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Case Report

Extensive extra genital Csillag's disease: a rare case report

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ABSTRACT

Lichen sclerosus et atrophicus (LSA), also called as Csillag's disease, is characterized by small, shiny, porcelain white, sclerotic papules, plaques and atrophic patches that can occur at any site on the skin including the mucosa. Most commonly found in genitalia, rarely occurring on extragenital skin. Predominantly seen in women with bimodal age distribution. Predominantly an interface dermatoses histopathologically consisting of epidermal atrophy, sclerosis /homogenization of collagen fibers and lichenoid inflammatory infiltrates in dermis. No effective treatment till date although topical steroids, calcineurin inhibitors, topical retinoids and systemic agents like hydroxychloroquine, mexthotrexate, PUVA shows varying inconsistent results. Here we report a case of 36-year-old female having LSA with extensive extra vulval involvement including face. No any other co-existing skin disorders. She showed partial improvement use of physical sunscreens, systemic antimalarials, oral methotrexate and antioxidants.

Keywords: LSA, Extensive, Extra genital

INTRODUCTION

Lichen sclerosus et atrophicus (LSA) also called as Csillag's disease is a benign immune mediated chronic muco cutaneous fibro inflammatory dermatosis of unknown aetiology and pathogenesis. It was described by Hallopeau in 1887 with anogenital (83% to 98%) and extragenital involvement (15% to 20%).[1-3] Extragenital LS (EGLS) can occur along with genital form. EGLS, present as white opalescent papules that may cluster as plaques and progressively result in parchment-like skin. It predominantly affects the vulval, perineal and perianal skin of prepubertal, perimenopausal and postmenopausal women. However, extra genital LSA is not uncommon and it was found in 805 of 5207 cases reviewed by Meffert et al.[1] Extra genital LSA is often noticed on the neck, shoulders, proximal extremities, upper trunk and thighs.[4] Although asymptomatic, some complaints of itching may be there. Uncommon sites include the palms, soles, scalp and face. LSA can be seen in association with morphea/scleroderma as overlapping lesions. Diagnosis is usually based on clinical suspicion followed by dermoscopy with hhistopathology of biopsied lesional skin confirming the diagnosis. Usually, extra genital LSA do not have carcinogenic potentials. However, possible malignant transformations have been described rarely.[5]

CASE REPORT

A 36-year-old female, presented to OPD with skin rashes for more than 7-year duration. She had occasional itching and burning. Its 1st developed over upper extremities on sun exposed areas and later on she developed over neck, nape, dorsum of foot and face. There was no significant past/family history. She was a known hypothyroid on thyroxine. General physical examination and systemic examination were unremarkable.

Local cutaneous examination revealed multiple ivory white atrophic papules, some coalesce to form plaques were seen over forehead, sides of face, neck, upper back, arms and forearms and dorsum of foot (Figure 1 A-F).

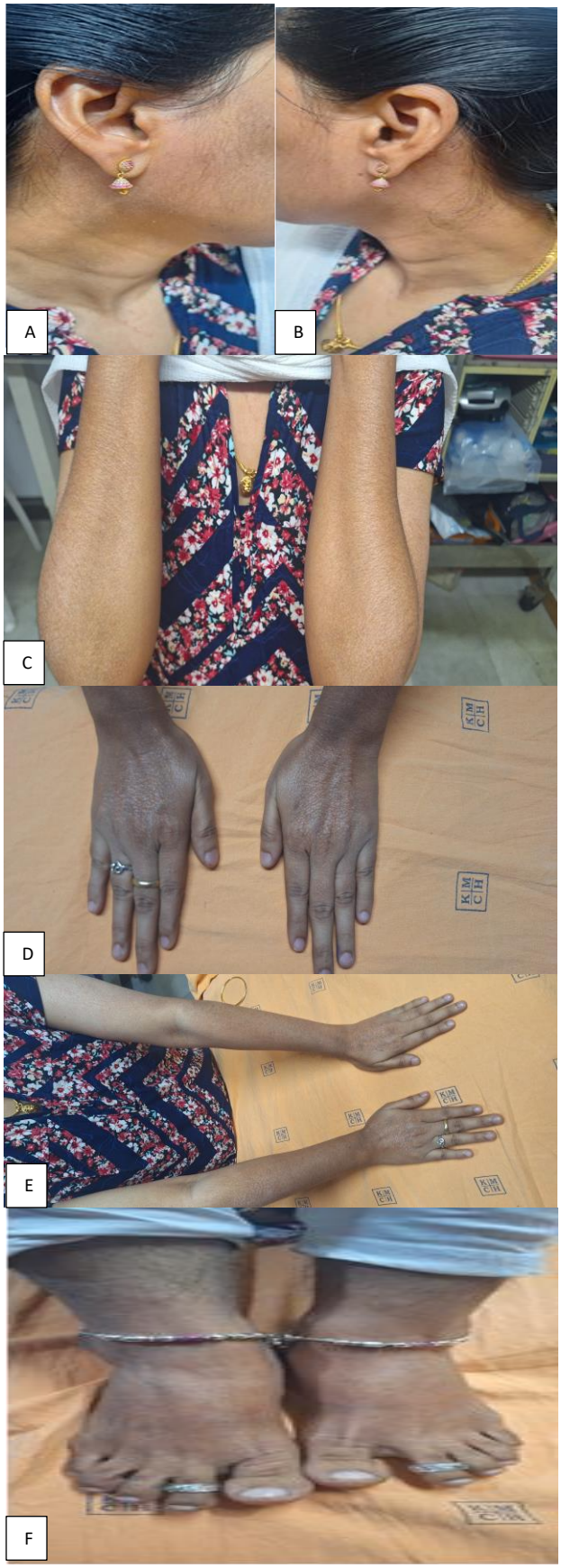

Figure 1 (A-F): Ivory white glistening papules, plaques and patches affecting face, neck, forearms, hands and dorsum of foot.

The plaques were gradually spreading over the years and becoming increasingly pruritic. Few patches showed atrophy. Koebnarisation was observed in few affected areas. Punch skin biopsy revealed findings diagnostic of LSA which included a thinned-out epidermis, loss of rete ridges, focal basal vacuolisation, perivascular mononuclear inflammatory infiltrate in the dermis with homogenisation and hyalinization of collagen (Figure 2 A and B). Immunohistochemical studies were not done. Routine haematological and biochemical investigations were within normal limits. Patient was started with topical clobetasol propionate cream, topical tacrolimus 0.1 % in ointment form with daily hydroxychloroquine (HCQS) 400 mgm initially for 4 weeks, presently on 200mgm and weekly oral methotrexate 7.5 mgm. Patient showed substantial improvement symptomatically and lesions regressed after 6 weeks. She is advised phototherapy (NBUVB) twice a week in the near future. She is under close follow up for recurrence and rarity of malignancy.

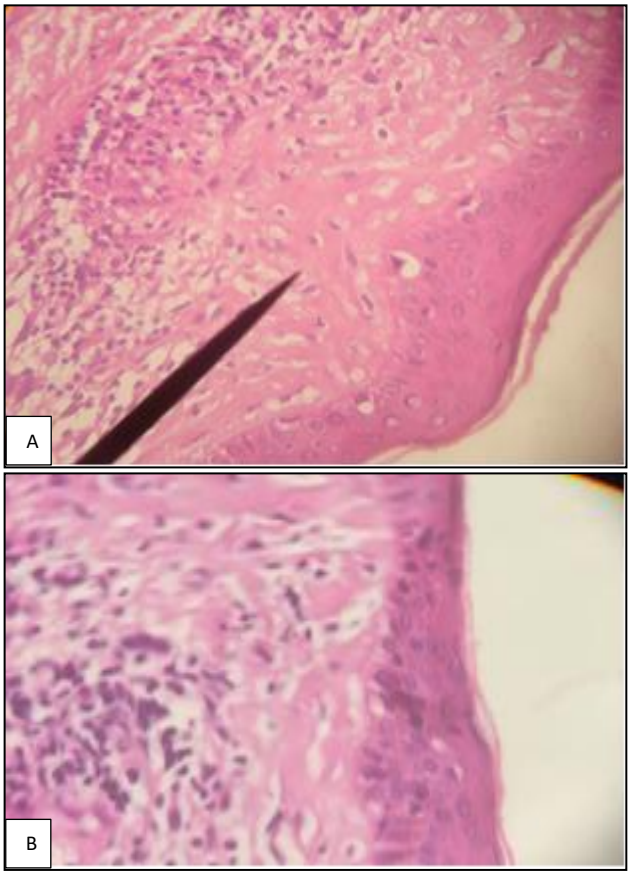

Figure 2 (A and B): H and E stain-epidermal atrophy, focal basal cell vacuolization, mononuclear inflammatory infiltrate in dermis.

DISCUSSION

Lichen sclerosus (LS) originally described by Hallopeau in 1887, is a relatively uncommon chronic auto immune inflammatory cutaneous disorder of unknown etiology. Immunoreactivity to extracellular matrix protein 1 has

been demonstrated in about 74% of the cases.[6] No racial predilection was observed. Familial clustering was observed in a few studies.[7] Occurs at any age with female preponderance. Lichen sclerosus is often associated with autoimmune diseases like hypothyroidism as noted in this case.[8] The prevalence of extragenital LSA may be underestimated as it is often asymptomatic. Extragenital lesions occur in about 15-420% of the patients. In the initial stage, it presents as interfollicular, pearly, polygonal papules, which merge to form atrophic, sclerotic plaques. As the disease progresses, follicular hyperkeratosis and telangiectasias are seen.[9] It occurs on the palms of the hand, soles of the feet, face, scalp and mouth. EGSL is also seen along the lines of Blaschko.[10] The Koebner phenomenon is noticed in LSA. Extragenital lesions are commonly seen in pre-existing scars and damaged areas.[2]

Rate of spontaneous resolution is lower than 25%.[11] Currently, there is no efficacious treatment available. Most patients are initially treated with potent topical corticosteroids. Various treatments including UVA1, NBUVB, PUVA, topical testosterone and estrogen, topical tacrolimus or pimecrolimus, antimalarial agents, penicillin, topical retinoids, and vitamins have been tried with varied efficacy.[12,13] Both UVA1 and NBUVB increase matrix-metalloproteinase levels in the skin explains its effectiveness in sclerosing diseases like LSA affecting epidermis and upper dermis.[14]

CONCLUSION

Our case was unique with extensive, extra genital involvement over face, anterior neck, nape, upper extremities and dorsum of foot in a female who responded partially to topical therapy, oral HCQS/methotrexate. This case is reported for its rarity and wide cutaneous involvement posing therapeutic challenges.

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