Upper-Mid Dermal Elastolysis: A case report

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ABSTRACT:

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Mid-dermal elastolysis (MDE) is a rare dermatologic disease affecting extracellular matrix fibers, particularly elastic fibers. It is characterized by asymptomatic, symmetrically distributed fine wrinkles with perifollicular papular protrusions. The diagnosis is confirmed by histologic elastic stains showing focal loss of elastic fibers strictly in the mid-dermal layer which helps differentiating the entity from other elastic fiber disorders.

We are hereby reporting a patient with atypically cutaneous and histological presentations of MDE. An 84 year old female presented with a history of localized wrinkles and skin-colored papules on both inframammary areas. The lesions are follicular origins and asymptomatic. The routine H&E stains revealed minimal changes but the elastic stains showed absent of elastic fibers in the upper and mid dermis sparing perifollicular areas. She lives in Bangkok, Thailand and there is no other family member experiencing the same presentation. The lesions share clinical and histologic features of both upper dermal elastolysis and mid dermal elastolysis which never been reported in previous literatures.

Key words: Upper-mid dermal elastolysis, Elastic fiber disorder

บทคัดย่อ :

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หน่วยผิวหนัง ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย

โรคมิดเดอร์มอล อิลาสโตไลซิส เป็นโรคผิวหนังที่พบได้น้อย เกิดจากความผิดปกติของเส้นใยอิลาสติคในชั้นหนังแท้ ประวัติและอาการแสดงของโรคได้แก่ รอยเหี่ยวย่นที่ผิวหนัง ร่วมกับตุ่มเนื้อยื่นบริเวณรอบรูขุมขน ซึ่งเกิดขึ้นอย่างสมมาตรทั้งสองข้างของร่างกายและมักไม่มีอาการ โดยการตรวจวินิจฉัยยืนยันทำได้ด้วยการตรวจทาง พยาธิวิทยาผิวหนัง ซึ่งเมื่อย้อมด้วยสารย้อมเส้นใยอิลาสติค พบว่ามีการลดลงและหายไปของเส้นใยอิลาสติคอย่างเฉพาะที่ในบริเวณชั้นหนังแท้ส่วนกลาง

รายงานฉบับนี้เป็นการนำเสนอผู้ป่วย หญิงอายุ 84 ปี มาตรวจรักษาด้วยประวัติ ตุ่มเนื้อยื่นบริเวณรอบรูขุมขนใต้ราวนมทั้งสองข้าง ไม่มีอาการผิดปกติใดและตรวจพบรอยย่นของผิวหนังบริเวณรอยโรคร่วมด้วย ผลการตรวจย้อมพิเศษทางพยาธิวิทยาผิวหนังพบว่ามีการหายไปของเส้นใยอิลาสติค ในชั้นหนังแท้ทั้งส่วนบนและส่วนกลาง แต่ยังคงไว้ซึ่งเส้นใย อิลาสติค บริเวณรอบรูขุมขนอยู่ ซึ่งอาการและอาการแสดงของผู้ป่วย ร่วมกับผลตรวจทางพยาธิวิทยาผิวหนัง แตกต่างจากโรคมิดเดอร์มอล อิลาสโตไลซิส ที่เคยรายงานมาก่อนหน้าที่ผ่านมา

คำสำคัญ: โรคมิดเดอร์มอล อิลาสโตไลซิส, ความผิดปกติของเส้นใยอิลาสติค

Introduction

Mid-dermal elastolysis (MDE) is a rare dermatologic disease affecting extracellular matrix fibers, particularly elastic fibers. It is characterized by asymptomatic, symmetrically distributed fine wrinkles with perifollicular papular protrusions[1]. The disease shows no familial clustering and, due to scarcity of the patients, most are case reports. While many hypotheses of pathogenesis have been postulated, such as aberrant of lysyl oxidase enzyme activity, anti-elastin antibody production and copper metabolism abnormalities, nevertheless, none of them is clinically proved in MDE patients[2]. The diagnosis is based primarily on histological section with elastic stain showing focal loss of the elastic fiber confined to mid-dermis which could help to differentiate the disease from other elastic fiber defects.

Case report

An 84 years old female presented with a history of multiple symmetrical skin-colored papules on both inframammary areas for several months. She denied any preceding symptoms as erythema or pain. The lesions never progressed to other sites of the body. Her underlying diseases are hypertension and diabetes. No other of her family members experienced the same manifestations.

Cutaneous examination revealed multiple yellowish to skin-colored follicular papules symmetrically arranged on both inframammary areas (Figure 1). The skin surrounding the lesions shows wrinkles and superficial epidermal atrophy without depression, out-pouching sac and skin redundancy.

Skin biopsy obtained from the papular lesion was stained with H&E and van Gieson elastic stain. In H&E section, the dermis was thickening. The papillary dermis displayed mild fibrosis with increasing collagen

fibers which are not sclerotic. There were insignificant perivascular inflammatory infiltration and no elastocytophagocytosis was detected. No abnormal material deposition was detected. The epidermis shows effacement of rete ridges. Additional elastic stain revealed markedly focally decreased elastic fibers in the upper and mid dermis while the perifollicular elastic fibers were preserved. (Figure 3&4).

The diagnosis was based on clinical manifestation and histopathological findings.



Figure 1



Figure 2

Figure 1, 2 demonstrate multiple symmetrical skin-colored follicular papules on both inframammary areas of the patient. Some areas showed wrinkles and superficial epidermal atrophy of the surrounding skin.

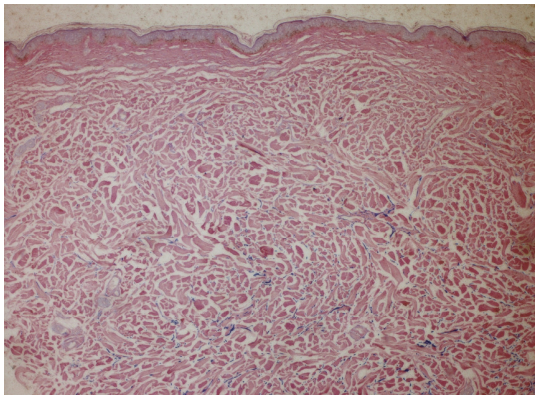


Figure 3

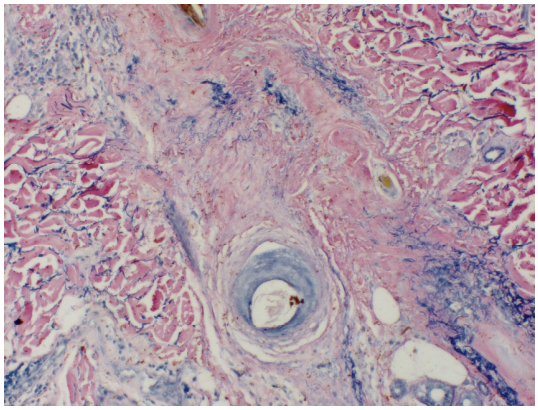


Figure 4

Figure 3, 4 show van Gieson elastic stain of the sample obtained from the papular lesions. There were markedly focally decreased elastic fibers in the upper and mid dermis comparing with the deeper dermis. The perifollicular elastic fibers were spared.

Discussion

Mid-dermal elastolysis (MDE), firstly described in 1977 by Shelly and Wood[3], is diagnosed mainly on histopathological basis, which reveals specific loss of elastic fibers in mid-dermis with no involvement of

more superficial papillary dermis and deeper reticular dermis including perifollicular elastic fibers[4]. Due to rarity of the disease, there are only few cases reported up to date. MDE is far more common among Caucasians with female predilection and median age of onset is 34 years (ranging from 6 to 70 years).[1] While the precise pathogenesis remains unclear, many hypotheses have been postulated, for instances, aberrant of lysyl oxidase enzyme activity, anti-elastin antibody production[5] and copper metabolism abnormalities.

Table 1 Differential diagnoses of elastic fiber disorders

Disorder	**MDE**	**Anetoderma**	**Annular elastolysis giant cell granuloma**	**Cutis laxa**	**Pseudoxanthoma elasticum-like papillary dermal elastolysis**
Clinical presentations	Fine wrinkles arranged parallel to skin cleavage lines and/or protruding perifollicular papules (may occur with peau d'orange appearance) Rare presentations : persistent reticular erythema Locations : trunk and proximal limbs	Pouch-like herniations of floppy skin Preceding inflammation may occur. Locations : trunk and proximal extremeties	Atrophic wrinkles in the center of erythematous bordering annular plaques Locations : sun-exposed areas as the face	Coarse wrinkles with loose redundant skin Complications of systemic elastolysis usually occur. Locations : face	Multiple non-follicular papules with yellowish hue, cobblestone appearance Clinical presentations resemble pseudoxanthoma elasticum. Locations : flexor forearms and axillae
Histologic findings	Band-like loss of elastic fibers in mid-dermal layer Variable degree of inflammatory cell infiltration Giant cells with elastophagocytosis may be observed.	Loss of elastic fibers in both papillary and reticular dermis Variable degree of inflammatory cell infiltration Giant cells with elastophagocytosis may be observed.	Decreased or total loss of elastic fibers in the center of lesions Granulomatous infiltration of the dermis with histiocytes, lymphocytes, and ginat cells	Granular degeneration and fragmentation of elastic fibers throughout the dermis Neutrophilic and lymphocytic infiltration with elastophagocytosis may be observed	Band-like loss of elastic fiber in papillary dermis

Previous reported cases displayed various clinical presentations. Ones of the most recognized clinical features were asymptomatic, symmetrical fine wrinkles with or without skin-colored perifollicular papular protrusions[6] affecting trunk and upper extremities, particularly in photodistributed areas. Other additional reported presentations are urticaria with erythematous papules and plaques and persistent reticular erythema[7].

Our patient presented with classic symmetrically arranged wrinkles and skin-colored follicular papules on both inframammary areas which were frequently reported in antecedent cases of MDE.

The typical histopathologic examination of MDE lesion with hematoxylin and eosin stain shows edema in mid-dermal layer and normal findings of upper papillary and deep reticular dermis, whereas elastic stains (such as Orcein stain, van Gieson stain) show band-like or focal loss of elastic fiber in mid-dermis. Comparing to our case, the H&E stain revealed edema in upper and mid-dermis which was confirmed by elastic van Gieson stain, displaying band-like absence of elastic stain all over upper and mid-dermis. In the perifolliculare subtype (MDE type II), elastic fibers are preserved in both papillary dermis and perifollicular areas. For this type of MDE, perifollicular papules are the most prominent clinical manifestation.

Anetoderma is an important differential diagnosis for MDE as both share clinical and histopathologic similarities of elastic fiber loss. However, due to focal loss of the fibers in both papillary and upper reticular dermis, important clinical features of anetoderma are depressions and sac-like protrusions occasionally accompanied with depigmented area.

Our patient presented with focal bilateral inframammary wrinkles with a few follicular papules without skin depression or out-pouching sac which were commonly seen in MDE. Contrasting to the clinical impression, the elastic stain showed markedly decreased to absent of elastic fibers in upper and mid dermis which was typical picture of anetoderma, however; the perifollicular fibers were still spared as in perifollicular subtype of MDE. In this case, clinic-pathological correlation shared features of both diseases.

There was an antecedent report of an 86-year-old white female presenting with yellowish papules and wrinkles of perilesional skin similar to those of our patient. The elastic stain showed absence of elastic fiber focally specifically in upper papillary dermis[8]. The lack of elastic fiber in upper dermis is not a histological feature of typical reported MDE, and the case was labeled as “Upper-dermal” elastolysis. Since the elastic stain of our patient revealed loss of elastic fibers in both upper and mid-dermis with perifollicular spare, these manifestations might be either on the spectrum between perifollicular type of MDE and anetoderma or a separated entity. This “upper-to-mid” dermal elastolysis condition has never been reported in earlier literatures.

Table 1 shows possible differential diagnoses which are group of disorders sharing defects of the elastic fiber including anetoderma, cutis laxa, annular elastolytic giant cell granuloma, and pseudoxanthoma elasticum-like papillary dermal elastolysis. The clinical and histological clues should be pulled together for guiding to the precise diagnosis.

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