

## Case Report

# Segmental lentiginosis with ipsilateral segmental vitiligo – a case report

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**Abstract** We report a patient with unilateral segmental lentiginosis of the right side of face and ipsilateral segmental vitiligo of right axilla. The patient also had a larger hyperpigmented macule on the same side of the face. This could have been a café au lait macule. We are reporting this case for its rarity.

**Key words**

Segmental lentiginosis, partial unilateral lentiginosis, segmental vitiligo.

### Introduction

Lentigines are small, circumscribed brown to dark brown to black, variegated to uniform coloured macules. They may be found as isolated macules in sun exposed areas or as multiple lesions on any cutaneous surface including palms and soles.<sup>1</sup>

A circumscribed grouping of lentigenes on one side of the body is called segmental lentiginosis.<sup>2</sup> The grouping may be localized or involve a larger area of the body.<sup>2</sup> This disorder is also known as zosteriform lentiginous naevus, partial unilateral lentiginosis (PUL) or lentiginous mosaicism.

Segmental vitiligo is characterized by unilateral depigmented macules in a dermatomal or quasidermatomal distribution.<sup>3</sup> Both disorders more commonly appear in childhood.<sup>4</sup>

We report a case of concomitant segmental

lentiginosis and segmental vitiligo on the same side of the body. We speculate that this paired manifestation supports the neurogenic hypothesis of the aetiology of vitiligo.

### Case report

A 10-year-old girl had hyperpigmented skin lesions present since birth on the right side of her face and progressive depigmented skin lesions in the right axilla since seven years of age.

On examination, the right side of her face had a cluster of brownish macules varying in size from 0.1 to 1mm and a single oval circumscribed brownish black macule measuring 2x1cm in size. These macules were present on a background of normal skin. There were depigmented macules of varying sizes with repigmenting margins on the right side of chest, right axilla and inner aspect of right arm (**Figure 1**).

There were no extracutaneous manifestations. The patient was started on topical fluticasone propionate 0.05% ointment for the vitiligo and asked to come for regular follow up.

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**Figure 1** Multiple depigmented patches in right axilla and café-au-lait-macule on right cheek.

## Discussion

The grouping of hyper-pigmented macules in this case made us to consider a differential diagnosis of segmental lentiginosis and speckled lentiginous nevus (nevus spilus). As the pigmented macules were present on a background of normal skin, a diagnosis of segmental lentiginosis was made. Segmental lentiginosis or PUL is usually diagnosed in young people and can even be present at birth.<sup>1</sup>

Multiple hypotheses exist concerning the aetiology of PUL with as yet no clear-cut answer. Some cases of PUL may be a form of segmental neurofibromatosis.<sup>1</sup>

The single light brown larger macule on the face could be lentigo lying outside the cluster of lentigines or a café au lait macule. Café au lait macules have been reported in association with PUL.<sup>1,2,5</sup>

A clinical diagnosis of segmental vitiligo was made based on the presence of depigmented macules with re-pigmenting margins in a segmental distribution.

Segmental vitiligo is a special type of vitiligo that has a stable course. It is not usually

associated with other autoimmune disorders.<sup>3,4</sup> It has been classified as type B in Koga's classification.<sup>6</sup>

The different hypotheses regarding the aetiology of vitiligo are autoimmune, neurogenic and self-destruct hypotheses. The dermatomal distribution and the presence of abnormalities of the terminal portions of peripheral nerves<sup>7</sup> in segmental vitiligo support the neurogenic hypotheses.

There are previous reports of the simultaneous occurrence of PUL and nevus depigmentosus.<sup>2,8,9</sup> To our knowledge the coexistence of PUL and segmental vitiligo has not been reported.

The simultaneous occurrence of PUL and segmental vitiligo in our patient may not be a mere coincidence. PUL has been postulated to a form of neurofibromatosis.<sup>1</sup> We feel that the coexistence of PUL and segmental vitiligo on the same side of the body further supports the neurogenic hypothesis of vitiligo.

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