

# Papular elastorrhexis: a rare acquired elastic tissue disease

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### Abstract

**Background:** papular elastorrhexis is a rare, acquired disorder of elastic tissue. The etiology of papular elastorrhexis is currently unknown, and few cases have been reported in the literature since the condition was first described in 1987. Papular elastorrhexis is clinically characterized by multiple small asymptomatic white or flesh coloured papules predominately located on the trunk and proximal extremities. Initial lesions are usually detected in the first two decades of life, and the condition has been reported more frequently in females. We present a case of a patient with papular elastorrhexis on her neck.

**Case Presentation:** a healthy Caucasian female in her 30s presented with a four-year history of asymptomatic 0.5 mm white papules on her lateral neck. She had no history of inflammation, acne, or trauma to the area, and had no family history of similar lesions. A full body skin exam was completed and was otherwise unremarkable. A punch biopsy was taken of the lesions on the lateral neck and sent for histopathological examination. Hematoxylin and eosin stains were normal, however, elastic stain showed a focal absence of elastic fibers in the papillary dermis and normal fibers in the reticular dermis consistent with a diagnosis of papular elastorrhexis.

**Conclusion:** the diagnosis of papular elastorrhexis is made based on both clinical and histopathological characteristics. The differential diagnosis of papular elastorrhexis includes papular acne scars, nevus anelasticus, Buschke-Ollendorff syndrome, mid-dermal elastolysis, eruptive collagenoma, and pseudoxanthoma elasticum. Papular elastorrhexis is a benign condition, but cosmetic concerns may be significant in some cases. Unfortunately, given the rarity of this condition, no rigorous data exist supporting the use of a particular treatment. Knowledge of the key and distinguishing features of papular elastorrhexis, as highlighted in this case, is critical to accurately diagnose this rare condition.

### Introduction

Papular elastorrhexis is a rare elastic tissue disease first described in 1987.<sup>1</sup> This uncommon condition is clinically characterized by multiple asymptomatic white or flesh coloured papules, typically appearing on the trunk and proximal extremities.<sup>2,3</sup> To date, less than 35 cases of papular elastorrhexis have been reported in the literature.<sup>4</sup> The etiopathogenesis of papular elastorrhexis is unknown.<sup>3</sup> The majority of cases have been reported in females, typically during the first or second decade of life.<sup>5</sup> Papular elastorrhexis appears to be sporadic and acquired, though one report has described a familial clustering of 3 cases.<sup>6</sup> We present a case of a patient with papular elastorrhexis on her neck, an atypical location for this condition.<sup>4</sup>

### Case

A healthy Caucasian female in her 30s presented with a four-year history of asymptomatic 0.5 mm white papules on her lateral neck (Fig. 1). She had not trialed any topical agents to the area at the time of initial assessment. She had no history of inflammation, acne, or trauma to the area, and had no family history of similar lesions. A full body skin exam was completed and was otherwise unremarkable. A punch biopsy was taken of the lesions on the lateral neck and sent for histopathological examination. Hematoxylin and eosin stains were normal; however, elastic stain showed a focal absence of elastic fibers in the papillary dermis and normal fibers in the reticular dermis consistent with a diagnosis of papular elastorrhexis. As the condition is benign, our patient opted for expectant management. Informed consent was obtained from the patient for publication.

### Differential diagnosis

The differential diagnosis for papular elastorrhexis includes papular acne scars, Buschke-Ollendorff syndrome, nevus anelasticus, eruptive collagenoma, pseudoxanthoma elasticum, and mid-dermal elastolysis.<sup>2,3</sup>

Our patient had no history of acne on the lateral neck. Given her noncontributory history, papular acne scars could reasonably be ruled out of the differential diagnosis. In addition, the typical histopathological appearance of papular acne scars is fibroblastic proliferation and elastic tissue changes surrounding pilosebaceous units, which was not consistent with the histopathology of our patient.<sup>4</sup>

Buschke-Ollendorff syndrome is a hereditary autosomal dominant disorder characterized by small hypopigmented connective tissue nevi with or without osteopoikilosis.<sup>2,3</sup> Unlike papular elastorrhexis,

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**Figure 1.** 0.5 mm white papules on the lateral neck of an asymptomatic woman in her 30s consistent on pathology with papular elastorrhexis.

Buschke-Olendorff syndrome is heritable and lesions typically present much earlier in life.<sup>4</sup> Our patient had no family history of similar lesions and first developed lesions in her early 30s, reducing the likelihood of Buschke-Olendorff syndrome.

Nevus anelasticus is a disorder characterized by multiple white or flesh-coloured 1-5 mm papules distributed asymmetrically over the chest and arms.<sup>7</sup> Histologically, an absence of elastic fibers from both the papillary dermis and upper reticular dermis is noted.<sup>7</sup> In our patient, elastic fibers were present in the reticular dermis, a finding more consistent with a diagnosis of papular elastorrhexis.

Eruptive collagenoma is characterized by multiple white or flesh-coloured 2-5 mm papules on the trunk, typically appearing in the first or second decade of life.<sup>4</sup> Histologically, reduced elastic fibers and thickened or homogenized collagen is noted. Collagen has always been reported as prominently increased in eruptive collagenoma, and either increased or normal in papular elastorrhexis.<sup>2</sup> In our patient, collagen was normal on histopathology.

Pseudoxanthoma elasticum is a connective tissue disorder that affects the skin, retina, and cardiovascular system. Skin lesions of pseudoxanthoma elasticum typically appear in the first or second decade of life as small, yellowish papules on flexural areas such as the neck, axillae, or groin. These papules can coalesce and cause a loss of skin elasticity.<sup>8</sup> Key histological features include mineralization and fragmentation of mid-dermal elastic fibers.<sup>9</sup> The lesions of our patient were not coalescing, and neither calcification nor mineral deposits were noted on histopathology.

Mid-dermal elastolysis is an acquired elastic tissue disorder that typically presents in females in the third to fifth decade of life.<sup>4</sup> Clinically, mid-dermal elastolysis is asymptomatic and appears as well-demarcated and symmetric lesions of fine wrinkling with or without perifollicular papules, usually on the trunk or upper extremities. Mild erythema, urticaria, and erythematous papules and plaques have also occasionally been reported.<sup>10</sup> Our patient did not have fine wrinkling of the skin.

## Discussion

Papular elastorrhexis is benign but can have cosmetic implications. No rigorous data exist supporting the use of a particular treatment. One case report documented transient improvement of lesions after intralesional injections of triamcinolone.<sup>11</sup> Other treatments such as isotretinoin, oral antibiotics, and topical tretinoin have not been found beneficial.<sup>4</sup> As such, expectant management is acceptable and often recommended.

Knowledge of the key and distinguishing features of papular elastorrhexis, as highlighted in this case, is critical to accurately diagnose this rare condition. Further research is required to elucidate the etiopathogenesis of this condition and to determine effective treatment strategies.

## Conflicts of interest

ML has no conflicts of interest to disclose.

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