direct suture are difficult. In that case, soft-tissue expansion with autologous bone grafting, autologous fat transplantation, dermal grafting, and en bloc autologous fat graft, can be used to fill the defect in the skin. However, the result often has proved unsatisfactory.[10] Autologous tissue cocktail injection appears to be a useful method for the correction of depressed atrophy of linear scleroderma en coup de sabre.[10]

In summary, we present a case of young female with classic linear morphea (en coup de sabre). She presented with scarring alopecia on scalp and brownish atrophic patch on forehead. She is not a known case of other autoimmune disease and has no abnormality in laboratory testing at presentation. The clinical and histopathological findings suggest the diagnosis of linear morphea. She has started her treatment with oral methotrexate and prednisolone. The treatment result is to be followed up.

References:

1. Bielsa Marsol I.,Update on the Classification and Treatment of Localized Scleroderma. Actas Dermosifiliogr. 2013; 104(8): 654-66.
2. Mohan SV.,Nittur V.&Steven KJ., Late-onset en coup de sabre of the skull. Skeletal Radiol. 2013; 42: 1447–50.
3. Amaral TN, Peres FA, Lapa AT, Marques-Neto JF,& Appenzeller S. Neurologic involvement in scleroderma: A systematic review. Semin Arthritis Rheum. 2013; 43(3): 335-47.
4. Taniguchi T.,Asano Y.,Tamaki Z.,Akamata K.,Aozasa N.,Nada S.,Takahashi T.,Ichimura Y.,Toyama T.,Sumida H.,Kuwano Y.,Miyanaba K.,Sato S., Histological features of localized scleroderma 'en coup de sabre': a study of 16 casesJ Eur Acad Dermatol Venereol. 2013 Oct 10.
5. Goh C.,Biswas A.,Goldberg LJ.,Alopecia with perineural lymphocytes: a clue to linear scleroderma en coup de sabre. J Cutan Pathol. 2012: 39: 518–20.
6. Pierre Louis M.,Sperling LC.,Hordinsky MK., Distinctive histopathologic findings in linear morphea (en coup de sabre) alopeciaJ Cutan Pathol. 2013: 40: 580–4.
7. Amaral TN, Marques Neto JF, Lapa AT, Peres FA, Guirau CR, Appenzeller S., Neurologic Involvement in Scleroderma en Coup de Sabre. Autoimmune Dis. 2012; 719685.
8. Thareja SK.,Sad hwani D.,Alan Fenske N.,En coup de sabre morphea treated with hyaluronic acid filler. Report of a case and review of the literature. Int J Dermatol. 2013 Oct 29.
9. Kraus V.,Lawson EF.,Von Scheven E.,Tihan T.,Garza J.,Nathan RG.,Cordoro KM.,& Waubant E., Atypical Cases of Scleroderma en Coup de Sabre. J Child Neurol. 2014; 29(5): 698-703.
10. Oh CK.,Lee J.,Jang BS.,Kang YS.,Bae YC.,Kwon KS.,Jang HS.,Treatment of Atrophies Secondary to Trilinear Scleroderma En Coup de Sabre by Autologous Tissue Cocktail Injection. Dermatol Surg. 2003; 29(10): 1073-5.