



## Case Report

### Linear Focal Elastosis in a Family

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#### ABSTRACT

*Linear focal elastosis is a disorder of elastic tissue characterized by yellowish horizontal asymptomatic palpable bands commonly on back. The condition is very less reported, easily overlooked by physicians and less known. We report the cases of LFE in a family which indicated the role of genetic inheritance in the etiology. Two young males 16 year and 18 years old presented with horizontal yellowish streaks over the back since last 5 months and 1 year respectively. They also reported similar lesions in their fathers. These lesions were characteristic of LFE in morphology.*

**Keywords-linear, focal, elastosis, family.**

#### INTRODUCTION

Linear Focal Elastosis (LFE) also known as 'elastoticstriae' is an uncommon elastotic disorder first described by Burket in 1989.<sup>1</sup> It is seen as multiple erythematous to yellowish horizontal streaks over the back generally more common in lower (68%) and middle back and thighs(14%). Other body sites can be involved less commonly such as lower limbs and shoulders. They are generally asymptomatic in nature although slightly palpable and indurated. It is usually misdiagnosed due to its resemblance to striae distensae or may be overlooked by patients and doctors due to its benign nature.

#### CASE REPORT

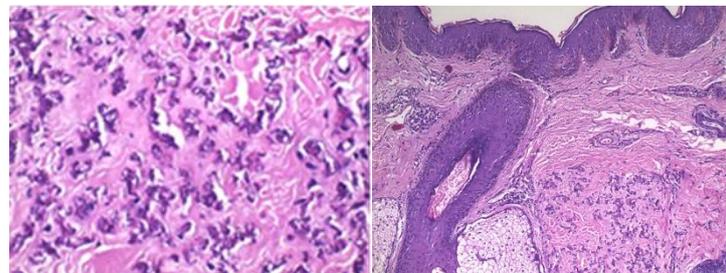
Two young males 16 year and 18 year old respectively presented to our OPD with multiple

skin coloured to yellowish horizontal asymptomatic streaks over back from last 5 months and 1 year respectively. The 16 year old male was son of the 18 year old male's brother. They both reported similar lesions in their fathers. They were related to each other as nephew and uncle as shown in the pedigree(figure 1).The pattern of inheritance can be autosomaldominant with incomplete inheritance as is depicted in the pedigree charting of this family though it still needs to be confirmed genetically. They had no keloidal tendency.

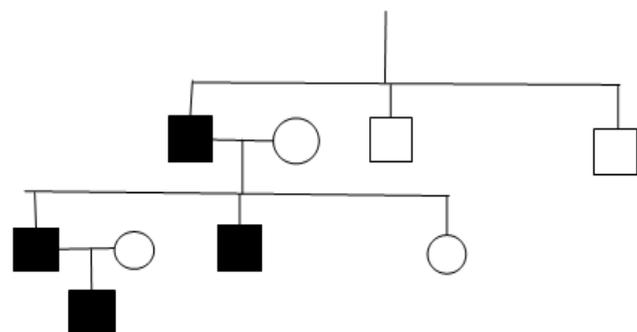
On examination over the lower back of both males, there were well defined erythematous to yellowish coloured horizontal plaques of variable sizes ranging from 5 to 15 cm in length and 0.5 to 1.5 cm in breadth approximately(figure 2,3).These

were palpable with a smooth surface and showed focal atrophy at places.

Histopathological examination showed an increase in the number of elastic fibres in upper and mid reticular dermis. The elastic fibers appear fragmented and curled giving appearance of ravelled wool.(figure 4a and 4b) There was no calcification or suggestion of transepidermal elimination.



**Fig.4** (a) Histopathology (10X) and (b) showing increased elastin in dermis



**Fig.1** Pedigree of involved persons in the family



**Fig. 2**— linear focal elastosis on trunk of 16 year old male



**Fig.3** linear focal elastosis on trunk of 18 year old mal

**DISCUSSION**

**Etiology**

LFE is thought to be a degenerative or regenerative process of striae distensae.<sup>1</sup> Though the causes for striae distensae such as steroids, obesity, cushing’s disease and sudden growth at puberty are not found to be associated with LFE. Increasing incidence in young patients suggest a role of intrinsic defects of elastic fibre metabolism. It is supposed to be associated with keloidal repair process of elastic tissues in striae distensae that is different from the keloid development mediated through TGF-β signaling pathway.<sup>2</sup> The occurrence of LFE in families has led to a suggestion of genetic basis in the development of LFE as is evident by the clustering of cases of LFE in a single family. Till date only three case reports of LFE with familial occurrence has been reported; one in a father and son and other in twins,<sup>3,4</sup> the third one in a brother and sister.<sup>5</sup>To the best of our knowledge there are no case reports of presence of LFE in three successive generations.

**Clinical Features**

LFE most commonly present as asymptomatic palpable horizontal streaks on lower and mid back.<sup>6,7</sup> In early stages it manifests as red atrophic band due to a continuum of elastolysis and elastosis differing only in stage of elastogenesis.<sup>8,9</sup> In later stages it manifests as yellow hypertrophic plaque due to focal increase in elastic fibres separating the dermal collagen bundles. Other sites involved are legs , thighs, face, axilla, shoulders though they are not so commonly involved.

### Histopathology

The middle and lower dermal collagen is separated by bluish grey, fine fibrillar material composed of thin wavy elastic fibres and fragmented elastic fibrebundles. Early lesions show elastolysis with decreased elastin and microfibrillar proteins while in the late lesions elastogenesis can be seen. The elastic fibres are present with fibroblasts and elastogenesis occurs in response to local trauma, UV light or following striae distensae. Dermal elastosis is seen in dermis corresponding to the areas with elastoticstriae seen clinically.

Histopathological differentials include pseudoxanthoma elasticum, connective tissue nevus, solar elastosis, elastofibroma and the clinical features are useful to differentiate these conditions from linear focal elastosis.<sup>10,11</sup>

### Diagnosis

Diagnosis is made on the basis of characteristic morphology and histopathology. It has to be differentiated from striae distensae which are generally red, white or purple atrophic bands as compared to LFE lesions which are raised and easily palpable. Striae distensae give a wrinkled appearance and are generally present on the abdomen, thighs, arms or breast. Other differential diagnosis should also be excluded such as pseudoxanthoma elasticum (PXE), anetoderma, linear xanthomas and solar elastotic bands. PXE are seen as yellowish papules coalescing to form cobble stone like plaques in flexures. Anetoderma shows loss of elastic fibers histopathologically while PXE shows calcified elastic fibres. Linear xanthomas shows foam cells on biopsy. Solar elastotic bands are generally present in photoexposed parts and shows solar elastosis on biopsy.

### Treatment

Generally no treatment is required due to the benign asymptomatic nature of LFE. It can only cause cosmetic problems and there are no systemic associations reported. Patients need to be explained and reassured about the condition. There is no effective treatment known till date.

### CONCLUSION

Linear focal elastosis is commonly misdiagnosed as striae distensae. It may be considered as an elastic tissue metabolism disorder with degenerative and regenerative processes occurring in different stages. Clustering in families suggest a genetic basis of the entity.

### Sources of support nil

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