What’s the diagnosis?

An otherwise healthy 54-year-old black man presented with a 10-year history of spotty pigmentary loss in a band on the left side of the abdomen, flank, and back. He denied a history of rash or inflammation in the area and had not experienced confluent depigmentation. He reported that initially he had only a few “white dots,” and over the next 5 to 7 years, he developed more of them confined within the same area. On presentation, he stated new areas of depigmentation had not developed in several years. The band was completely asymptomatic and had not been treated with any prescription or over-the-counter medications.

On examination he had multiple 2- to 3-mm confetti-like depigmented macules that seemed to be centered around follicles in a band with blaschkoid distribution extending across the left side of the abdomen, flank, and back. The band did not cross the midline and similar lesions were not present elsewhere.

A punch biopsy of one of the depigmented macules revealed a markedly diminished number of melanocytes along the junction as well as a decrease in melanin, which was confirmed by Melan-A and Fontana stains, respectively.
The Diagnosis: Blaschkoid Punctate Vitiligo

Based on the patient’s clinical appearance as well as the histologic findings, the diagnosis of vitiligo was made. Although vitiligo is certainly not uncommon and punctate vitiligo is a known clinical presentation, punctate vitiliginous depigmentation conforming to lines of Blaschko is unique. Follicular repigmentation in a patch of vitiligo potentially could lead to this “spotty” appearance, but our patient maintained that the band was never confluent and that small macules arose within normally pigmented skin. The patient’s adult age at onset makes this case even more unusual.

Follicular repigmentation in vitiligo is fairly well understood, as the perifollicular pigment is formed by upward migration of activated melanoblasts in the outer root sheath. Follicular depigmentation as well as selective or initial loss of melanocytes around hair follicles in early vitiligo has not been described. It is unclear if the seemingly folliculocentric nature of the patient’s vitiliginous macules was a false observation, coincidental, or actually related to selective melanocyte loss around follicles.

Blaschkoid distribution has been described in numerous skin disorders and is known to be based on genetic mosaicism. Most of these disorders are X-linked and/or congenital. However, many acquired skin conditions have been described exhibiting blaschkoid distribution, such as vitiligo, psoriasis, lichen planus, atopic dermatitis, and mycosis fungoides.

Confetti-like depigmentation has been described as an unusual clinical variant of vitiligo. It also has been reported after psoralen plus UVA therapy in patients with more classic vitiligo, numerous domestic chemicals, and in association with mycosis fungoides. In these cases, punctate lesions were disseminated, symmetric on extremities, or limited to areas exposed to chemicals.

REFERENCES