Quiz
What are these plaques?

Amor Khachemoune, Shahbaz A Janjua
Ayza Skin and Research Centre, Lalamusa

An 8-year-old, otherwise healthy, girl presented with 1-month history of an eruption of mildly pruritic, rough plaques over the skin of her elbows, knees and upper lateral thighs. Her personal and family medical histories were unremarkable; particularly there was no history of atopic diathesis. She denied the use of any medication in the recent past.

On physical examination, there were multiple, symmetrically distributed, 2 cm to 6 cm round to oval plaques comprised of multiple, grouped, monomorphous, follicular, keratotic, skin colored, 1 mm to 2 mm papules affecting the skin over her elbows (Figure 1), knees and upper lateral thighs. Each papule was bearing a central, protruding, thorny, hair-like, 1-mm keratotic spine (Figure 2). Although, the papules were grouped into plaques, discrete follicular, keratotic papules were also present in the vicinity of the plaques. The lesions were not biopsied, as the consent could not be obtained from her parents.

Address for Correspondence
Dr. Shahbaz A. Janjua, MD,
Ayza Skin and Research Centre, Lalamusa.
E mail: dr_janjua@hotmail.com
**Diagnosis**

Lichen spinulosus

Lichen spinulosus (LS) is a rare, idiopathic, follicular, keratotic dermatosis occurring mostly in children, adolescents and young adults. The male to female ratio is 2:3.¹ No race predilection or seasonal variations have been noted. This condition was first recorded in the literature by Crocker in 1883. He named it “lichen pilaris seu spinulosus.” Since then, there were few case reports until Friedman recorded 35 cases in the Philippines.¹ Characterized by an acute development of symmetrically distributed follicular, keratotic papules that are sharply grouped in round to oval plaques varying in size from 2 cm to 6 cm, LS can appear on the extensor surfaces of elbows and knees, the neck, buttocks, abdomen and the trochanteric regions. Very rarely, the eruption can be generalized. A typical LS papule is 1 mm to 3 mm in diameter with skin-colored conical projection and a central 1-mm to 2-mm keratotic spine. The eruption may be asymptomatic or only mildly pruritic. The lesions may spring up simultaneously or in crops.

**Etiology and pathology**

Although the exact etiology of LS is uncertain, it has been described as a follicular reaction pattern to various environmental agents such as infections, toxins, chemicals and drugs.² It has been attributed to atopy,³ but clear evidence is lacking. Because lesions occur on pressure areas, trauma has been considered a cause, although clinical findings do not suggest koebnerization.¹ LS is also described in patients with HIV infection,³⁴ and in association with Crohn’s disease.⁵ In short, LS remains an idiopathic dermatosis.

On histopathology, LS lesions show a central, infundibular, keratotic plug dilating the follicle and protruding above the epidermal surface. Follicular hyperkeratosis, parakeratosis and acanthosis may be present. Dermal perifollicular and perivascular lymphocytic infiltrate is also seen under light microscopy. It is pertinent to note that LS is histopathologically indistinguishable from keratosis pilaris.

**Differential diagnosis**

Differential diagnosis of LS is broad and includes other follicular keratotic dermatoses such as the following dermatoses:

- **Keratosis pilaris** may closely mimic LS, but is a common dermatosis, of slow onset, appearing most commonly in the first decades of life. Seasonal variation with winter exacerbation is another differentiating point. Morphologically, it is differentiated from LS by the even but diffuse distribution of smaller, keratotic follicular papules, each with a protruding or embedded central hair, occurring commonly on the extensor surfaces of proximal limbs. There are many clinical subtypes with keratosis pilaris alba being the most common and differentiated from keratosis pilaris rubra, another subtype, by the absence of perifollicular erythema. There is a frequent association of keratosis pilaris with ichthyosis vulgaris.

- **Pityriasis rubra pilaris** may be familial or acquired with acute onset but is differentiated from LS by characteristic reddish-orange or salmon-colored scaling, palmoplantar keratoderma and keratotic follicular papules on the dorsal surfaces of the proximal phalanges, elbows and wrists.
**Darier’s disease** is an autosomal dominant genodermatosis, characterized by the symmetrical appearance of follicular keratotic, skin-colored papules on the seborrheic areas. The papules, which are pruritic, ultimately turn yellowish brown and greasy. Palms are commonly involved and the nails undergo changes consisting of red and white longitudinal stripes, ridging and nicking at the distal ends, and are often pathognomonic. Mutations in the gene ATP2A2 (Darier’s gene) have been described to be responsible for the disease. The affected gene encodes for the calcium pump, so the mutation alters cytosolic calcium levels, resulting in disruption of the desmosomes and keratin filaments.

**Phrynoderma** is described as cutaneous manifestations of vitamin A deficiency characterized by follicular hyperkeratosis affecting anterior thighs and posterior arms due to keratinization of the hair follicles giving the general appearance of “toad skin.” Vitamin A deficiency could be primary, mainly due to inadequate intake, or less commonly could be secondary to interruption with the absorption, storage and transport of the vitamin as in celiac disease, cirrhosis of the liver and surgical resection of the duodenum. Children with chronic malnutrition are especially prone to the effects of the hypovitaminosis A. Other manifestations include night blindness, xerophthalmia, keratomalacia and keratinization of the mucosal epithelia of the gastrointestinal and urinary tracts. Eye changes are the earliest signs detected and consist of retinal dysfunction and xerosis of the cornea and conjunctiva. Advanced disease is characterized by the presence of superficial foamy patches of epithelial debris on the bulbar conjunctiva; these are commonly called Bitot’s spots.

**Treating LS**
Lichen spinulosus does not usually spontaneously remit and may persist for several years, if left untreated. However, the lesions may involute gradually over several years. Treatment aims to address the cosmetically unappealing disfigurement caused by the disease. It includes topical application of keratolytic agents including salicylic acid, lactic acid and urea available in different formulations and concentrations. In one study, overnight application of 6% salicylic acid gel under occlusion for 2 weeks and without occlusion for 8 weeks resulted in significant reduction of the keratotic lesions. Topical steroids may be added if pruritus is also present. In another study, treatment with 12% lactic acid lotion and triamcinolone in a moisturizing base also resulted in significant improvement. Topical retinoids have no role in treating LS.

**References**