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Original Article

Upsurging cases of en coup de sabre: Distinctive clinical manifestations in a series of 41 patients

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Abstract

Background En coupe de sabre is not uncommon connective tissue disorder of children and adult and often end with marked deep scarring that starts very early and ending with scarring of the skin and scarring alopecia of scalp.

Objective The aim of current study is to screen for patients with en coup de sabre among cases with morphea and to evaluate the different clinical and histopathological features of the disease.

Methods This is cross sectional descriptive clinical study where all patients with morphea were screened for en coup de sabre and their clinical manifestations including scarring alopecia. Full clinical history and examination were carried for all patients that were seen during the period from 2014-2023 years. Biopsies for histopathological evaluation were carried out.

Results A total number of 181 patients with different types of morphea were seen. En coup de sabre was detected in 41 (22.65%) cases with female to male ratio was 2.72:1. The age onset of the disease was ranged from 4-28 years. Five (12.19%) were observed cases with face only but no scalp involvement and scarring alopecia was detected in 36(87.8%) of patients. One of the distinctive early features of the en coup de sabre of the face was the presence of rectangular mostly hyperpigmented and sometimes hypopigmented bands. Histopathological assessment showed mild acanthosis with normal dermal-epidermal junction but with pigmented basal layer while the dermis revealed mild inflammatory reaction with collagen deposition in early lesions while complete skin sclerosis was noticed in late lesions.

Conclusion En coup de sabre is a disease of children and young ages and mostly seen among females. There are early and late distinctive clinical and histopathological pictures. It is common cause of scarring of the face and scalp. Early detection of these patients is mandatory to prevent permanent complications.

Key words
En coupe de sabre; Morphea; Localized scleroderma.

Introduction

Morphea is an inflammatory cutaneous disorder of unknown etiology with fibrosis limited to the skin and underlying tissue which may lead to functional and cosmetic impairment.[1] It was a rare skin problem among Iraqis in the 1970s but the disease has increased since the 1980s and increased tremendously after 1991and it is no longer a rare variety today.[2]

Morphea can be divided into five types: Circumscribed, generalized, linear, deep and mixed morphea.[3]

Circumscribed morphea presents as single or

multiple oval/ round lesions limited to the epidermis and dermis while generalized morphea is characterized by ≥4 lesions in at least 2 of 7 anatomic sites. Linear morphea is the most common subtype during childhood or adolescence with a broad spectrum of clinical manifestations. It usually appears on the head (en coup de sabre) or extremities, but it can affect the trunk and it may involve the dermis, subcutaneous tissue, muscle or even bone. Deep morphea involves the dermis, subcutaneous tissue, fascia, and muscle with poorly circumscribed plaques. Mixed morphea is a combination of any of the previous types of morphea.[3]

En coup de sabre' (ECDS) is an uncommon form of localised scleroderma affecting the frontoparietal (forehead) area of the head.[4] The condition typically presents with a linear band of fibrotic skin on the scalp and present as a localised alopecia.[5] The incidence of ECDS is fewer than 0.5 per 100,000 people,[6] with females and Caucasians more commonly affected than males and other ethnicities.[7] Two-thirds of cases develop in childhood and studies have found the average age of onset between 6.9 and 13.6 years.[8, 9]

While the initial etiology of ECDS is unknown, the disease is caused by an autoimmune reaction causing inflammation leading to fibrosis and sclerosis of the affected skin and underlying tissues.[4] Studies have implicated a number of triggers, including sporadic autoimmune reactions, genetic mosaicism, trauma, injection and Borrelia infection.[10] There is no identified genetic origin, but genetic factors likely play a role in susceptibility, as the disease is more prevalent in families with a history of other autoimmune conditions.[11]

As with other types of localised scleroderma, studies suggest that elevated levels of Th1 cytokines promote an inflammatory reaction at the affected site, recruiting and activating T-lymphocytes and monocytes to the frontoparietal area.[12] In some patients, lesions in ECDS appear to follow Blaschko's lines,[13] representing pathways of cell migration and proliferation during embryonic development. Therefore, some researchers have suggested that ECDS is the result of genetic mosaicism (multiple genetically distinct cell populations arise within the same embryo, as seen in other conditions affecting skin pigmentation).[14] However, this is disputed by many studies, while others suggest that the disease develops along a dermatome (skin fed by a single nerve root).[15]

Progression from the inflammatory phase to the fibrotic phase is mediated by Th17 cytokines, where the blood flow becomes restricted and vasculature becomes damaged in the affected region and tissue fibrosis factors become elevated.[12,16,17] Th2 effector cytokines such as IL-4, IL-6 and TGB-β promote the proliferation of fibroblasts and the release of excess collagen and extracellular matrix proteins causing the thickened, hardened and discoloured band of fibrotic skin observed in the condition.[16,17]

The progression of ECDS often presents with depression of the frontal bone, atrophy and calcification of underlying tissues, and leads to neurological complications. These may include brain lesions (more commonly on the same side as the skin lesions), seizures, ocular defects, movement disorders, behavioral changes,

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headaches and migraines.[18-20] Early treatment of en coup de sabre with immunosuppressant such as methotrexate may help to slow disease development and prevent subsequent neurological symptoms.[21] However, in some cases, neurological symptoms have been reported before the onset of localized scleroderma.[22]

So the aim of the study is to screen for patients with ECDS among cases with morphea and evaluate the cutaneous manifestations including scarring alopecia among these patients.

Patients and methods

This is a cross-sectional descriptive clinical study where 181 patients with different types of morphea were screened for ECDS and the associated scarring alopecia during the period from March 2014-March 2023.

A complete medical history was obtained for all patients, including the following information: name, age, sex, address, occupation, patient complaints (e.g. pruritus, pain or burning), course and duration of disease, age at onset, associated systemic symptoms, associated autoimmune disease, and past medical, surgical, family and drug history.

A clinical examination is performed to determine the type, color, number, size and anatomic location of the lesion.

The diagnosis was achieved by clinical examination and supported by skin biopsy for hemotoxillin and eosin staining and pathological examination.

Results

A total number of 181 patients with different types of morphea were seen, their ages ranged between 4–65 years with mean±SD, 30±15.345, and their skin type was Fitzpatrick III and IV. There were 141 (77.9%) females and 40 (22.09%) males with the ratio of 3.52:1.

En coup de sabre was detected in 41 (22.65%) cases, their ages ranged from 2-32 years with a mean of 20 years with 31 (75.6%) cases under 18 years of age, with 30 (73.17%) females, 11 (26.82%) males, and female to male ratio was 2.72:1. The age at onset of the disease was ranged from 4-28 years with mean of 14 years and in 30 (73.17%) cases, onset of the disease was during childhood.

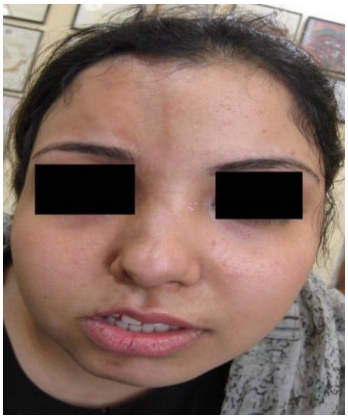

Figure 1 17-years old female with ECDS showing an atrophic, depressed rectangular band-like patch involving right side of the scalp, forehead and extending down into right side of the nose and upper lip.

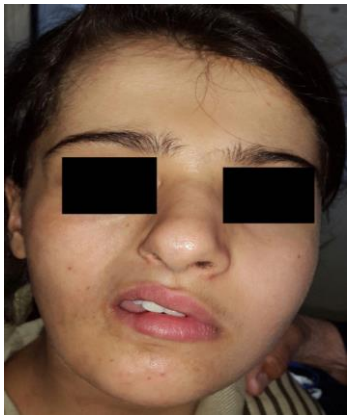

Figure 2 16-years old female patient with ECDS showing hemiatrophy of the right side of the face.

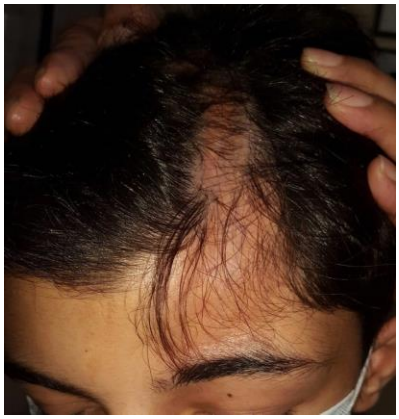

Figure 3 11-years old female with ECDS the scalp showing pigmented atrophic skin with scarring alopecia

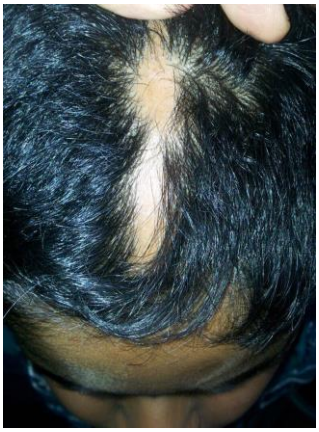

Figure 4 10-years old male patient with ECDS of the mid-scalp showing linear scarring alopecia.

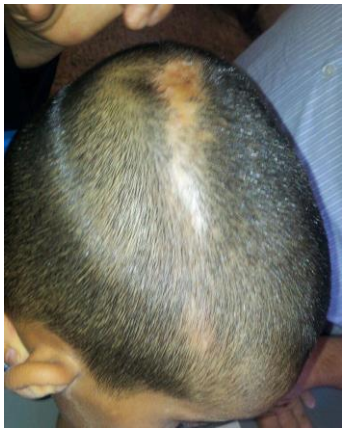

Figure 5 7-years old male with ECDS showing linear moth eaten scaring alopecia of the scalp

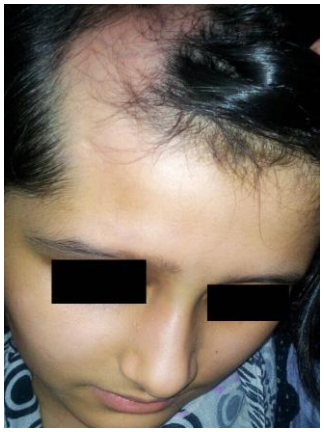

Figure 6 12-years old female patient ECDS showing wide area of scarring alopecia at the right side of the scalp extending into forehead

The duration of ECDS lesions ranged from 4 months to 5 years with a mean of 2 years. ECDS lesions were seen as sclerosed patches most commonly in the frontoparietal scalp and/or the para- median forehead and in a lesser extent in the face **(Figures 1-8)**. These lesions might eventually end with depressed, sclerosed, pigmented bands and causing atrophy of the underlying bone. When the lesion involved the scalp, there was often atrophy of the underlying bone and scarring alopecia as seen in 36 (87.8%) cases **(Figures 3-6)**. These scarring areas had started as moth eaten sites and coalesced together to end with big scarred patches **(Figure 6)**. All cases presented with unilateral involvement of the scalp or face and non of them had bilateral skin involvement.

Five (12.19%) patients showed only face involvement with no scalp scarring. **(Figures 2&8B)**. One of the distinctive and characteristic early features of the en coup de sabre of the face was the presence of rectangular mostly hyperpigmented and sometimes hypopigmented bands **(Figures 7A&8)** that progressively changed into sclerosed and atrophic patches with depressed underlying bone. No important systemic manifestations were observed in any patient.

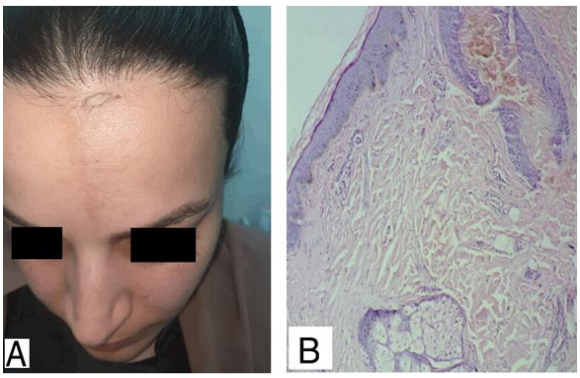

Figure 7 Showing a patient with very early rectangular pigmented lesion of ECDS (a) with her histopathology revealing pigmented basal layer and increased dermal collagen deposition with reduction of skin appendages. Hematoxylin and Eosin (H&E) stain 10X.

The histopathological assessment showed that, in the early lesion, there was mild acanthosis with normal dermal-epidermal junction but with pigmented basal layer while the dermis revealed mild to moderate, superficial and deep perivascular lymphocytic infiltration. In addition, there is a large amount of collagen deposition in the reticular dermis, which extended down to occupy the panniculus. **(Figures 7B&9)**.

While in late lesions, there were epidermal atrophy, focal damage of the dermal-epidermal junction, few melanophages in the hyalinized papillary dermis, scattered perivascular lymphocytic infiltration and increased collagen

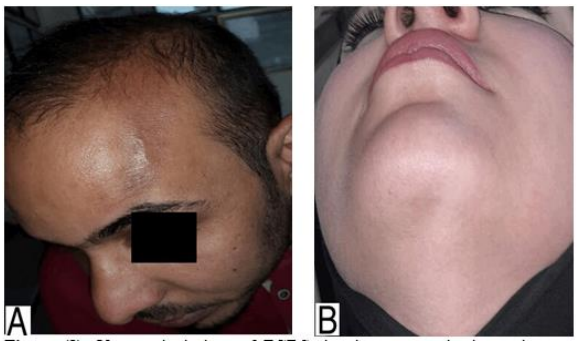

Figure 8 Very early lesions of ECDS showing rectangular hyperpigmented patch (A) and rectangular hypopigmented patch (B).

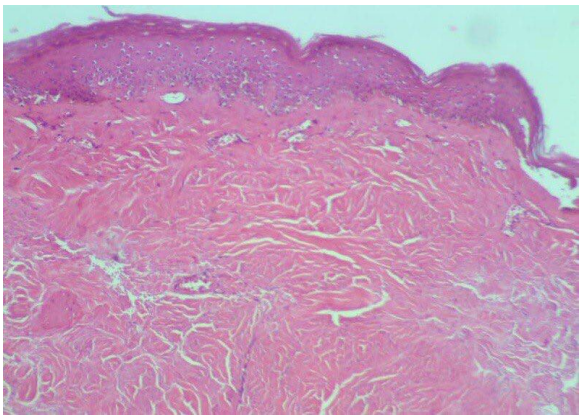

Figure 9 Early sclerosed lesion of ECDS showing epidermal acanthosis, perivascular infiltrate, vascular dilatation and increased collagen deposition. H&E stain 10X.

deposition with complete dermal sclerosis **(Figure 10)**.

Discussion

Morphea is a rare inflammatory disorder of the skin in Western countries and effects both children and adults.[23]

Morphea is an inflammatory disorder with usual childhood onset and female predominance. Its etiopathophysiology is not well known; but there is primary microvascular injury and abnormal T-lymphocyte response, which stimulate collagen overproduction and fibrosis. En coup de sabre is a specialized form of linear morphea, for which the pathogenesis is extremely complex and not yet fully understood.

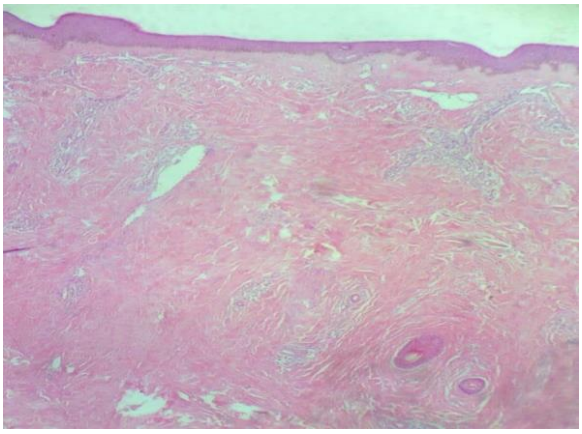

Figure 10 Late sclerosed lesion of ECDS showing mild atrophy of the epidermis, perivascular infiltrate, vascular dilatation and increased collagen deposition. H&E stain 4X.

Epidemiologic studies of morphea have recorded its incidence as 0.4 to 2.7 per 100,000 populations.[6,24] Morphea was a rare skin disorder in the 1970s in the Iraqi population.[2] Since 1980s morphea has been increasing and there was marked upsurge in cases after the Gulf War 1991. This increment happened in concordance with the rise in the incidence of other skin diseases especially, mycosis fungoids and many hematological malignancies.[25-28] The cause behind this tremendous increase of morphea and other skin diseases could not be well explained but it could be attributed to increased exposure to radiation from depleted uranium and chemical weapons during the subsequent wars in Iraq since early 1991. As en coup de sabre is considered a variant of morphea and its etiopathogesis, there was also definite increases in their cases and also could be attributed to repeated wars and uranium exposure.

The pathogenesis of morphea is not well known but the most accepted theory is multifactorial, involving both genetic and environmental factors that ultimately lead to damage of small blood vessels, release of profibrotic cytokines, and disruption of the balance between collagen synthesis and destruction.[29] and this similarly applied for ECDS.

In the present study a total number of 181 patients with different types of morphea were seen, their ages ranged between 4–65 years with mean±SD, 30±15.345 and there was predominance of females. These results were comparable to older study by the same author.[2] While en coupe de sabre was detected in 41 (22.65%) cases, their ages ranged from 2-32 years with a mean of 20 years with 31 (75.6%) cases under 18 years of age, with 30 (73.17%) females, 11 (26.82%) males and the female to male ratio was 2.72:1.

The present work showed that the mean age at the onset of ECDS morphea was 14 years and in 75.6% of cases, the age was below 18 years with female predominance (73.17%). These results were consistent with morphea cases as seen in the present work. These results were comparable to other studies and support a wildly pediatric onset of ECDS morphea, confirming that it is the disease of children.[6,8,9,30,31]

The clinical manifestation of ECDS were comparable with what has been published[4,7,19,31] but two distinctive new features were described in the present study. One of these characteristic feature was the rectangular bands affecting the face in the early course of the disease. While the second one was the hyperpigmented and sometimes hypopigmented bands that were observed in the early age of ECDS and then progressively changed into sclerosed and atrophic patches with depressed underlying bone.

ECDS can result in neurological, ocular, and dental complications.[18-20] Fortunately, for unknown reasons, none of our patients developed important systemic manifestations.

The histopathological results of our cases in early and late skin lesions were similar to ordinary morphea[2] and were comparable to other studies.[2,29]

Conclusion

En coup de sabre is increasing over the years and went parallel with tremendous sharp rise in cases of morphea. It is a common feature of morphea as it is seen in 22.65% of the total morphea patients while scarring alopecia is very common feature of ECDS as observed in 87.8 % of cases. There is a distinctive feature that was observed in the present work as the disease initially started as pigmented rectangular band as early manifestation and overtime changed into depressed fibrotic band with involvement of the underlying bone. The histopathological picture started as early changes, mainly basal pigmentation with dermal inflammatory reaction with collagen deposition. While the late changes revealed complete sclerosis with absence of skin appendages.

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References

1. Careta M, Romiti R. Localized scleroderma: clinical spectrum and therapeutic update. *Anais Brasileiros de Dermatologia*. 2015;**90(1)**:62-73.
2. Sharquie KE, Noaimi AA, Eman T. Abdulqader ET,Aljanabi WK. Clinical and histopathological evaluation of pigmented morphea with new insight in relation to etiopathogenesis of the disease. *Am J Dermatol Venereol*. 2020;**9(2)**:21-6.
3. Laxer RM, Zulian F. Localized scleroderma. *Curr Opin Rheumatol*. 2006;**18**:606-13.
4. Lausen SR, Daugaard-Jensen J, Lauridsen EF, Kjaer I. Localised scleroderma en coup de sabre affecting the skin, dentition and bone tissue within craniofacial neural crest fields. Clinical and radiographic study of 6 patients. *Eur Arch Paediatr Dentist*. 2019;**20**:339-50.
5. Graham PM, Gupta N, Altman DA. En coup de sabre. *Cutis*. 2019;**103(1)**:34-6.

6. Peterson LS, Nelson AM, Su WPD, Mason T, O'Fallon WM, Gabriel SE. The epidemiology of morphea (localised scleroderma in Olmsted County 1960-1993. *J Rheumatol*. 1997;**24(1)**:73-80.
7. Niklander S, Marin C, Martinez R, Esguep A. Morphea "en coup de sabre": an unusual oral presentation. *J Clin Exp Dent*. 2017; **9(2)**:e315-e318.
8. Chiu YE, Vora S, Kwon EM, Maheshwari M. A significant proportion of pediatric morphea en coup de sabre and Parry-Romberg syndrome patients have neuroimaging findings. *Pediatr Dermatol*. 2012;**29(6)**:738-48.
9. Tolkachjov SN, Patel NG, Tollefson MM. Progressive hemifacial atrophy: a review. *Orphanet J Rare Dis*. 2015;**10**:39.
10. Kreuter A. Localised scleroderma. Dermatologic therapy. 2012; 25(2):135-147.
11. Saracino AM, Denton CP, Orteu CH. The molecular pathogenesis of morphea: from genetics to future treatment targets. *Br J Dermatol*. 2017;**177(1)**:34-46.
12. Kurzinski K, Torok KS. Cytokine profiles in localised scleroderma and relationship to clinical features. *Cytokine*. 2011;**55(2)**:157-64.
13. Soma Y, Fujimoto M. Frontoparietal scleroderma (en coup de sabre) following Blaschko's lines. *J Am Acad Dermatol*. 1998;**38(2)**:366-8.
14. Wiebel L, Harper JI. Linear morphea follows Blaschko's lines. *Br J Dermatol*. 2008;**159**:175-81.
15. Gambichler T, Kreuter A, Hoffman K, Bechara FG, Altmeyer P, Jansen T. Bilateral linear scleroderma "en coup de sabre" associated with facial atrophy and neural complications. *BMC Dermatol*. 2001;1:9.
16. Badea I, Taylor M, Rosenberg A, Foldvari M. Pathogenesis and therapeutic approaches for improved topical treatment in localized scleroderma and systemic sclerosis. *Rheumatology*. 2009;**48**:213-21.
17. Kreuter A. Localised scleroderma. *Dermatol Therapy*. 2012;**25(2)**:135-47.
18. Amaral TN, Neto JFM, Lapa AT, Peres FA, Guirau CR, Appenzeller S. Neurologic Involvement in Scleroderma en Coup de Sabre. *Autoimmune Dis*. 2012:**2012**:719685.
19. Khamaganova I. Progressive Hemifacial Atrophy and Linear Scleroderma En Coup de Sabre: A Spectrum of the Same Disease? *Frontiers in Medicine*. 2018;**4**:2017.
20. Garofalo Gomez N, Novoa Lopez L, Gomez Garcia AM, Mendez Mendez M. Linear scleroderma en coup de sabre and epilepsy: presentation of a case in a child. *Neurologia*. 2012;**27(7)**:449-51.
21. Rattanakaemakorn P, Jorizzo JL. The efficacy of methotrexate in the treatment of en coup de sabre (linear morphea subtype). *J Dermatolog Treat*. 2018;**29(2)**:197-9.
22. Inci R, Inci MF, Ozkan F, Ozturk P. Frontal linear scleroderma en coup de sabre associated with epileptic seizure. *BMJ Case Rep*. 2012;**2012**:bcr2012007837.
23. Sehgal VN, Srivastava G, Aggarwal AK, Behl PN, Chou-dhary M, Bajaj P. Localized scleroderma/morphea. *Int J Dermatol*. 2002; **41**:467.
24. Murray KJ, Laxer RM. Scleroderma in children and adolescents. *Rheum Dis Clin North Am*. 2002;**28**:603-24.
25. Al-Hamamy H, Sharquie KE, Naomi A, Abdulwahhab W. Mycosis Fungoides in Iraqi patients-clinical, histopathological and immunohistochemical study. *J Cosmet Dermatol Sci Appl*. 2015;**5(2)**:116-24.
26. Sharquie KE, Al-Jaralla FA, Al-Saadawi AR. Hyperpigmented lichenoid mycosis fungoides as a new variant: clinical and histopathological evaluation. *Our Dermatol Online*. 2020;**11(e)**:e168.1-e168.5
27. Sharquie KE, Al-Jaralla FA, Taha S F. Mycosis fungoides in Iraqi population; changing in frequency, demographic and clinical subtypes. *J Pak Assoc Dermatol*. 2022;**32(3)**:510-16.
28. Sharquie KE, Jabbar RI. Triple therapy of Kaposi's sarcoma using oral propranolol,oral zinc sulfate and oral acyclovir as new initiative study. *J Pak Assoc Dermatol*. 2023;**33(1)**:72-7.
29. Fett N, Werth VP. Update on morphea: part I. Epidemiology, clinical presentation, and pathogenesis. *J Am Acad Dermatol*. 2011; **64(2)**:217–30.
30. Zulian F, Athreya BH, Laxer R, *et al*. Juvenile localized scleroderma: clinical and epidemiological features in 750 children. An international study. *Rheumatology (Oxford0*. 2006;**45**:614-20.
31. Tollefson MM, Witman PM. En coup de sabre morphea and Parry-Romberg syndrome: a retrospective review of 54 patients. *J Am Acad Dermatol*. 2007;**56(2)**:257-63.