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Concomitant Segmental Vitiligo and Segmental Morphea in an 8-year-old boy

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Sir,

Segmental vitiligo (SV) and segmental morphea are unilateral cutaneous disorders, with an autoimmune pathogenesis. The association of the two generalized forms has been largely reported, while the combination of the two segmental variants is uncommon.

An 8-year-old boy referred to our hospital for vitiligo and morphea lesions at his trunk. As referred by his parents, the SV lesion was present at birth and was stable. The morphea lesion appeared 2 years before, initially with peripheral lilac ring and previously treated with topical steroids. On physical examination, confluent achromic lesions, with jagged border, were present at the left scapular region, with dermatomal distribution, while a homolateral sclerotic patch was present at the left lumbar region [Figure 1]. A cutaneous biopsy of the sclerotic lesion was performed, histologically confirming the diagnosis of morphea: epidermal atrophy and dermal thickening and collagen hyalinization, with lymphocytic infiltration, were present [Figure 2]. Routine laboratory exams were in normal range; thyroid function tests and celiac disease antibodies tests were negative. The patient is under topical treatment with vitamin E cream, with partial improvement of the sclerotic lesion.

Our case is of interest because of its rare association of these two autoimmune conditions. A study on 1098 vitiligo individuals has shown that 20% of patients presented comorbid autoimmune diseases. Between them, 0.2% of individuals had linear morphea.^[1]

Many reports describe cases of concomitant generalized vitiligo and morphea. Instead, the association between SV and segmental morphea had been rarely reported^[2-4] and is summarized in Table 1.

The explanation of this association is still unclear.

SV is a localized vitiligo with typical distribution along dermatomes or Blaschko's lines. Both SV and classic vitiligo are autoimmune conditions with

T-cell-mediated degeneration of melanocytes. Recent studies indicate that SV is due to an autoimmune attack against a mosaic region.^[5] Also, morphea is considered an autoimmune disorder, with collagen density and sclerosis. Neurological, traumatic and infectious agents have been suggested as trigger factors of the autoimmune attack.

Differential diagnosis of both conditions is summarized in Table 2. In our patient, the vitiligo lesion was clearly identifiable, since its achromic macular aspect with jagged border, while the morphea lesion could be confused with other sclerotic conditions, including lichen sclerosus, radiodermatitis, and intralesional

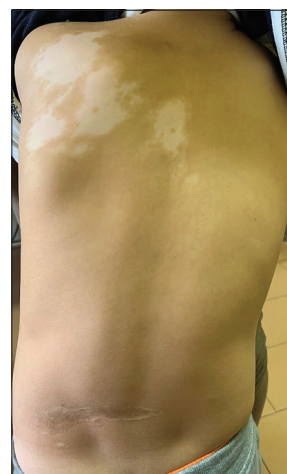


Figure 1: Vitiligo of the left scapular region and sclerotic patch of the left lumbar region

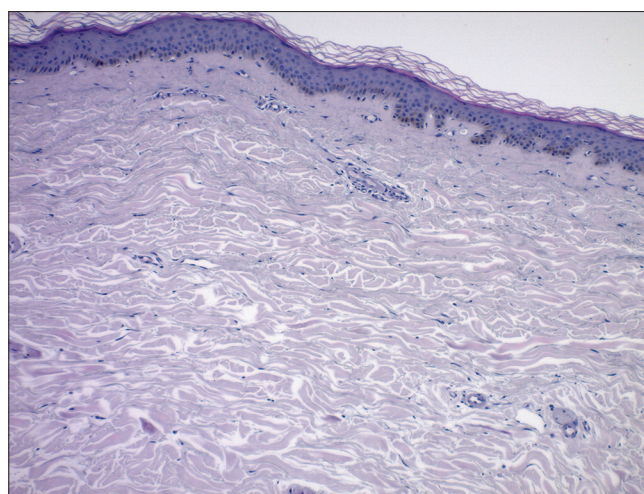


Figure 2: Hematoxylin-eosin stain 10×; epidermal atrophy; thickening and collagen hyalinization in the deep dermis, with mild inflammatory infiltration of lymphocytes, which are distributed around blood vessels. Atrophy of adnexal structures, particularly the pilosebaceous units, with hypertrophic hair erector muscle. The vascular changes are thickening of the walls of small blood vessels and narrowing of their lumen

Table 1: Reports of concomitant segmental vitiligo and segmental morphea

Authors	Sex	Age (years)	Vitiligo	Vitiligo region	Morphea	Other signs
Ubaldo, 2019	Female	22	SV	Left periorbital region with eyebrows poliosis	En coup de sabre (left forehead)	Cleft lip
Yadav, 2014	Female	18	SV	Left upper limb-left chest	Segmental morphea of right upper limb and right abdomen	
Janowska, 2013	Male	46	SV	Left upper limb-left trunk Right chin and neck	Parry-Romberg syndrome (left cheek, jaw, eye, ear)	
Bowen, 2010	n.a.	n.a.	SV	n.a.	En coup de sabre	
Bonifati, 2006	Male	21	SV	Left trunk	Linear dermatomeric morphea of the left upper limb	
Creus, 1994	Female	11	SV	Left neck and trunk	Parry-Romberg syndrome	

SV: Segmental vitiligo; n.a.: not available

Table 2: Main differential diagnosis of vitiligo and morphea

Vitiligo	Morphea
Piebaldism	Lichen sclerosus
Tuberous sclerosis	Radiodermatitis
Postinflammatory leukoderma	Vitiligo
Posttraumatic leukoderma (e.g., burns)	Sclerodermiform basal cell carcinoma
Pityriasis alba	Idiopathic or secondary lipoatrophy
Pityriasis versicolor	Necrobiosis lipoidica
Ito's hypomelanosis	Chronic graft-versus-host disease
Mycosis fungoides	Lipodermatosclerosis
Melanoma-associated leukoderma	Porphyria

steroid-induced lipoatrophy. However, our patient did not report anamnestic data possibly related with the lesion, such as previous radiotherapy or injective treatments. Additionally, the histological exam ruled out other disorders.

In consideration of previous reports, and of the similar immunological pathogenesis, a common immunological pathophysiology is hypothesized. Concerning the segmental distribution, the currently accepted theory suggests that both diseases result from genetic mosaicism, with mutations in some ectodermal cells during the embryonic life.^[2,5] The disorder develops when cells resulting from mosaicism are exposed to a still unclear causative agent.^[2,5]

We report this case to add to the knowledge of the association of these two segmental disorders, both representing a condition of genetic mosaicism and suggesting common immunological pathogenic factors.

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Conflicts of interest

There are no conflicts of interest.

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