Extragenital lichen sclerosus et atrophicus within a skin graft scar

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INTRODUCTION

Lichen sclerosus et atrophicus (LSA) is a rare chronic relapsing skin condition typically arising in postmenopausal women and prepubescent children in the genital area. Only 6% of cases occur in extragenital skin without concomitant anogenital involvement.¹ LSA can exhibit the Koebner phenomenon. Although various types of inciting events have been documented for the development of extragenital lichen sclerosus et atrophicus, we present the first case, to our knowledge, to occur within a skin graft donor site. Significantly, this is the fourth reported case of koebnerizing extragenital LSA without coexisting or previous genital involvement.

CASE PRESENTATION

A 78-year-old woman with a history of diabetes, chronic kidney disease, and stage 1A breast cancer status postlumpectomy presented for evaluation of a new itchy and painful rash of the right posterior thigh and subsequently the bilateral inner thighs for 6 months’ duration. Symptoms initially developed within a skin graft scar on the right posterior thigh, which was taken during early childhood to repair a vaginal wall birth defect called Rokitansky-Mayer-Küster-Hauser syndrome. She denied ever having genital involvement.

Examination found a large, well-circumscribed ivory white atrophic scar with a thin peripheral erythematous scaly plaque on the right posterior thigh (Fig 1, A). The bilateral medial thighs were diffusely atrophic with ill-defined xerotic plaques with a cigarette paper texture (Fig 1, B). A genital examination was unremarkable.

Histologic sections from the right posterior thigh and the right medial thigh showed an atrophic epidermis with compact hyperkeratosis and focal areas of thickened granular layer. In the dermis was mild edema and homogenization of the papillary dermis with a scant perivascular lymphocytic inflammatory infiltrate (Fig 2). Based on the clinical and histopathologic features, extragenital LSA within a skin graft site scar was diagnosed. She was treated with clobetasol 0.05% ointment and calcipotriene 0.005% ointment twice daily. At her 2-months follow-up, the patient had significant symptomatic and cosmetic improvement (Fig 3).

DISCUSSION

Lichen sclerosus et atrophicus (LSA) is a chronic relapsing skin condition characterized by early inflammation followed by chronic scarring and skin atrophy. LSA can occur at any age, but there is a typical bimodal onset in prepubescent children and postmenopausal women. Etiology of LSA remains unclear, but the literature suggests a likely autoimmune phenomenon in a genetically predisposed individual. Previous infections, trauma, and occlusive moist environments are also contributing factors. The lesions of LSA are typically ivory or porcelain-white shiny papules and macules that coalesce to form sclerotic plaques, as seen in our patient.
Although classically seen in the anogenital area (80%-98%), it can be seen in extragenital sites in 15% to 20% of patients. Extragenital sites, although far less common, are most often seen on the buttocks, thighs, breasts, submammary area, and neck. Only 6% of LSA are isolated to extragenital lesions, as seen in our case.

Extragenital LSA can exhibit the isomorphic or Koebner phenomenon in which typical lesions of the disorder occur after inciting events. Few case reports are available supporting this concept, with LSA found within postirradiation sites, cholecystectomy scars, vaccination sites, jellyfish stings, herpes zoster scars, sun burns, sites of friction, and injection sites among others. Although various inciting events have been documented, we believe this patient to be the first documented case of LSA involving a skin graft donor site.

Most cases that show koebnerization have concurrent or a history of genital LSA. To the best of our knowledge, this patient represents the fourth reported case of extragenital LSA, with a known inciting event, developing without concomitant or preexisting anogenital LSA. Four cases of koebnerized extragenital LSA without anogenital involvement have been reported previously: in a vaccination site, an old burn scar, a cholecystectomy scar, and a surgical scar.

The timeframe varies among koebnerizing cases of extragenital LSA. Some appear within months of the trigger, and others develop several years after the trigger. In a case series of 6 of patients with LSA in postirradiation sites, patients had lesions anywhere from 2 to 12 years after radiation. Meffert and Grimwood reported a case in 1994 of LSA developing 50 years after a childhood burn injury. This case was the longest time lapse reported to date until our patient, who had LSA at least 70 years after the skin graft was initially taken.
Unlike anogenital LSA, extragenital LSA does not carry an increased risk of squamous cell carcinoma. Little is known about the long-term complications of extragenital LSA and the risk of repeated koebnerizing lesions developing in areas of trauma. First-line treatment for extragenital LSA consists of high-potency topical corticosteroids and emollients. Although extragenital lesions are not as responsive as genital lesions, our patient has noted significant improvement with these measures in only 2 months time. Second- and third-line treatment options include topical calcineurin inhibitors, systemic retinoids, surgery, phototherapy, photodynamic therapy, and fractional ablative lasers.

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