INTRODUCTION

Parry–Romberg syndrome (PRS) is a rare neurocutaneous syndrome characterized by progressive shrinkage and degeneration of the tissues beneath the skin, usually on one side of the face also called as progressive hemifacial atrophy. It is an autoimmune disease, specifically a variant of localized scleroderma. The disorder has a female to male predilection of 3:2. It was described first by Parry in 1825, then Romberg in 1846 and Eulenberg in 1871.

Left side of the face is involved in more than 85% of cases. It commonly affects the eyes and is sometimes (15%) associated with neurological disorders such as trigeminal neuralgia, facial paresthesia, headache, and focal epilepsy. Concomitant occurrence of discoid lupus erythematosus (DLE) and morphea in the same skin lesion is exceptional, defined as overlap syndrome with two or more different connective tissue disease concurrently or consecutively. A 32-year-old female developed DLE on a long-standing lesion of scleroderma over left temporal area with characteristics histopathological changes. She was treated with oral antimalarials and steroids which halted the progress of the disease.

Key words: Discoid lupus erythematosus, linear, Parry–Romberg syndrome, scalp

CASE REPORT

A 32-year-old female working in the farm presented with a complaint of tightening and thickening of the skin over left cheek, forehead and scalp with loss of hairs and throbbing pain since almost 8 years. She had h/o elevated scaly lesion over the left side of the scalp which were itchy since 5 years. She also complained of edema and pain over the right side of the face since 15 days. No history of cough, cold, dyspnea, chest pain, palpitations, nausea, vomiting, abdominal pain or joint pain was present. No history of any seizure-like symptoms or cognitive impairment was given. Family history was unremarkable. Cutaneous examination showed well-defined linear atrophic plaque over left forehead, maxillary region that is, left cheek and frontal area of the scalp [Figure 1]. Cicatricial alopecia with 2-3 hyperpigmented plaques and mild scaling over left temporal region [Figure 2]. Indurated and edematous plaque over right cheek and forehead [Figure 1]. Buccal...
mucosa showed hyperpigmentation over both sides. The neurological examination was normal. Complete blood count, erythrocyte sedimentation rate, liver function test, renal function test and coagulation tests were normal. Clinically patient was labeled as Parry–Romberg disease and a scalp biopsy was taken from the scalp lesion to confirm the diagnosis. The section showed in part changes of late sclerotic stage of morphea characterized by presence of thickened, closely packed and hypocellular collagen bundles in the reticular dermis. The papillary dermis also shows the presence of homogeneous collagen [Figure 3].

The other area shows the hyperkeratosis with follicular plugging [Figure 4]. Thinning of stratum malphighii, focal hydropic degeneration of basal cells and squamatization of basilar keratinocytes [Figure 5]. Stroma showed lymphocytic infiltrate around hair follicles and in interstitial pattern. Changes showed development of DLE in a case of morphea.

Patient was not ready for various immunological investigation and computed tomography (CT) scan. Direct immunofluorescence microscopy was not done due to lack of facility. Patient was given chloroquine 200 mg daily with oral steroids prednisolone 20 mg daily in a tapering dose for 15 days. Inflammatory lesions responded with subsidence of erythema and edema. Topically betamethasone valerate (0.05%) was started, and gradually patient was shifted to tacrolimus 0.1% ointment. Patient is better with no further progress of the disease.

**DISCUSSION**

Scleroderma “en coup de sabre” is a linear localized form of morphea, affecting the frontoparietal scalp and forehead in stripe-like sclerotic plaques. The skin appears hard, hyperpigmented, shiny and with alopecia. Progressive atrophy is generally localized to a small area of the skin corresponding to the temporal or buccinator muscles. Our patient presented with atrophy of the left side of the face with inflamed plaque on the right side. The disease has been reported to affect both sides of the face in 5-10% of the cases. Neurologic symptoms include epilepsy, migraine, facial pain and brain lesions on CT and magnetic resonance imaging as well as heterochromia and enophthalmous.[8] Migraine and facial pain (trigeminal neuralgia) are the commonest neurological conditions with a frequency reaching 52%. Epilepsy has been described in 10% of the patients, sometimes associated with brain abnormalities ipsilaterally to the skin lesions.[8]

Our patient had throbbing pain which was episodic in nature without any other brain morphology. There are reports associating these neurological symptoms with peripheral sympathetic nervous system or trigeminal nerve abnormalities, and others support the presence of vascular dysgenesis.[7] The most recent and reliable theory is that of a genetic alteration in the first stage of embryogenesis of the central nervous system. Other causes include viral or bacterial infections, loss of cervical sympathetic nerve, following peripheral neuritis of the trigeminal nerve, trauma, endocrine disturbances, and hereditary.[4]

Immunosuppressive drugs such as methotrexate, corticosteroids, cyclophosphamide, and azathioprine are used. Surgical modality includes autologous fat transfer or fat grafts, temporal fascia flaps, cartilage grafts, bone grafts, orthognathic surgery, and bone distraction.[9]

Overlap syndrome are defined as disorders that satisfy diagnostic criteria of two or more different connective tissue disease concurrently or consecutively. Concomitant occurrence of chronic cutaneous lupus erythematosus and morphea in the same skin lesion is exceptional. Our case where patient first developed linear morphea, encoupe de sabre with involvement of scalp and forehead a relatively rare entity, consecutively developed lesions of DLE. Presence of thickened, closely packed and hypocellular collagen bundles in the reticular dermis were suggestive of localized scleroderma while hyperkeratosis with follicular plugging thinning of stratum malphighii, focal hydropic degeneration of basal cells and squamatization of basilar keratinocytes showed development of DLE on lesion of morphea. Such association has been reported by Khelifa et al.[7] also over face and scalp. Umbert and Winkelmann[6] Marzano et al.,[9] and Julià et al.[11] also reported such association but lesions were on the arms rather than over face. All cases were treated with oral antimarialar and topical steroids with partial improvement. The lesions were along the line of Blaschko’s in all cases. Blaschko’s lines are thought to reflect cell migration and clonal expansion during embryogenesis of the skin. Genetic mosaicism and microchimerism causes skin lesions in a linear distribution in scleroderma. DLE pathophysiology favors keratinocyte apoptosis as the initiating event. Seitz et al.[8] suggested linear variant of DLE, which can be explained by presence of genetically variant keratinocyte lacking protective factor ultraviolet induced damage, showing aberrant cytokine expression.

The occurrence of DLE in patient with PRS has been rarely reported. Our patient developed. DLE on a long-standing lesion of scleroderma over left temporal area. Knowledge

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of this peculiar association is important from the point of view of management and prognosis.

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