CLINICAL VIGNETTE

Papular Elastorrhexis: A Case Report and Review of the Literature

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Abstract

Papular elastorrhexis (PE) is an uncommon, acquired elastic tissue disorder with an unclear nosology. To date, there have been only 22 cases of PE described in the literature. PE may be an under-reported condition and the paucity of reported cases may stem from the asymptomatic nature of the disease, the absence of extra-cutaneous involvement and the lack of a well-established set of clinical and histopathologic diagnostic criteria. Clinically, lesions are most commonly described as asymptomatic, firm, non-follicular, yellow to white to skin-colored papules involving the trunk and upper extremities. Less commonly reported areas of involvement include the lower extremities, neck, occiput and mandible [1,2]. The most consistently reported histopathologic features of PE are fragmented and decreased dermal elastic fibers. Herein we present a case of PE affecting the upper trunk and occipital-cervical areas of a 61-year-old female, perform a review the PE literature and discuss the nosology of the entity.

Patient Presentation

A 61-year-old woman presented to dermatology clinic with an eight-month history of asymptomatic papules that first appeared on her neck. The patient reported a progressive increase in the number of lesions and the area of lesional involvement. She denied preceding inflammatory skin lesions. Physical exam revealed non-follicular, skin-colored to hypopigmented papules on her upper back, posterior and lateral neck and occipital scalp (Figure 1). No other abnormal cutaneous findings were noted. Ophthalmologic exam was showed no significant abnormalities. Complete blood count, basic metabolic panel, erythrocyte sedimentation rate, liver, and thyroid function tests were all normal.

Histopathology

Haematoxylin and eosin staining of lesional tissue demonstrated a patchy superficial perivascular lymphocyte-predominant infiltrate (Figure 2). Focal papillary dermal pigment incontinence was also noted (Figure 3). Homogenization of dermal collagen fibers was not observed. The epidermis and dermis appeared otherwise unremarkable. Verhoeff–Van Gieson stain revealed a relative reduction of elastic fibers localized to the mid to lower dermis (Figure 4). Intense elastic fiber fragmentation was noted in the same area (Figure 5).

Discussion

There are numerous elastic tissue disorders characterized by decreased elastic tissue[3]. Of these disorders, there are five entities characterized by both decreased and fragmented elastic tissue[3]. PE is one of these five entities. When Bordas, et al first described PE in 1987, the characteristic lesions were clinically described as firm, yellowish, isolated, painless, oval-shaped papules 2-5 mm in diameter[4]. The lesions lacked a perifollicular arrangement and did not have the bladderlike softness characteristic of anetoderma. The lesions were distributed over the abdomen, chest and back of a 17-year-old male and initially appeared when the patient was age 14. Histopathologic examination of tissue sections obtained from three separate lesional biopsies demonstrated no abnormalities on hematoxylin-eosin. However, tissue sections stained with acid orcein revealed a relative decrease in elastic fibers density and intense fragmentation of elastic fibers within the mid to lower dermis.

Since this first report, 21 additional cases of PE have been described in the literature (Table 1). The age at diagnosis ranged from 4 – 47 years, with an average age at diagnosis of 21.2 years. 14 patients were female and 8 patients were male. All reports describe the lesions as being asymptomatic in nature and firm in consistency. The color of PE lesions described in the majority of cases is white. Lesions are less commonly reported as being skin-colored, hypopigmented and yellow[1,4-7]. The lesional size is typically less than 5mm. Most reports describe the lesions as being non-follicular in distribution. The trunk and upper extremities are the most common areas of involvement. The neck and occiput are less common areas of involvement[2]. Extracutaneous involvement has not been reported. On histology,

reported biopsies demonstrate decreased and fragmented elastic fibers. Furthermore, the elastic fiber abnormalities lack folliculo-centricity. The reported distribution of elastic fiber abnormalities within the dermis ranges from localized to the papillary dermis, the mid dermis and/or the reticular dermis. Collagen fibers have been reported as being normal, homogenized, condensed and/or fibrotic. Biopsies from seven patients showed a perivascular lymphocytic or lymphohistiocytic infiltrate[1,2,7]. A biopsy from one patient revealed decreased melanin granules but a normal number of melanocytes within the epidermis[8]. Biopsies from two patients demonstrated the presence of increased interstitial mucin surrounding an area of elastic fiber reduction and fragmentation[9].

The most consistently reported characteristics of PE are[1]: a relative decrease in dermal elastic fibers[2]; intense fragmentation of dermal elastic fibers[3]; absence of folliculocentricity[4]; asymptomatic nature[5]; firm consistency. The differential diagnosis of PE therefore includes four other elastic tissue disorders histologically characterized by decreased and fragmented elastic fibers. These entities include: nevus anelasticus (NA), anetoderma, acquired cutis laxa and post-inflammatory elastolysis and cutis laxa. These aforementioned elastic fiber disorders have clinical characteristics and additional histologic characteristics that distinguish them from PE (Table 2).

The nosology of PE is unclear. Bordas, et al. suggested that PE was a variant of NA, noting that histologically both NA and PE are characterized by decreased and fragmented of dermal elastic fibers [4]. However, Bordas, et al also specified histologic and clinical features that distinguished PE from NA. These distinguishing PE characteristics included[1]: elastic fiber fragmentation more prominent than elastic fiber reduction[2]; lack of lesional folliculo-centricity[3]; no tendency for lesions to coalesce into patches[4]; onset of disease during puberty. Sears, et al. suggested that PE is a variant of connective tissue nevus, noting that histologically both connective tissue nevus of the collagen type and PE are characterized by decreased dermal elastic fibers[10]. However, the literature now reveals that elastic fiber reduction in the context of connective tissue nevi of the collagen type is most likely due to a dilutional effect from increased collagen fibers[11]. Furthermore, there is no elastic fiber fragmentation associated with the collagen type of connective tissue nevus. Schirren, et al. suggested that PE was an abortive form of dermatofibrosis lenticular disseminata (DFLD). DFLD is a connective tissue nevus of the elastic type associated with Buschke-Ollendorff syndrome (BOS)[12]. There are two variants of DFLD, the more common papular type and the less common plaque type. While the papular variant of DFLD demonstrates clinical similarities to PE, it differs histologically by having increased amounts of broad, interlacing elastic fibers in the dermis without fragmentation[5,8]. The plaque variant of DFLD may exhibit decreased elastic fibers, similar to PE. Unlike PE however, the plaque variant of DFLD is not associated with elastic fiber fragmentation and there is a tendency for papules to coalesce into plaques[8]. There have been no reported cases of PE associated with either osteopoikilosis or a LEMD3 mutation, further arguing against the association between PE and BOS. That said, a genetic basis for the expression of PE remains a possibility. Schirrin et al reported PE involving two family members[12].

A lesional biopsy from our patient demonstrated histologic features compatible with that described by Bordas, et al. There was a relative reduction (Figure 4) and intense fragmentation of elastic fibers (Figure 5) observed within the mid to deep dermis. We additionally noted a patchy superficial perivascular lymphocyte-predominant infiltrate and focal dermal pigment incontinence. While Bordas, et al did not describe the presence of an inflammatory infiltrate in his original report, three other groups have observed a perivascular lymphocytic or a lymphohistiocytic infiltrate in the context of PE[1,2,7]. No other reports describing PE have documented focal dermal pigment incontinence. This finding raises the possibility of a preceding inflammatory skin disease or simple traumatic/irritational effect at the lesional site. Given that our patient adamantly denies preceding inflammatory skin changes, the finding of dermal pigment incontinence is more likely due to a traumatic effect.

Our patient's clinical attributes fall within the spectrum of clinical findings reported in association with PE. Like the majority of PE cases, our patient is female. Also similar to most PE cases described in the literature, our patient has asymptomatic, non-follicular, discrete, skin-colored to hypopigmented papules. While the upper back is a common location for PE lesions, the neck and occiput are less commonly reported sites of involvement. The most unusual clinical feature of our patient is her age. At 65 years old, the patient is the oldest reported patient with PE. The distribution of our patient's skin lesions and her age raise the possibility of white fibrous papulosis of the neck (WFPN)/Papillary dermal elastolysis (PDE). However, the presence and prominent of elastic fiber fragmentation of the mid to

reticular dermis argues against the diagnosis of WFPN/PDE.

The diagnosis of PE is challenging given the paucity of reported cases and the lack of stringent clinical and histologic diagnostic criteria. Our patient exhibits some features unusual for PE including her age, involvement of the cervical-occipital region and the presence of focal dermal pigment incontinence. However, the combined clinical and pathologic features of our patient are most consistent with the uncommon elastic tissue disorder, PE. At present the etiology and pathogenesis of PE is unknown. There is no evidence to support the association of PE with either NA, connective tissue nevus of the collagen type or DFLD/BOS. PE is most likely a distinct elastic tissue disorder.

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Figure Legend

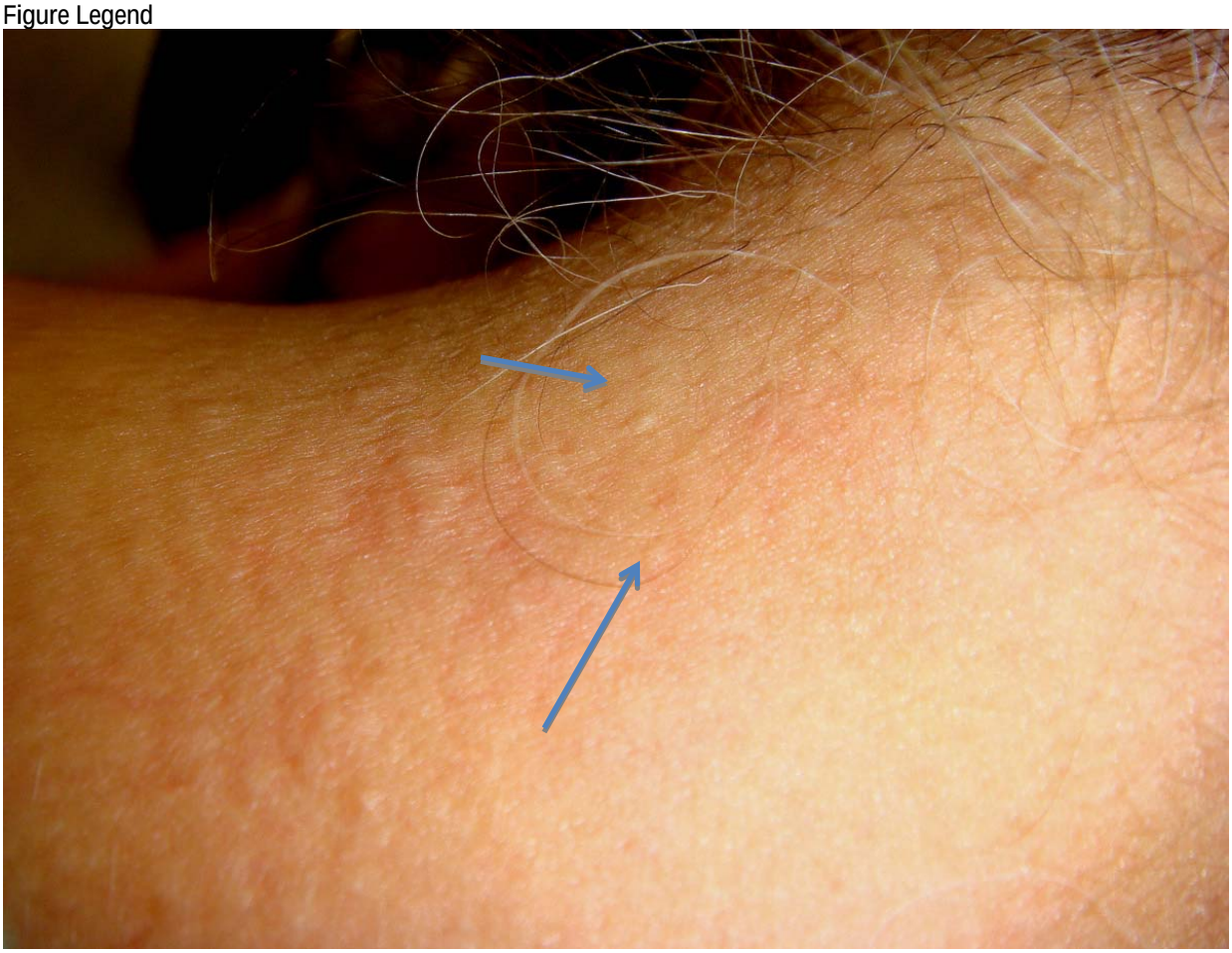

Figure 1: Non-follicular, skin-colored to hypopigmented papules involving the upper back.

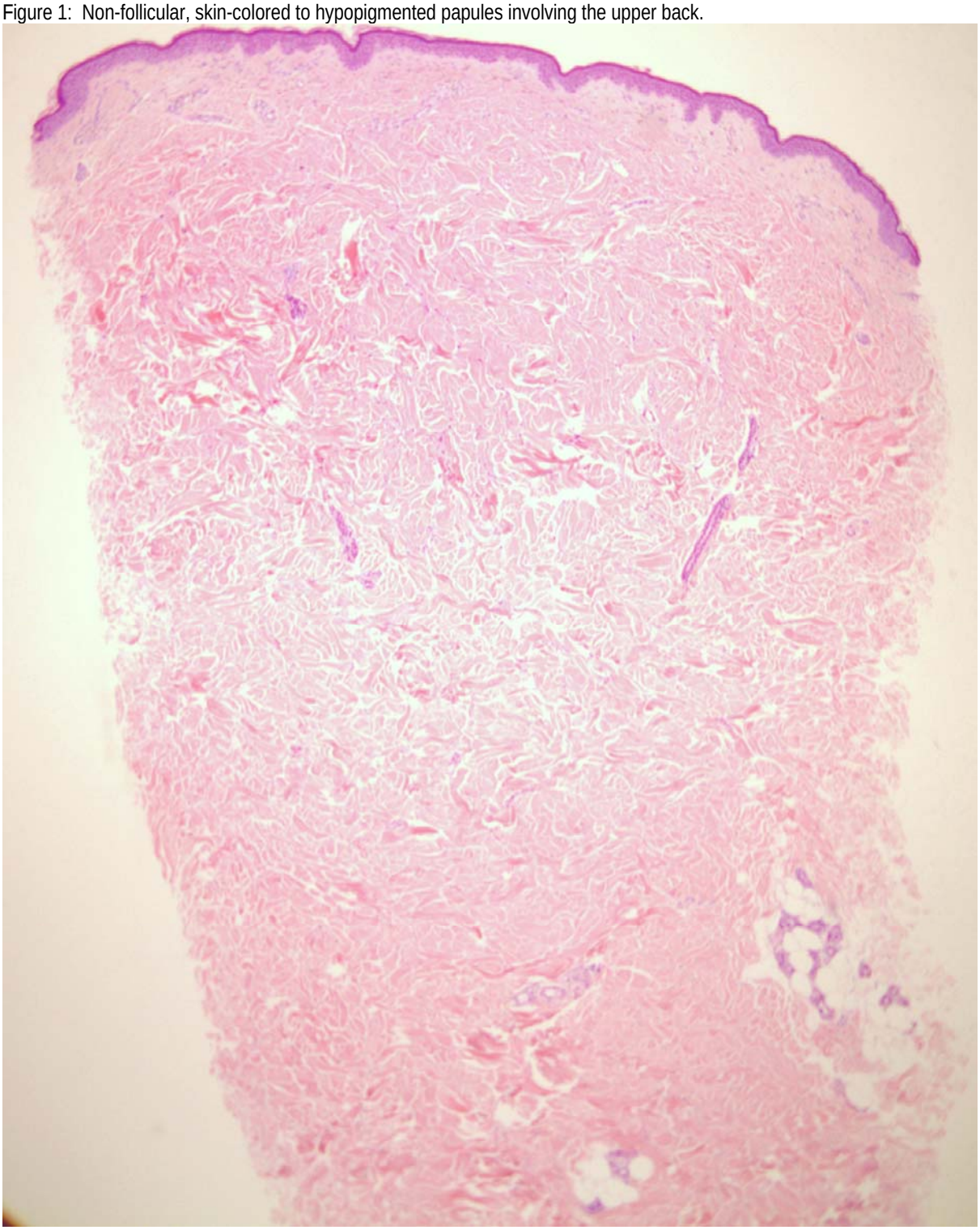

Figure 2: Hematoxylin and eosin staining at 40x reveals a fairly unremarkable epidermis and dermis.

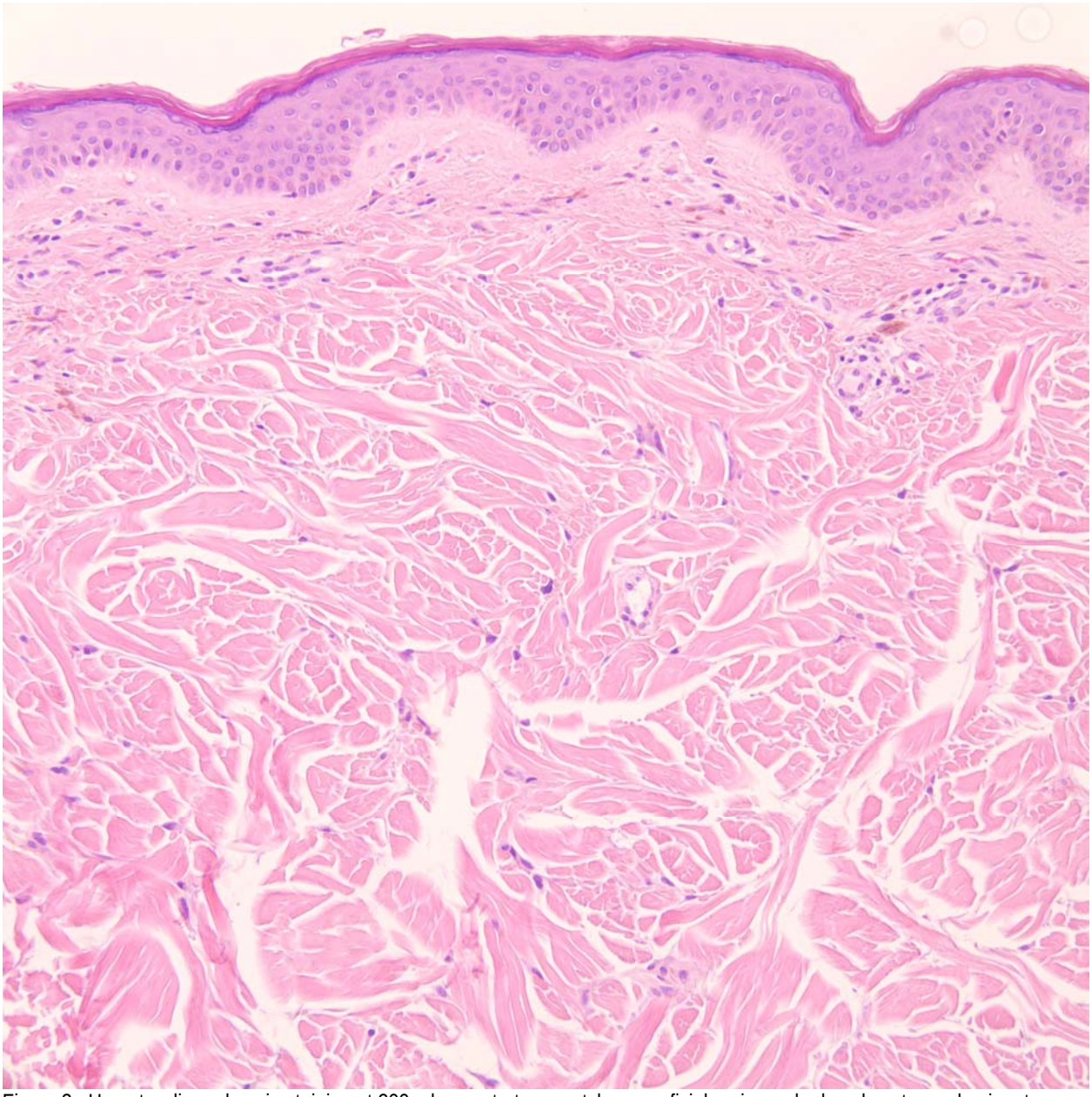

Figure 3: Hematoxylin and eosin staining at 200x demonstrates a patchy superficial perivascular lymphocyte-predominant infiltrate and focal papillary dermal pigment incontinence.

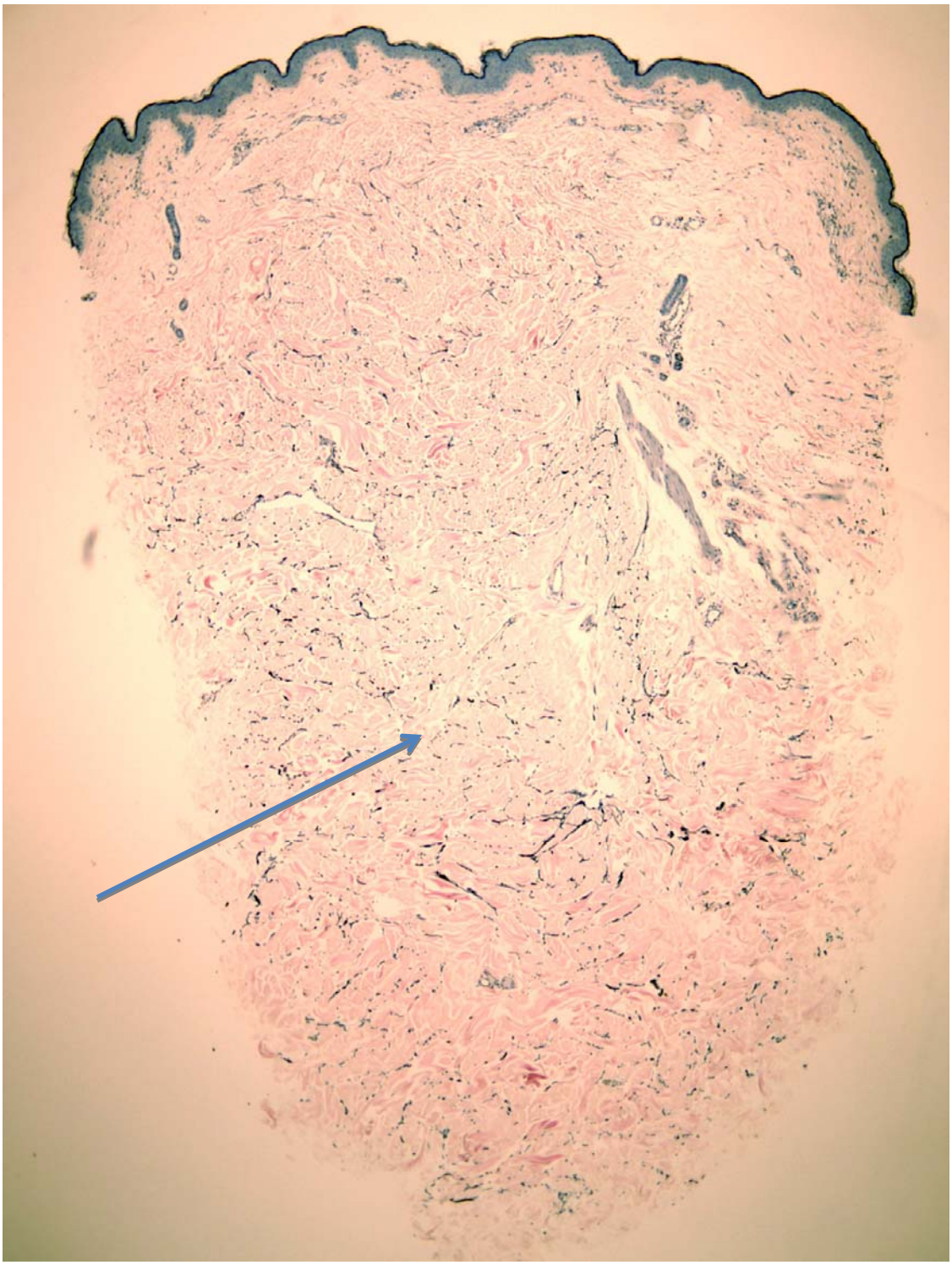

Figure 4: Verhoeff–van Gieson staining at 40x highlights decreased elastic fibers within the mid to deep dermis.

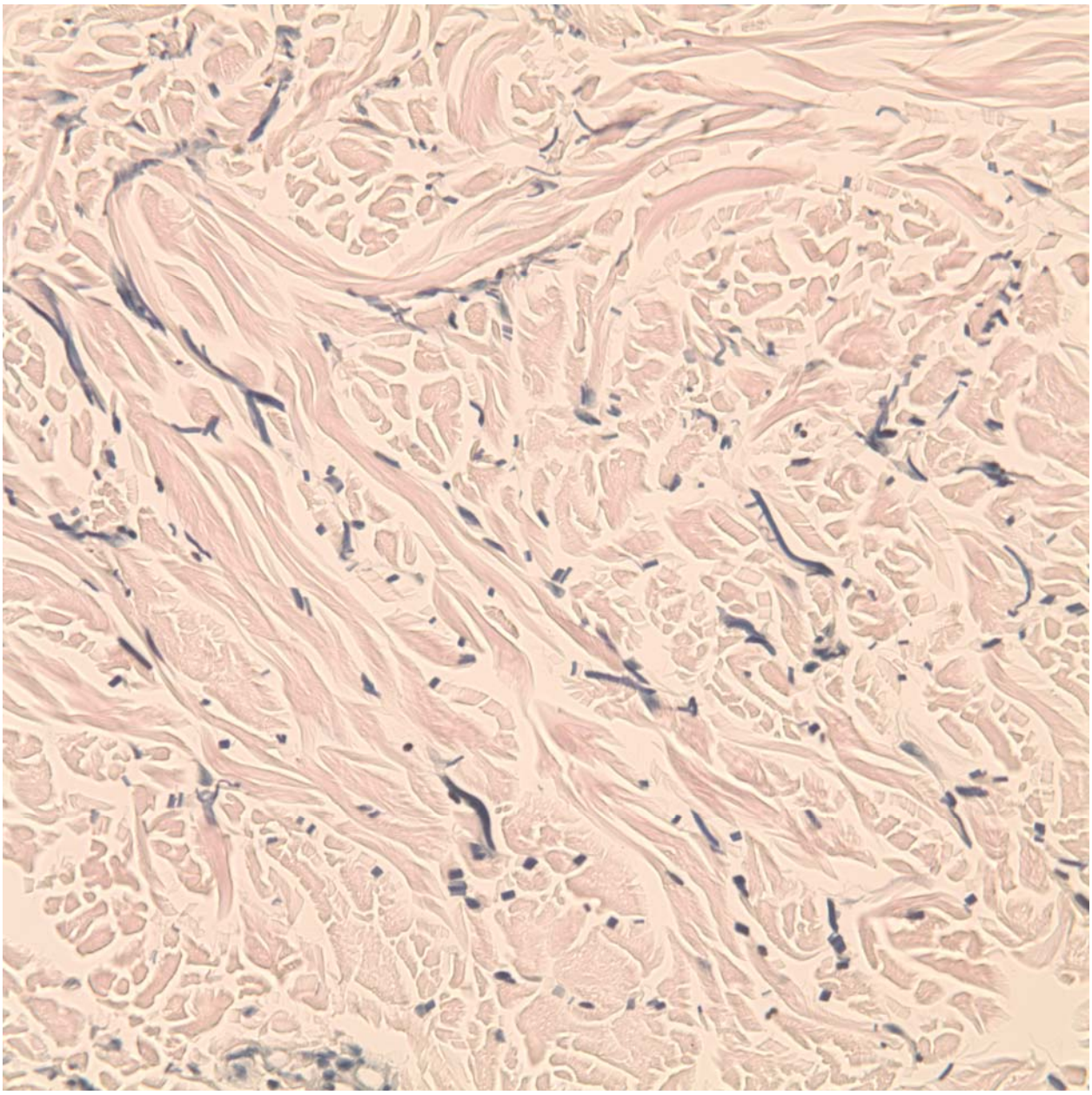

Figure 5: Verhoeff–van Gieson staining at 400x highlights intensely fragment elastic fibers within the mid to deep dermis.