

Piezogenic Papules in Ehlers-Danlos Syndrome

A 2-year-old girl was referred to our clinic for evaluation of Ehlers-Danlos syndrome. She had a history of easy bruising, and the physical examination showed hematomas, stretchy skin of velvety texture, and hyperflexible joints, suggestive of the presumed diagnosis. Her father had full-blown Ehlers-Danlos syndrome of the classic type featuring skin hyperextensibility, widened atrophic scars, and joint hypermobility. In both patients, piezogenic papules appeared on the medial and lateral aspects of the heels when standing upright with full weight (Figure).

Piezogenic papules develop when high compressive stress causes herniation of subcutaneous fat into the dermis.¹ The heels are a typical “locus minoris resistenciae.”

Piezogenic papules are common in adults with an occupation that involves standing and in athletes such as marathon runners or weightlifters. However, in children and especially in toddlers, these papules are an unusual finding.

Reports suggest an association of piezogenic papules and Ehlers-Danlos syndrome in which impaired collagen synthesis predisposes to various other kinds of herniation.^{1,2} Ehlers-Danlos syndrome summarizes a heterogeneous group of rare hereditary disorders with the common feature of connective

tissue weakness. Ultrastructural abnormalities of collagen fibrils thereby lead to symptoms in organs frequently exposed to distention such as the skin, ligaments, joints, vessels, heart, colon, and uterus.³ The current classification of Ehlers-Danlos syndrome distinguishes the classic type with skin hyperextensibility, joint hypermobility, and tissue fragility from a hypermobility type, a vascular type, and additional rare types.⁴

In a small study that included 29 patients with Ehlers-Danlos syndrome, piezogenic papules were found in 34.5% of patients but in not a single person of a sex- and age-matched control group.² Piezogenic papules may be painful and occasionally lead to consultation for this otherwise harmless condition. Because these papules are uncommon in toddlers, they might serve as a red flag for Ehlers-Danlos syndrome in this age cohort. ■

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Figure. Piezogenic papules on the medial aspects of the heels in a 41-year-old patient with Ehlers-Danlos syndrome (top) and his 2-year-old daughter (bottom).