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ORIGINAL ARTICLE

The Importance of Collagen Tissue in Papular Elastorrhexis, Eruptive Collagenoma, and Nevus Anelasticus

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Background: Papular elastorrhexis (PE), eruptive collagenoma (EC), and nevus anelasticus (NA) are described as multiple small papules with decrease, fragmentation, or lack of dermal elastic fibers. These diseases are suggested to be the same entity. The change of collagen fibers in the conditions has not been addressed to date. **Objective:** We compared the clinical features of the 3 diseases and investigated changes in the collagen fibers involved. **Methods:** Twenty-four cases of PE, 12 cases of EC, and 2 cases of NA found in PubMed and the Korean database were reviewed. Changes in dermal collagen fibers in 10 cases with histological figures were investigated. **Results:** There were significant similarities between the 3 entities in terms of their clinical features. Four patients with PE and 2 with EC with fine, dense collagen fibers were women who had multiple white to hypopigmented, slightly indurated to firm, millimeter-size papules on the trunk and/or extremities that progressed gradually after developing in the patients' first to third decades. **Conclusion:** The 3 conditions are the same clinical entity in our opinion; such cases with fine, dense collagen manifest typical features. **(Ann Dermatol 28(2) 210～215, 2016)**

-Keywords-
Collagen fiber, Connective tissue nevus, Elastic fiber, Eruptive collagenoma, Nevus anelasticus, Papular elastorrhexis

INTRODUCTION

Papular elastorrhexis (PE), eruptive collagenoma (EC) and nevus anelasticus (NA) have been described as multiple small papules with decrease, fragmentation, or lack of dermal elastic fibers. The conditions were first described in 1987, 1966, and 1921, respectively[1-3]. The term *nevus anelasticus* was introduced by Staricco and Mehregan[4] in 1961 as a more adequate expression for describing the entity named by Lewandowsky[3] as *naevus elasticus regionis mammariae* in 1921.

It is suggested that PE, EC, and NA might be the same entity. Bordas et al.[1] suggested that PE was a variant of NA in the first reported case of PE. In addition, it was suggested that EC is inseparable from PE[5]. Other authors have suggested that NA and EC should be considered variants of PE[6].

Ryder and Antaya[7] reviewed previous reports of the 3 entities and found remarkable similarities between them. All 3 begin to appear before the age of 20 years, and the sites of the lesions are mainly the trunk and upper extremities. A lack of history of trauma, inflammation, family history, or extracutaneous manifestations is also a common feature. All 3 showed a decreased amount of elastic tissue in biopsy specimens. Therefore, the authors concluded that PE, EC, and NA are the same entity.

Although changes in elastic fibers have been well evaluated in the 3 conditions, changes in collagen fibers have not attracted attention. There have been inconsistencies in

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previous reports' descriptions of collagen fibers. Some authors reported homogenized, thick collagen fibers. On the other hand, others reported that the collagen tissue was normal. We experienced an interesting case in 2011 and reported it as PE[8]. The patient had fine, compacted collagen fibers in the upper dermis, where elastic fibers were reduced and fragmented, which prompted our interest in changes in collagen fibers as an important histological feature in these diseases.

We reviewed previous reports of PE, EC, and NA, and changes in collagen and elastic fibers were investigated.

MATERIALS AND METHODS

We searched reports of PE, EC, and NA in PubMed and a Korean database. Demographic information and clinical manifestation of reported cases of the 3 diseases were summarized, and similarities and differences between the 3 conditions were analyzed.

We selected cases with histological figures from which we could evaluate the status of dermal collagen and elastic fibers. Changes in dermal collagen and elastic fibers of the select cases of the 3 diseases were summarized and compared.

According to the status of dermal collagen fibers, the se-

Table 1. Demographic information and clinical manifestation of reported cases

Variable	Papular elastorrhexis (n=24)[1,6,8-21]	Eruptive collagenoma (n=12)[22-32]	Nevus anelasticus (n=2)[7,33]
Sex (male:female)	7:17	5:7	0:2
Average age, yr (range)	21.0 (4～45)	23.9 (12～42)	16.5 (16～17)
Average age at onset, yr (range)	15.9 (3～32) (n=20)	16 (2～38) (n=10)	12.5 (12～13)
Family history			
Yes	3 (from single family)		0
No	16	8	2
Progression pattern			
Rapid	1	2	
Slow	10	5	1
Lesion			
Number			
Single	0	0	0
Multiple	24	12	2
Relation with hair follicle			
Perifollicular			
Nonfollicular	20		1
Size			
Papules only	21	4	2
Papules with nodules and/or plaques		8	0
Hardness			
Soft		1	1
Slightly indurated to firm	9	8	1
Hard	1		0
Color			
Whitish to hypopigmented	24	2	1
Yellowish		1	
Skin-colored		7	1
Erythematous		1	
Location			
Scalp	1	0	0
Face	1	3	0
Neck	2	1	0
Trunk	22	10	2
Upper extremities	11	5	1
Lower extremities	6	4	0
Radiographic examination			
Normal	19	8	
Abnormal			

lected cases were reclassified into 3 groups: (i) normal collagen group, (ii) fine, dense collagen group, and (iii) thick, dense collagen group. Demographic information and clinical manifestations for the 3 groups were summarized and compared.

RESULTS

Twenty-four cases of PE[1,6,8-21], 12 cases of EC[22-32], and 2 cases of NA[7,33] were found. One case of EC[24] was reported in Korean in the Korean literature, and the others were in English. Demographic information and clinical manifestations for the cases are summarized in Table 1.

There were significant similarities between the 3 entities. All 3 diseases showed female predominance, and the average onset age was mid second decade. There was no family history except in 3 cases of PE from a single family[20]. The lesions progressed gradually in most cases. The lesional features of number, hardness, and location showed no significant difference between the 3 diseases. All cases had multiple lesions, and most lesions were slightly indurated to firm. The trunk and extremities were the major involved sites. All PE and NA cases in which relationships with hair follicles were described had nonfollicular lesions. Relationships with hair follicles were not mentioned in the EC cases. All lesions were papules of several millimeters in size in PE and NA, but 8 cases of EC had nodules and/or plaques along with papules. All PE lesions were whitish to hypopigmented, whereas 7 cases of EC had skin-colored lesions. There were no abnormal radiologic findings.

Among the reports, we selected only cases with histological figures in which we could evaluate the status of dermal collagen and elastic fibers. Six cases of PE[6,8-10,16,18], 3 cases of EC[24,25,27], and 1 case of NA[33] were selected. Changes in dermal collagen and elastic fibers in those cases are summarized in Table 2. The dermal area in which elastic fiber was reduced and fragmented was the upper dermis in 9 cases except in 1 case of NA, in which elastic fiber alteration was observed from the upper to deep dermis. Changes in collagen fibers in the elastic fiber-reduced area could be classified into 3 patterns: (i) normal collagen, (ii) fine, dense collagen, and (iii) thick, dense collagen. Four cases of PE[8,10,16,18] and 2 cases of EC[24,27] had fine, dense collagen tissue. In 2 cases of PE[6,9] and 1 case of NA[33], collagen fibers did not show any change; and only 1 case of EC[25] had the thick, dense collagen fibers.

The 10 selected cases were reclassified into 3 groups according to the status of collagen fibers in the elastic fiber-reduced dermal area: (i) normal collagen group, (ii) fine, dense collagen group, and (iii) thick, dense collagen group. Demographic information and clinical manifestations for the 3 groups are summarized in Table 3. The fine, dense collagen group had all women, and the average onset age was 13.3 years (range, 3～25 years). Three cases progressed gradually, but 1 case progressed rapidly, with all lesions developing within 1 week. In all cases, multiple asymptomatic, less-than-1-cm-size, scattered papules developed on the trunk and/or extremities. Exceptionally, a few lesions also developed on the face and neck in 1 patient. The lesions were whitish to hypopigmented and slightly indurated to firm in most patients.

Three cases in the normal collagen group differed from cases in the fine, dense collagen group in their clinical manifestations. The first case[9] reported as "eruptive PE" was the only male patient and had the latest onset age among the 10 selected cases. The involved sites of face and scalp also differed from the main involved areas. The lesions had developed rapidly, which was also unusual. The second patient[33] was a 17-year-old female who had papules that were grouped and localized on the right areola. Elastic fiber alteration was observed from the upper to deep dermis in the biopsy specimen. Judging from the clinical figure, the last patient[6] seemed to have numerous lesions.

One case with thick, dense collagen[25] had a unique clinical manifestation: dozens of lesions were localized on the

Table 2. Changes in dermal collagen and elastic fibers in cases with histological figures in which the status of dermal collagen and elastic fibers could be evaluated

Variable	Papular elastorrhexis (n = 6)[6,8-10,16,18]	Eruptive collagenoma (n = 3)[24,25,27]	Nevus anelasticus (n = 1)[33]
Involved dermal area	Upper dermis	Upper dermis	Upper and deep dermis
Collagen fiber			
Normal	2	0	1
Fine dense	4	2	0
Thick dense	0	1	0
Elastic fiber	Reduced and fragmented	Reduced and fragmented	Reduced and fragmented

Table 3. Demographic information and clinical manifestations of three groups classified according to the status of collagen fibers in the elastic fiber-reduced dermal area

Variable	Normal collagen (n=3)[6,9,33]	Fine dense collagen (n=6)[8,10,16,18,24,27]	Thick dense collagen (n=1)[25]
Sex (male:female)	1:2	0:6	0:1
Average age, yr (range)	21.7 (17～30)	15.5 (9～26)	15
Average age at onset, yr (range)	18.3 (12～29)	13.3 (3～25)	15
Family history			
Yes	0	0	0
No	3	6	1
Progression pattern			
Rapid	1	1	0
Slow	2	3	1
Lesion			
Number			
Multiple but not numerous	1	6	0
Numerous	2	0	1
Size			
Papules only	3	6	0
Papules with nodules and/or plaques	0	0	1
Relation with hair follicle			
Perifollicular			
Nonfollicular	2	3	
Hardness			
Soft		1	0
Slightly indurated to firm	1	4	1
Color			
Whitish to hypopigmented	3	5	0
Skin-colored	0	1	0
Erythematous	0	0	1
Location			
Scalp	1	0	0
Face	1	1	0
Neck	0	1	0
Trunk	2	6	1
Upper extremities	0	6	0
Lower extremities	0	6	0
Radiographic examination			
Normal	2	4	
Abnormal			

left trunk in a zosteriform distribution. Peculiarly, the color of the lesions was erythematous, and some lesions were confluent.

DISCUSSION

We searched reports of PE, EC, and NA in PubMed and the Korean database and found 24 cases of PE, 12 cases of EC, and 2 cases of NA. Demographic information and clinical manifestations of the 3 diseases had significant similarities on comparative evaluation. EC has been known to present with an acute onset[2]. However, 5 of the 7 cases of EC whose progression pattern was described progressed gradually, as in PE and NA. NA was first described as perifollicular lesions[3]. However, 1 of the 2 NA cases included had nonfollicular lesions, and a relationship with hair follicles was not described for the other patient.

We selected 10 cases with published histological figures in which we could evaluate the status of dermal collagen and elastic fibers. The cases were reclassified into 3 groups according to collagen tissue changes in the dermal area where elastic fibers were reduced and fragmented: (i) normal collagen group, (ii) fine, dense collagen group, and (iii) thick, dense collagen group.

Six cases in the fine, dense collagen group consisting of 4

cases of PE and 2 cases of EC showed similar clinical features. The condition developed in women between the 1st and 3rd decades, and the lesions progressed gradually. There were multiple asymptomatic, white to hypopigmented, slightly indurated to firm, less-than-1-cm-size, scattered nonfollicular papules on the trunk and/or extremities. Electron microscopic examination was performed in 1 case of that group[27]. Scanning electron microscopy of the dermis showed that collagen fibers did not form bundles. Collagen fibers were individualized, forming waved, compact masses resembling noodles, and hence, empty spaces normally seen between dermal collagen bundles disappeared. On the other hand, normal collagen bundles were easily observed in the normal skin, and empty spaces were seen among the bundles.

Three cases in the normal collagen group differed from the fine, dense collagen group in clinical manifestations. Each case had peculiar clinical features with few similarities. One case in the thick, dense collagen group also had clinical features that differed from other cases.

Ryder and Antaya[7] suggested that PE, EC, and NA are the same entity. Our conclusions corroborated that suggestion. Furthermore, the fine, dense collagen group showed typical clinical and histological features of this condition. Until now, decrease in elastic tissue has been emphasized, but changes in collagen fibers also should be emphasized because collagen tissue mainly contributes to the formation of the lesions. It is possible that the decrease in elastic tissue is secondary to the change in collagen bundles—such as a dilutional effect.

Connective tissue nevi are acquired dermal connective tissue hamartomas characterized predominantly by an imbalance in the relative amount and distribution of collagen, elastin, or proteoglycans[34]. The nevi are classified according to histological aspects. When collagen predominates, they are called collagenomas. Lesions in which elastic tissue predominates are called elastomas[34,35]. Therefore, we suggest *papular collagenoma* as a new name for the disease because it represents the disease's clinical and histological features.

Four cases with normal collagen or thick, dense collagen showed different clinical features as well as histological features from the fine, dense collagen group. We considered them to be conditions different from papular collagenoma or its variant.

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