

# Transient Skin Rippling in an Infant

Mia A. Mologousis, BS; Elena B. Hawryluk, MD, PhD



A healthy, full-term, 5-month-old infant boy presented to dermatology for evaluation of an intermittent, asymptomatic, rippled skin texture of the left thigh that resolved completely between flares. The parents noted fewer than 10 intermittent flares prior to the initial presentation at 5 months. Physical examination of the patient's skin revealed no epidermal abnormalities, dermatographism, or subcutaneous nodules, and there was no positive Darier sign. A subsequent flare at 9 months of age occurred concurrently with fevers up to 39.4 °C (103 °F), and a corresponding photograph (quiz image) provided by the parents due to the intermittent and transient nature of the condition demonstrated an ill-defined, raised, rippled plaque on the left lateral thigh.

## WHAT'S YOUR DIAGNOSIS?

- Becker nevus
- congenital smooth muscle hamartoma
- infantile transient smooth muscle contraction of the skin
- mastocytoma
- shagreen patch

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From the Department of Dermatology, Boston Children's Hospital, Massachusetts, and the Department of Dermatology, Massachusetts General Hospital, Boston. Mia A. Mologousis also is from Tufts University School of Medicine, Boston. Dr. Hawryluk also is from Harvard Medical School, Boston. The authors report no conflict of interest.

Correspondence: Elena B. Hawryluk, MD, PhD, Massachusetts General Hospital, 50 Staniford St, Ste 200, Boston MA 02114 (ehawryluk@mgh.harvard.edu).

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**THE DIAGNOSIS:****Infantile Transient Smooth Muscle Contraction of the Skin**

A diagnosis of infantile transient smooth muscle contraction of the skin (ITSMC) was made based on our patient's clinical presentation and eliminating the diagnoses in the differential. No treatment ultimately was indicated, as episodes became less frequent over time.

The term *infantile transient smooth muscle contraction of the skin* was first proposed in 2013 by Torrelo et al,<sup>1</sup> who described 9 newborns with episodic skin rippling occasionally associated with exposure to cold or friction. The authors postulated that ITSMC was the result of a transient contraction of the arrector pili smooth muscle fibers of the skin, secondary to autonomic immaturity, primitive reflexes, or smooth muscle hypersensitivity.<sup>1</sup> Since this first description, ITSMC has remained a rarely reported and poorly understood phenomenon with rare identified cases in the literature.<sup>2,3</sup> Clinical history and examination of infants with intermittent transient skin rippling help to distinguish ITSMC from other diagnoses without the need for biopsy, which is particularly undesirable in the pediatric population.

Congenital smooth muscle hamartoma is a benign proliferation of mature smooth muscle that also can arise from the arrector pili muscles.<sup>4</sup> In contrast to ITSMC, a hamartoma does not clear; rather, it persists and grows proportionally with the child and is associated with overlying hyperpigmentation and hypertrichosis. The transient nature of ITSMC may be worrisome for mastocytoma; however, this condition presents as erythematous, yellow, red, or brown macules, papules, plaques, or nodules with a positive Darier sign.<sup>5</sup> Although the differential diagnosis includes the shagreen patch

characteristic of tuberous sclerosis, this irregular plaque typically is located on the lower back with overlying peau d'orange skin changes, and our patient lacked other features indicative of this condition.<sup>6</sup> Becker nevus also remains a consideration in patients with rippled skin, but this entity typically becomes more notable at puberty and is associated with hyperpigmentation and hypertrichosis and is a type of smooth muscle hamartoma.<sup>4</sup>

Our case highlighted the unusual presentation of ITSMC, a condition that can easily go unrecognized, leading to unnecessary referrals and concern. Familiarity with this benign diagnosis is essential to inform prognosis and guide management.

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