

Morphea and vitiligo-A very uncommon association

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Sir,

Morphea represents a localized form of scleroderma where there is predominant skin involvement, with occasional involvement of subjacent muscles. However, it usually spares the internal organs [1]. Morphea has been associated with several autoimmune diseases like mixed connective tissue disease, dermatomyositis, pemphigus, myasthenia gravis, bullous pemphigoid, systemic sclerosis, Hashimoto's thyroiditis, etc [2,3]. The association of morphea with vitiligo has been rarely reported. In this article we report a 29 year female with concomitant vitiligo and plaque type morphea over her lower back.

CASE REPORT

A 29 year female reported to our dermatology department with a chief complaint of brownish hyperpigmentation, thickening and hardening of the skin on the lower back on the left side for the last three months. There is history of mild pruritis. There is no history of any discoloration in the digits on exposure to the cold. There is no history of sour eructation's, epigastric discomfort or constipation. There are no similar complaints of hardening of skin on hands and feet. She denied any history of preceding trauma or application of some topical medications prior to the complaints. There is no such history in her family members. The patient gives history of vitiligo for the last five years localized to scalp with associated whitening of hair. On examination, there was an ill-defined hyperpigmented, indurated, shiny plaque around the size of 8cm × 4cm on the lower back towards the left side extending up to the midline (Fig. 1). The plaque

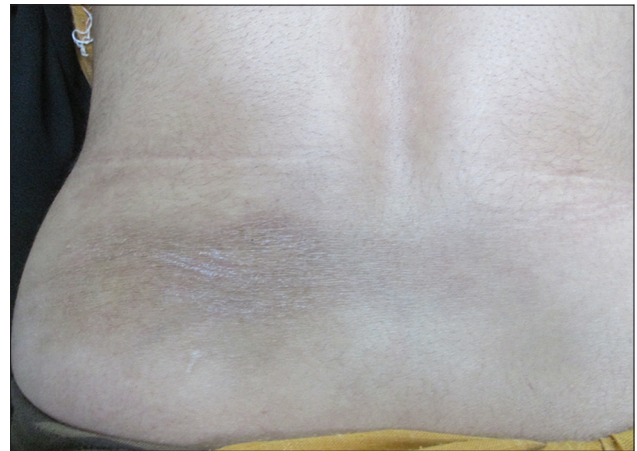


Figure 1: Ill-defined brownish hyperpigmented indurated plaque over lower back.

showed loss of appendages. There was no associated digital pallor or cyanosis. Nail fold capillaroscopy was unremarkable. The scalp on the occipital region of the patient revealed a hypo pigmented patch about the size of 5cm × 4cm with associated leucotrichia (Figs 2 and 3). A 5mm punch skin biopsy on the back was sent for histopathological examination which showed atrophy of the epidermis, perivascular lymphocytic and plasma cell infiltrates in the dermis and subcutaneous tissue. There were thickened and closely packed bundles of collagen. The adnexal structures were scanty. The typical clinical findings and further supported by histopathology confirmed the diagnosis of morphea in our patient. Her routine laboratory tests were unremarkable. Her anti-nuclear antibody was within normal limits. Her thyroid function tests were unremarkable. She was prescribed topical tacrolimus 0.1% ointment to be applied twice daily. After three months of treatment, her skin pigmentation and induration improved.

How to cite this article: Arif T, Hassan I, Nisa N. Morphea and vitiligo-A very uncommon association. Our Dermatol Online. 2015;6(2):232-234.

Submission: 23.10.2014; **Acceptance:** 25.01.2015

DOI: 10.7241/ourd.20152.62



Figure 2: Vitiliginous patch with associated leucotrichia on the occipital region.



Figure 3: Close view of the vitiliginous patch and the associated leucotrichia.

DISCUSSION

Scleroderma comprises a spectrum of disorders which is characterized by thickening and/or hardening of the skin eventually leading to the fibrosis of the tissues. Broadly, it has been divided into the systemic and localized forms. The localized type of the scleroderma is called as morphea. In morphea, there is predominant skin involvement, with occasionally involving the subjacent muscles. The internal organs are usually spared in morphea in contrast to the systemic sclerosis [1]. However, extra cutaneous features have been reported from childhood morphea cases [4]. Paterson, et al has classified morphea into morphological types viz., plaque, linear, generalized, deep and bullous [5,6]. Our patient had plaque type of morphea.

Morphea is considered to be an immune-mediated disease which has been suggested by various studies. This

is supported by increased levels of circulating cytokines in the patients of morphea including interleukin-2 (IL-2) receptor, soluble CD4 and CD8, CD23, CD30, IL-6 receptor, IL-13 and toxic necrosis factor (TNF), soluble vascular cell adhesion molecule-1 (VCAM-1) and E-selectin, antiendothelial cell antibodies, etc. Various organ-specific auto antibodies have been demonstrated in the serum of these patients and their relatives. Another evidence is the association of morphea with other autoimmune diseases. Morphea has been seen in association with carpal tunnel syndrome, nephritis, dermatomyositis, pemphigus, primary biliary cirrhosis and myasthenia gravis [2]. The association of morphea with vitiligo is an uncommon one and hence inspired the authors to report the same.

Bonilla-Abadia (2012), et al have described a peculiar case of morphea as a part of multiple autoimmune syndrome (MAS) in which a 53 year-old female patient with plaque type morphea over legs was associated with vitiligo, pneumonitis, autoimmune thrombocytopenic purpura and central nervous system vasculitis [7]. When three or more well-defined autoimmune diseases are present in a single patient, the condition is known as MAS [8]. However, in our case, there was no systemic involvement other than the associated vitiligo. Moreover, her anti-nuclear antibody and thyroid function tests were unremarkable. However, the antibody profile specific for other organs was not done.

Generally, the prognosis of morphea is considered to be good. Rarely, it has been reported to evolve into the systemic sclerosis. The disease activity may last for three to four months in most cases [9]. However, regular follow up is warranted to screen for the development of other concomitant systemic autoimmune disorders like MAS.

The treatment of morphea has been updated. Various treatment options include topical tacrolimus, Imiquimod, phototherapy, calcipotriol in combination with betamethasone dipropionate, cyclosporine, D-penicillamine, photopheresis, etc [10]. Our patient was already receiving treatment for vitiligo. For morphea, she was prescribed topical tacrolimus 0.1% ointment applied twice daily. After three months of treatment, skin pigmentation and induration of the plaque on the back improved.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

Written informed consent was obtained from the patient for publication of this article and any accompanying images.

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Source of Support: Nil, **Conflict of Interest:** None declared.