Early onset linear focal elastosis in a Turkish boy

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A 14-year-old boy applied to our outpatient clinic with a three-month history of lesions on the back. The physical examination revealed linear, yellow, stria-like palpable bands surrounded by a slight erythema extending horizontally across the back. Histopathological examination of the lesional skin demonstrated coarseness in collagen bundles and homogenization and decrement in elastic fibers with van Gieson stain. These features were consistent with linear focal elastosis. In the literature, linear focal elastosis is mainly reported in the elderly. We describe here a case of early linear focal elastosis arising in a 14-year-old Turkish boy.

Key words: elastotic striae, back, linear focal elastosis.

Linear focal elastosis (LFE) is a relatively rare disease of the elastic tissue which was first described in 1989 by Burket et al.[1] in three white men with onset after age 60. Since then, more than 20 cases of different ages have been described in the literature. We describe here an additional case of early LFE arising in a 14-year-old Turkish boy.

Case Report

A 14-year-old boy applied to our outpatient clinic with a three-month history of lesions on the back, which were asymptomatic. The patient denied recent weight gain or loss. The physical examination revealed linear, yellow, stria-like palpable bands surrounded by a slight erythema extending horizontally across the back (Fig. 1). There was no scaling. Histopathological examination of a skin biopsy taken from the band demonstrated coarseness in collagen bundles and homogenization. Within these homogenized areas, aggregated fine elastic fibers were observed with van Gieson stain (Fig. 2).

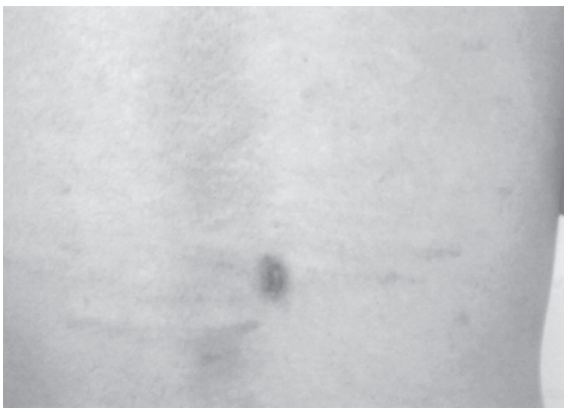

Fig. 1. Linear bands on the lumbar area.

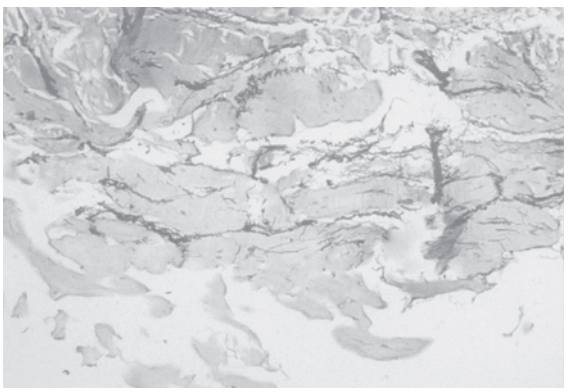

Fig. 2. Aggregated fine elastic fibers (Van Gieson stain × 200).

Discussion

The first case reports of LFE suggested a late onset, but analysis of the recent cases in the literature and ours show a younger onset[2-5]. LFE occurs mainly in patients of Asian descent, but cases with diverse ethnic backgrounds have

also been reported[6,7]. The disease has a male predominance[8-13]. The representative clinical picture is stria-like linear but yellow and palpable indurated bands localized on different portions of the back that extend horizontally[3,8,10-14]. In a few cases, face and leg localization was described[2,4,8,11]. The lesions are asymptomatic and patients are usually not aware of onset of the skin lesions. There is no history of trauma, weight changes or steroid use[6,7]. Some patients described years of duration[14-18]. As duration of the lesions was not well documented in all reports, it is not possible to know the exact time of onset of the disease. There is a lack of data in the literature about the course of the disease. Association with any other skin or systemic disease is accepted to be unusual for LFE[7]. In the literature, there are two cases of LFE associated with adjacent striae distensae[12,14,19]. The majority of cases described in the literature including ours had solely LFE, supporting the hypothesis that LFE arises de novo in the skin.

The etiopathogenesis of the disease is not known. The possible mechanisms proposed today are genetic influences, solar exposure, a nevoid or hamartomatous condition or keloidal repairing process taking place in the late stages of striae distensae[7,15,18]. Localization of LFE lesions suggest that they are not induced by solar exposure. Recently, Akagi et al.[11] reported that they observed similar histologic features in LFE and pseudoxanthoma elasticum in a case who had lesions of both diseases on different areas of the body. They postulated a causative relationship between LFE and pseudoxanthoma elasticum.

Linear focal elastosis shows a focal increase in the number of elongated, fragmented, morphologically immature and mature elastic fibers on histopathological examination[4,13,15,17,20,21]. Electron microscopy demonstrates numerous elongated and fragmented elastic fibers[18]. According to Choi et al.[3], LFE is a dynamic process in which elastolysis and degeneration of elastic fibers occur initially and reactive elastogenesis develops in the static stage. Age, disease duration, presence of erythema and histopathological findings of a decreasing number of the elastic fibers in our patient resemble Choi's case, who had active lesions of LFE.

The differential diagnosis includes disorders of the elastotic tissue such as striae distensae, pseudoxanthoma elasticum, dermatofibrosis lenticularis disseminata and solar elastotic bands. Table I shows the characteristic features

Table I. Differential Diagnosis of Linear Focal Elastosis

Disorder	Epidemiology/feature	Clinical features	Histopathology
Linear focal elastosis	M>F, 7-89 y; no racial predilection	Asymptomatic, yellow or red linear plaques across lumbar spine	Massive, wavy, fragmented basophilic elastic fibers throughout reticular dermis
Striae distensae	Topical/systemic steroid use, pregnancy, weight gain	White, red, or violaceous atrophic bands; axillae, abdomen, thighs, arms, breasts	Flattened epidermis, abnormal collagen fibers, and variable changes in elastic tissue
Pseudoxanthoma elasticum	>40 year, M, exposed to saltpeter; F, obese, multiparous blacks	Coalescing papules in antecubital fossae, periumbilical hyperkeratotic plaques	Fragmented, curled, thickened mineralized (calcified) elastic fibers in mid and deep reticular dermis
Elastosis perforans serpiginosa	M>F, <30 y; no racial predilection	Asymptomatic or pruritic pink, scaly papules and arcuate plaques on head, trunk, and extremities	Increased elastic tissue in papillary dermis with transepidermal elimination of elastotic fibers, inflammatory and keratinous debris
Elastofibroma	F>M, 35-94 y; 2/3 cases in Japanese patients	Slowly growing subcutaneous nodule adjacent to scapula	Fragmented elastic fibers studded with globular aggregates of elastic material appearing serrated
Solar elastosis	Age-related increase in prevalence; more common in whites, photodistribution	Gray to yellow, thickened or atrophic, telangiectatic, fine to coarse wrinkles	Basophilic staining of hypertrophic tangled fibers and amorphous elastotic aggregates in the papillary and upper reticular dermis
Solar elastotic bands	Mainly in Caucasians, sun-exposed skin	1-1.5 cm in diameter, nodular skin-colored bands	Actinic elastosis
Dermatofibrosis lenticularis disseminata	No race predilection	Oval, skin colored papules	Poorly demarcated area of increased dermal collagen, elastic fibers; normal/ decreased, increased
Perforating calcific elastosis	Middle aged, obese, multiparous women	Periumbilical, gradually enlarging, well demarcated, hyperpigmented patch or plaque	Numerous altered elastic fibers in the reticular dermis

F: Female. M: Male.

of these diseases[1,6,7]. White or violaceous bands of atrophic skin in striae distensae, calcified elastic tissue fibers in pseudoxanthoma elasticum, photodistributed nodular skin-colored bands in solar elastotic disease and skin-colored papules in dermatofibrosis lenticularis disseminata contrast sharply with clinical features and histological findings in LFE lesions[1,6,7]. No effective treatment for LFE is known.

This is the first case report of LFE from Turkey in the pediatric population. LFE is a newly recognized disease and we believe that accumulating case reports in the literature will help us to learn more about both the etiopathogenesis and the real epidemiological characteristics of the disease.

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