Co-occurrence of linear scleroderma en coup de sabre and segmental vitiligo on the face

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DESCRIPTION

A woman in her 30s came with complaints of white-coloured lesions on the left side of her face and multiple brown-coloured lesions on the face, neck and back for 1 year. Lesions were insidious in onset and gradually progressive and not associated with itching or pain. There was no history of trauma, family history of similar lesions or topical application of any products on the area.

On examination, multiple hypopigmented macules and patches were seen on the glabella, cheek and upper lip region on the left side of the face, suggestive of segmental vitiligo (SV) (figure 1). Lesions were seen under Wood's lamp as chalky white fluorescence (figure 2).

Hyperpigmented patch with linear atrophy on palpation was seen on the left side of the forehead and hyperpigmented patches with interspersed hypopigmented macules with atrophic and shiny skin seen on the left side of the neck, left scapula and infrascapular region, suggestive of linear scleroderma (figure 1).

Vitiligo of the segmental type affects a particular segment of the body characteristically demarcated at the midline. Autoimmune diseases are less commonly associated with SV, compared with vitiligo vulgaris, but certain skin conditions which follow a linear pattern, such as linear morphea, have often been reported to be associated with SV. Most case reports of this association have a remarkable similarity. Many linear genetic conditions such as lichen striatus, blaschkitis, linear scleroderma and linear lupus erythematosus follow a certain pattern along the Blaschko's lines due to genetic mosaicism, where an immune reaction towards a mutated cell is suspected. All these diseases usually start at a young age and have a temporary disease activity, followed by spontaneous resolution, with response to treatment with methotrexate effective in most cases with few recurrences.[1]

Despite some case reports of the coexistence of SV and linear scleroderma, this phenomenon is still rare.[2 3] It is remarkable that two mosaic disorders affecting different cell lines and embryogenic origins have a tendency to co-occur on the same body area. Various treatments such as oral corticosteroids, methotrexate, ciclosporin, Janus kinase inhibitors, supplements and phototherapy have been tried in the treatment of vitiligo.[4] Multiple treatment options such as methotrexate and intravenous/oral steroids have been used, which led to

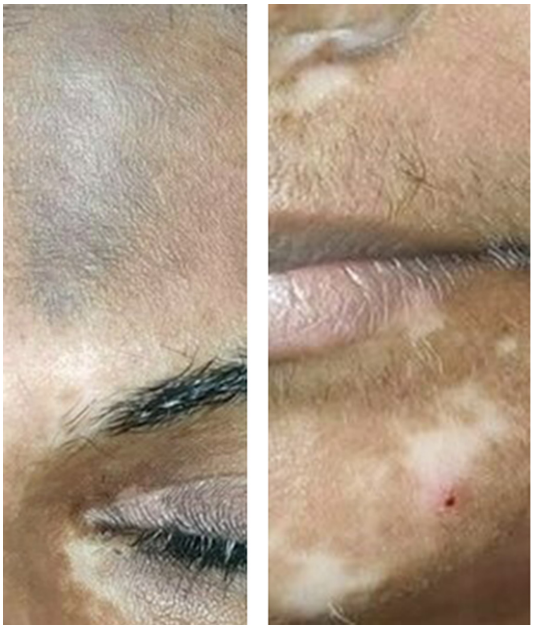

Figure 1 Linear scleroderma en coup de sabre and segmental vitiligo on the left side of the face.

Figure 2 Lesions of vitiligo have a chalky white fluorescence on Wood's lamp examination.

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Patient's perspective

The sudden appearance of lesions over the face has affected my self esteem and reduced my quality of life. The nature of both the diseases, the chronicity of the conditions has added to my emotional trauma.

Learning points

- Although the exact pathogenesis of both conditions is still not clear, an autoimmune mechanism may explain the co-occurrence.
- Cutaneous mosaicism could explain the distribution of lesions in segmental vitiligo and scleroderma en coup de sabre.
- The simultaneous appearance of segmental vitiligo and scleroderma en coup de sabre on the same side of the face is a rare occurrence.

rapid control of disease progression and significant improvement.[5] Although there are many reports on the coexistence of linear scleroderma and SV at separate sites in the same patient, it is rare that these two conditions occurred at the same time and on the same side of the face in a patient.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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