

# Acral speckled hypomelanosis



Jumana Aldhalaan, MBBS,<sup>a</sup> Yasser A. Ghobara, MBBS,<sup>b</sup> Ahmed Alisaa, MBBS, FRCPC,<sup>b</sup> and Mohammed I. AlJasser, MBBS, FRCPC<sup>c</sup>  
*Riyadh, Saudi Arabia*

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## INTRODUCTION

Hypopigmentation is seen in a wide range of skin conditions. The morphology and distribution of hypopigmented lesions help in narrowing down the differential diagnosis. Several diseases present with guttate, speckled, or confetti hypopigmented macules, and cases of speckled hypopigmentation limited to acral areas have recently been described.<sup>1-4</sup> In this report, we describe a 9-year-old girl with acral speckled hypopigmentation and discuss the differential diagnosis.

## CASE PRESENTATION

An otherwise healthy 9-year-old girl of Middle Eastern origin presented with white spots on her hands and feet that had persisted for more than a year. The lesions were asymptomatic but slowly increasing in number with time. There was no family history of any similar lesions.

Physical examination showed multiple well-defined, 2- to 4-mm hypopigmented guttate macules symmetrically affecting the dorsum of both hands and feet with no atrophy (Fig 1). The hypopigmented macules extended, but with less severity, to the forearms and legs (Fig 2). The lesions on the hands and feet were more grouped compared with the lesions on the forearms and legs. Skin lesions were not accentuated under a Wood light. There was no involvement of any other site. Examination results of the hair, nails, and mucous membranes were normal. Based on the clinical findings, a diagnosis of acral speckled hypomelanosis was made. A skin biopsy was offered for histologic confirmation but was refused by the parents. The patient had been treated previously at another hospital with

tacrolimus ointment for several weeks, with no improvement. Narrowband ultraviolet B phototherapy was subsequently started at the same hospital. Significant improvement was noted after 6 months of therapy but was followed by relapse within a few weeks after therapy completion. Another trial of narrowband ultraviolet B phototherapy was discussed with the parents at our clinic.

## DISCUSSION

Speckled, guttate, or confetti hypopigmentation is seen in several dermatoses, including idiopathic guttate hypomelanosis, idiopathic confetti-like leukoderma, familial white lentiginosis, Darier disease, Cole disease, and tuberous sclerosis complex.

Idiopathic guttate hypomelanosis is a common condition that presents with well-defined depigmented macules on the extremities of adults secondary to cumulative sun exposure. Lesions described in idiopathic confetti-like leukoderma and familial white lentiginosis are mainly truncal in distribution, with the latter being more prevalent in those of African descent.<sup>5,6</sup> Truncal guttate hypopigmentation in Darier disease is usually accompanied by other features, including greasy keratotic papules in seborrheic areas, acral keratoses, and V-shaped scalloping of the distal part of the nail.<sup>7</sup> Cole disease is characterized by palmoplantar hyperkeratotic papules and guttate hypopigmented macules affecting the trunk and extremities.<sup>8</sup> Tuberous sclerosis complex is a neurocutaneous disorder with neurologic, cutaneous, cardiac, and renal manifestations. Dermatologic manifestations include hypopigmented ash-leaf macules, shagreen patches, and angiofibromas.<sup>9</sup>

From the College of Medicine, King Saud bin Abdulaziz University for Health Sciences<sup>a</sup>; Derma Clinics<sup>b</sup>; and Division of Dermatology, King Saud bin Abdulaziz University for Health Sciences, Riyadh.<sup>c</sup>

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Correspondence to: Jumana Aldhalaan, MBBS, National Guard, Ar Rimayah, Riyadh, Saudi Arabia. E-mail: [jaldhalaan@gmail.com](mailto:jaldhalaan@gmail.com).

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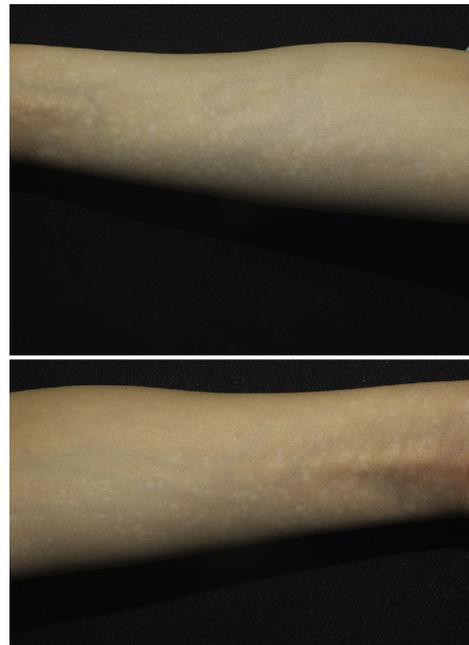
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**Fig 1.** Acral speckled hypomelanosis. Multiple 2- to 4-mm guttate hypopigmented macules symmetrically distributed on the dorsum of both feet. Note the less severe extension of macules to both legs.

Dermatoses characterized by acral pigmentary changes include reticulate acropigmentation of Kitamura, dyschromatosis symmetrica hereditaria, and acromelanosis albo-punctata. Reticulate acropigmentation of Kitamura typically presents with reticulate hyperpigmentation on the hands and feet associated with marked atrophy.<sup>10</sup> Dyschromatosis symmetrica hereditaria manifests in early childhood with a combination of hypopigmented and hyperpigmented macules on the dorsal hands and feet.<sup>10</sup> Acromelanosis albo-punctata is characterized by the generalized hyperpigmentation of the skin and confetti-like hypopigmented macules found mainly on the hands and feet.<sup>11</sup> These hypopigmented macules are also found to a lesser extent in flexural areas.

The clinical picture of our patient, with speckled hypopigmented macules primarily affecting acral areas, does not fit any of the aforementioned entities. There are a few reported cases of pure acral hypomelanosis. Six cases similar to ours were reported in the literature with speckled hypomelanotic macules on normal skin background and no other associated features. The first case described a 14-year-old girl with speckled hypopigmentation on both hands and feet.<sup>1</sup> Unlike our patient, this girl had a positive family history. A skin biopsy specimen showed a significant decrease in the number of melanocytes. Three other reports described 5 patients who presented with similar lesions and a negative family history.<sup>2-4</sup> One of the reported changes was unique: histopathologic evaluation showed a normal melanocyte count with



**Fig 2.** Acral speckled hypomelanosis. Multiple well-defined hypopigmented guttate macules scattered over both inner forearms. Note the more grouped distribution of the lesions distally.

macromelanosomes.<sup>4</sup> All cases share a similar morphology and distribution of hypopigmentation and an early disease onset, which is in agreement with the findings in our case. This pattern of hypopigmentation was named *speckled acral hypopigmentation* or *acral speckled hypomelanosis* by the authors.

The main feature differentiating acral speckled hypomelanosis from other entities is the peculiar distribution of the macules over the hands and feet. Examination of the hypopigmented macules in acral speckled hypomelanosis under a Wood light does not show accentuation. Furthermore, histologic examination shows a decreased or normal number of melanocytes, unlike in vitiligo, in which melanocytes are absent. Unlike other reported cases, our patient had a unique distribution of macules extending with less severity to the forearms and legs.

In conclusion, we present a case of acral speckled hypomelanosis characterized by isolated acral guttate hypopigmented macules on normal background skin color. We support the opinion of other authors that this is most likely a novel entity of acral pigmentary alteration. Future clinical and genetic studies are needed to better characterize this condition.

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