

# Dermatology Digest

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**FIGURE 1**  
Sharply demarcated hyperpigmentation

## Puzzling brown area on toddler's skin

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### ▶ CASE

A 24-month-old mixed Asian Caucasian female presented with an asymmetrical “darkening” of the skin along the anterior trunk. The mother noticed the skin changes when the child was around 15 months old; the pigmentation contrast has increased slightly since then. The toddler had no significant medical history except for moderate eczema since infancy. There was no associated pruritus or discharge, and no other areas of the body were affected. The patient was in the 75% percentile for both height and weight. There was no direct sun exposure to the area.

On physical examination, a sharply demarcated, homogenous, hyperpigmented macular area was visible on the patient's left side (see Figure 1). It started above the nipple line and ran to the genitalia with the medial border at the umbilicus. The pigmentation faded along the axillary line, and there was no visible hyperpigmentation on the posterior trunk. The only other significant finding was a Mongolian spot on the buttocks. The neurologic examination revealed no focal deficits.

### ▶ WHAT IS YOUR DIAGNOSIS?

- *Giant café au lait spot*
- *Segmental neurofibromatosis*

- *Segmental vitiligo*
- *Segmental pigmentation anomaly*

### ▶ DISCUSSION

The diagnosis is segmental pigmentation anomaly (SPA). A perplexing disorder, SPA was first described in the Israeli population by Metzker and colleagues<sup>1</sup> and may manifest as hypopigmentation, hyperpigmentation, or both. Histologic examination shows basal hyperpigmentation only. The exact incidence is unknown because of the large number of pigmentation disorders that may look similar—that is, they may be segmental, dermatomal, or run along the lines of Blaschko. This condition was originally termed *segmental pigmentation disorder* by Metzker,<sup>1</sup> but Orion and colleagues renamed it *segmental pigmentation anomaly*.<sup>2</sup>

Although giant café au lait macules (CALMs) may be linked to genetic mosaicism, this patient's lesion did not have the clinical appearance of a giant café au lait spot, which usually manifests as a round or oval macule ranging in color from dark to light brown (“coffee with milk”) and may have smooth or irregular borders. These spots tend to be present along the buttocks and do not usually appear in such a homogenous manner as seen in this patient. Segmental neurofibromatosis may also manifest with hyperpigmentation, but the hyperpigmentation is not as homogenous as it is in SPA and is usually accompanied by other clinical features, such as axillary freckling and smaller CALMs.<sup>1</sup> Segmental vitiligo manifests with hypopigmentation, and typically the hypopigmentation is complete.

**TREATMENT** SPA is a benign condition that requires no specific treatment. In this case, the diagnosis was made clinically. The condition may fade during adolescence. Sunscreen and observation were recommended, with no follow-up required. **JAAPA**

**Joe R. Monroe, PA-C, MPAS, department editor**

### REFERENCES

1. Metzker A, Morag H, Weitz R. Segmental pigmentation disorder. *Acta Dermatol Venereol* (Stockh). 1983;63:167-169.
2. Orion E, Matz H, Wolf D, Wolf R. Café au lait has hue of its own. *Dermatol Online J*. 2003;9(5):1-8.