Case Report

Generalized morphea, lichen sclerosis et atrophicus associated with oral submucosal fibrosis in an adult male

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ABSTRACT

Generalized morphea is a disease characterized by wide-spread sclerosis of the skin. A 39-year-old man presented with history of multiple pigmented and bound-down plaques on the body along with mucosal involvement. Dermatological examination showed multiple indurated and sclerosed plaques with follicular plugging in few of them and gross thickened eroded and glazed tongue. The constellation of these findings with histopathological correlation led us to diagnosis of this spectrum of cutaneous involvement. The coexistence of localized morphea with lichen sclerosis et atrophicus has been reported earlier but existence of these entities with submucosal fibrosis in a same patient is documented here and is the first of its kind.

Key words: Generalized morphea, Lichen sclerosis et atrophicus, Oral submucous fibrosis

INTRODUCTION

Generalized morphea is a disorder of unknown etiology. The onset of disease is insidious with development of plaques resembling those of localized morphea. These plaques are commonly much larger than those seen in localized disease, being many centimetres in diameter. The coexistence of morphea and lichen sclerosis et atrophicus (LSA) in the same patient suggests that these lesions represent a spectrum which may reflect similar etiologic events or closely related pathologic processes in these two diseases.[1] Oral submucous fibrosis (OSM) is a long-lasting disease of the oral mucosa that mainly appears to be caused by chronic exposure to the constituents of areca nut.[2] It is characterized by development of white and indurated changes of the cheeks, lips, palate, and tongue due to fibrosis leading to restriction of mouth opening. The involvement of oral mucosa with submucosal fibrosis is unknown in morphea or LSA. We describe a case with coexistence of these of skin lesions and concomitant mucosal involvement.

CASE REPORT

A 39-year-old man presented with complaints of multiple pigmented and bound-down plaques on the skin of 5-years duration, followed by thickening of tongue with erosions of 3-years duration and ulceration of hands with associated deformities of 9-months duration. The lesions initially started as asymptomatic plaques over arms, followed by similar lesions on trunk, lower extremities, hands, and the face. He also noted development of white-colored patches in few of these lesions and progressive tightening of affected skin. There was thickening of tongue and difficulty in protrusion of tongue with few erosions. There was gross ulceration over the dorsa of hands and contracture deformities of all the fingers that prevented him to perform his normal daily activities. He did not complain of any bluish discoloration of fingers, dyspnea, dysphagia, weight loss, or any other systemic complaint. He was a chronic alcoholic and a smoker for last 18 years. There was no history of any betel nut chewing at any time, though he attributed the changes of the oral cavity to an amalgam dental filling that was removed within a few months of start of the disease. Despite removal, the process of thickening of tongue continued. There was no similar complaint in any other family member.

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He was moderately built and nourished and general physical as well as systemic examination did not reveal any findings. On cutaneous examination, there were multiple, well-defined, large skin colored to hyperpigmented, indurated, and sclerosed plaques distributed all over the body [Figure 1]. On the face, there was a single large plaque involving the left eye with subsequent ectropion [Figure 2]. The facial expressions were preserved. Another large plaque extended from nape of the neck to occipital region leading to scarring alopecia [Figure 3]. There was involvement of pinnae with similar changes. In addition, few plaques exhibited hyperpigmented margins, adherent dark scales, follicular plugging, and depigmentation [Figure 4]. The dorsal surface of both hands showed ulceration on the medial aspect with dirty unhealthy crusted granulation tissue [Figure 5]. The skin of hands was adherent and there

Figure 1: Multiple sclerosed plaques on the body

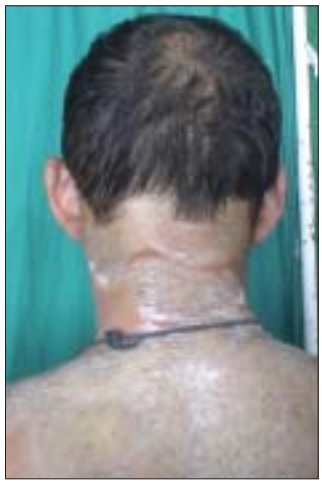

Figure 3: Morphea plaque on nape of neck

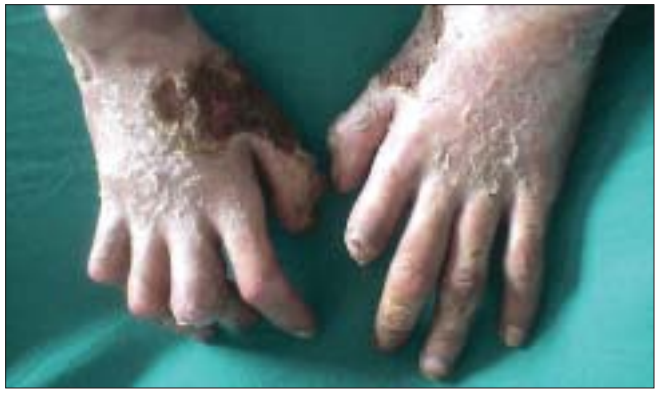

Figure 5: Morphea plaques on hands showing contractures and ulceration

Figure 2: Morphea plaque on left eye with subsequent ectropion

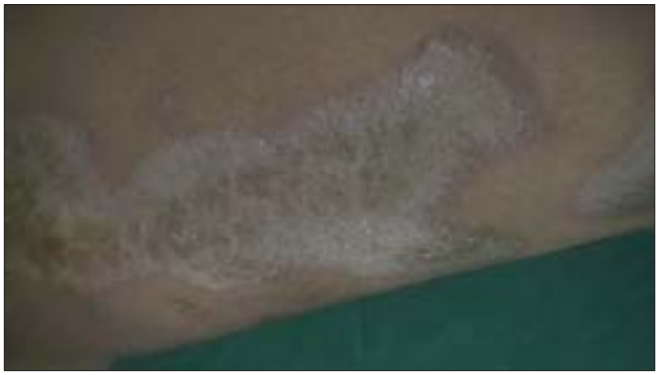

Figure 4: Lesion showing follicular plugging, scales, and depigmentation

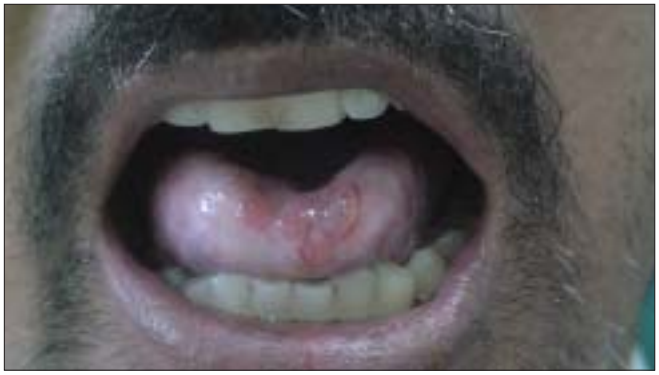

Figure 6: Thickened and glazed tongue

was fixed flexion deformities of all the fingers. The nails were thickened and yellowish. There were no ragged cuticles or digital pitted scars.

Oral cavity examination showed thickened and glazed tongue with erosions at few places [Figure 6]. There was difficulty in opening of the mouth. He was unable to touch the hard palate with tip of his tongue as well as there was inability to protrude the tongue. There was fibrosis of both soft palate and tongue. There were few similar depigmented lesions over shaft and glans penis. There was no difficulty in retracting the prepuce but the shaft of penis was turned unidirectionally.

Investigations revealed Hb of 13 gm/dL, TLC of 6700/mm^3 with normal differential and platelet count. The urinalysis and serum biochemistry were normal. Tests for rheumatoid factor, antinuclear antibody, AntiHCV, and HbsAg were negative. Spirometry and barium swallow were normal. Abdominal ultrasound showed fatty liver changes. X-ray of wrist and metacarpophalangeal and interphalangeal joints showed periarticular erosions, osteopenia, and flexion deformity. Skin biopsy was done from four regions: (1) biopsy from forearm showed thinned out epidermis with flattened rete ridges. Dermis had mild to moderate perivascular lymphoplasmacytic cell infiltrate; in addition, dermis was completely collagenized with hyaline deposition describing changes of morphea; (2) biopsy from lesion with follicular plugging showed hyperkeratotic epidermis with flattened rete ridges. Dermis was densely sclerosed with mild perivascular inflammatory infiltrate. The adenexal structures were normally placed in the deeper dermis. This was consistent with lichen sclerosis et atrophicus; (3) biopsy from tongue showed no epidermal changes and mild chronic inflammatory infiltrate in subepithelium, that too in addition was heavily collagenized describing changes of submucosal fibrosis; and (4) ulcer edge biopsy was composed of sheets of acute inflammatory cells, fibrin, plasma, few dermatophytes, and bacterial colonies. The infiltrate was concentrated more around blood vessels with endothelial swelling but there were no changes of fibrinoid necrosis or malignancy.

After detailed history, examination, and investigations, final diagnosis of generalized morphea with LSA and submucosal fibrosis was made and the patient was administered dexamethasone pulse therapy in a dose of 140 mg daily for three days along with antibiotic course and supportive treatment. On his first follow-up visit, there was mild subjective improvement in his tightness of the skin and he is on further treatment with pulse therapy.

DISCUSSION

Scleroderma is a group of chronic autoimmune diseases with skin thickening as the hallmark of the disease. Traditionally, the term "scleroderma" encompasses two groups: localized scleroderma and systemic sclerosis. Generalized morphea and LSA are now classified under localized sclerosis by Peterson *et al*, due to histological similarities between two and the frequent occurrence of LSA with the other morphea subtypes.[3] Generalized scleroderma is characterized by wide-spread sclerosis of the skin in which plaques become confluent and affect more than two body sites. Sometimes, it may develop as an extension of localized morphea or be associated with lesions of LSA.[4] Both these disorders have female preponderance and mainly affect children and young adults.

The etiology of these diseases is still unclear and their relationship is obscure. Vascular damage and an increased level of adhesion molecules and cytokines are seen in early lesions of morphea. There is increased expression of type 1 collagen in morphea. Collagen synthesis is increased in both LSA and morphea, with increased noncollagenous protein glycosaminoglycan in morphea.[5] Coexistence of these entities have been described initially by Uitto *et al*, in 10 patients in the year 1980. Thereafter, there are only sporadic case reports in the literature.[6,7]

OSF is a chronic debilitating and a premalignant condition of the oral cavity. The pathogenesis of the disease is not well established. Epidemiological evidences strongly indicate the association of the betel quid (BQ) habit and OSF. Various findings indicate the disease to be a consequence of disturbances in the homeostatic equilibrium between synthesis and degradation of extracellular matrix (ECM), wherein collagen forms a major component. Transforming growth factor-beta (TGF-beta) is a potent stimulator of production and deposition of the ECM.[8] Various studies have taken into account the role of chewing or smoking tobacco in development of OSF. It was inferred that neither chewing nor smoking tobacco increased the risk of developing OSF.[9] On the other hand, it was also found that it was frequency of chewing and not the total duration that was directly

correlated to OSF.[10] In addition to tobacco use, intake of specific/unknown nutrients may have a role in the development of oral precancerous lesions.[11]

Our patient had extensive fibrosis limited to skin and mucosal surfaces. Over a period of five years the disease progressed rapidly without any systemic involvement supporting the clinical impression that morphea is a benign, self-limited disease with progression to systemic form being a rare occurrence. However, morphea can be very disabling and in a study by Peterson *et al*, conducted in Olmsted County, disability was observed in 11% of individuals affected with morphea.[12] The extension of the lesions of LSA over the dorsal and palmar aspect of the hands was associated with formation of severe contractures and ulceration leading to disability. This could be due to acute inflammatory process causing fibrosis of the affected site. Various other causes of ulcers such as vasculitis and malignancy were excluded histopathologically. Radiological changes like periarticular erosions, osteopenia, and flexion deformities have been reported with localized or generalized morphea as seen in our patient.

Our patient was a chronic smoker and an alcoholic. The development of OSM in him may be an independent occurrence although the basic pathology in all these diseases is related to excess collagen deposition, but the involvement of oral mucosa has not been reported with either generalized morphea or LSA. There was no history of BQ chewing at any time but there was history of dental amalgam filling prior to onset of disease in the oral cavity. But even subsequent to its removal, the changes of OSM continued and there are no reports to suggest dental filling as its cause. On the contrary, there are a few studies in literature to indicate intake of alcohol or tobacco smoking or even other unknown dietary factors as the inciting factors. A similar case of localized morphea with OSM has been described earlier but the changes occurred secondary to BQ habit.[13]

This patient has many unusual presentations that are difficult to explain despite good history and examination and serves worth reporting. Therefore to conclude, morphea and LSA may be associated in a same patient and even transitional forms may occur; LSA is now categorized as subepidermal morphea; and OSM may have other causes apart from BQ chewing.

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