Periumbilical Pseudoxanthoma Elasticum

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We report a case of periumbilical pseudoxanthoma elasticum in a 73-year-old female. It is characterized by a dark-yellowish plaque present on the periumbilical area of the lower abdomen. The plaque is well-demarcated and its surface is grooved or fissured, atrophic and verrucoid. This localized lesion of the periumbilical area occurs in a multiparous woman with no history of hereditary systemic pseudoxanthoma elasticum.
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Key Words: Multiparous women Periumbilical peudoxanthoma elasticum

Pseudoxanthoma elasticum is a heritable, systemic disorder of connective tissue characterized by degeneration of elastic tissue mainly affecting the skin, eyes and cardiovascular systems.[7] Recently there have been reports of a localized, acquired form of cutaneous pseudoxanthoma elasticum. Localized skin lesions occur predominantly in middle-aged, multiparous, obese black women.[1-3] The lesions are typically located superior to the umbilicus and are associated with transepidermal elimination.[1-6] The acquired form of the disease is distinguished from the usual pseudoxanthoma elasticum by a negative family history, localized cutaneous lesions, late onset, and an absence of a angioid streak or other expected systemic manifestations. However, skin lesions are histologically similar to those seen in the inherited type of the disease.

To our knowledge, a few cases of this condition have been reported in the English literature[1-5, 9] and we could not find any cases in the Korean literature. We report a case of pseudoxanthoma elasticum limited to the periumbilical area occuring in a non-black multiparous woman with no history of hereditary systemic pseudoxanthoma elasticum.

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REPORT OF A CASE

A 73-year-old multiparous, gravida 9, woman had a slightly elevated verrucous plaque on the periumbilical area for about 50 years. The plaque was first noticed as a walnut-sized, hyperpigmented periumbilical plaque after her third delivery. She had been healthy, except for this skin lesion, which had been growing slowly during her multiple pregnancies. She did not experience any subjective symptoms. She had no family history of similar lesions.

On physical examination a dark-yellowish keratotic plaque(20 x 15 cm) on the periumbilical area of the lower abdomen was observed. It was well-demarcated and reticulated with an irregular border. The lesion had a firm, verrucous surface with individual confluented keratotic papules(Fig. 1). No angioid streaks were seen on ophthalmologic examination. The patients blood pressure was 130/90 mm Hg and findings from routine laboratory studies were within normal limits.

A routine histologic section from the periumbil-

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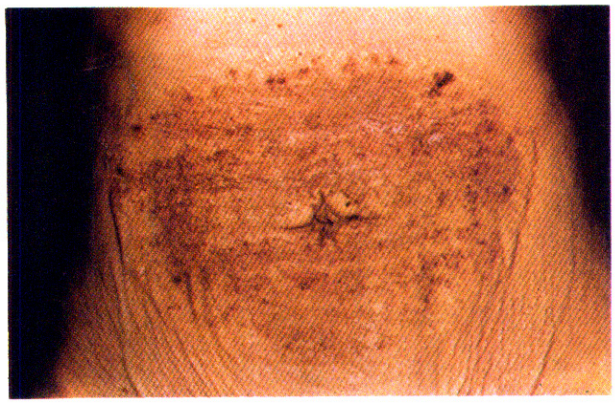

Fig. 1. Well-circumscribed verrucous periumbilical plaque.

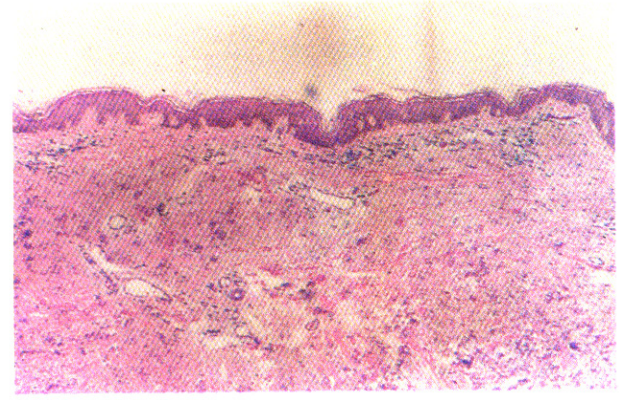

Fig. 2. Short and irregularly clumped fibers, which are basophilic stained, in the middle and lower dermis (H & E stain, × 100).

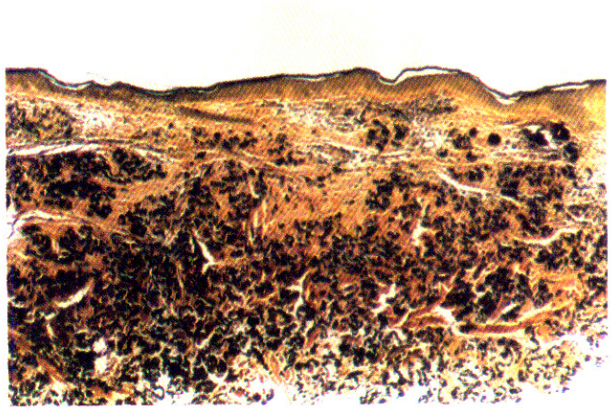

Fig. 3. Rigid basophilic fibers and granular debris were positive in Verhoeff-van Gieson stain (× 100).

ical plaque was stained with hematoxylin-eosin and Verhoeff-van Gieson. Histopathologic examination of the hematoxylin-eosin stained specimen showed altered elastic fibers in the middle and lower dermis. The fibers had a well-defined margin of basophilia(Fig. 2). The verhoeff's stained section showed these elastic fibers to be short, thickened and clumped(Fig. 3).

DISCUSSION

Pseudoxanthoma elasticum (PXE) is a familial disease characterized by widespread degeneration of elastic tissue involving the skin, eyes, gastrointestinal tract, and blood vessels.[7] It can be inherited as an autosomal dominant or recessive trait.[8]

In recent years there have been reports of a localized, acquired form of pseudoxanthoma elasticum.[1-5] This localized disorder characteristically occurs as a well-defined periumbilical plaque in multiparous, obese, middle aged black women.

The etiology of this localized form of PXE is unclear. It has been proposed that this localized form of PXE is unrelated to hereditary PXE.[1,5] Hicks et al suggested that localized PXE is an acquired lesion resulting from the effects of obseity and multiple pregnancy on elastic tissue. The traumatic effect of repeated pregnancies on the elastic tissue of the abdominal area may have determined the localization of lesion and may contribute to the development of disorder. In addition obesity might accelerate the disease process. Neldner et al[5] reported a patient with localized acquired cutaneous PXE who suffered from chronic active hepatitis with ascites. They hypothesize that localized PXE is acquired lesion after abdominal distension from ascites.

In our case the patient was a multiparous woman whose lesion was located in the periumbilical region. In a biopsy specimen the histologic changes characteristic of PXE were noted. However, there were no signs of hereditary systemic pseudoxanthoma elasticum. Therefore, our case supports the idea that repeated local cutaneous trauma leads to the development of periumbilical PXE.

There are other possible causes of periumbilical PXE. The threshold of trauma related change in elastic fibers may differ for people of black and light skin color groups. Therefore, genetic differences cannot be excluded.[1] Some patients in reported cases of localized PXE were found to have hypertension[1,3,6] and an angioid streak.[3,6] Recently

Goldstein and Lesher[9] presented a case of periumbilical PXE with systemic manifestations of the eyes and cardiovascular system. They suggested that patients with periumbilical pseudoxanthoma elasticum have, at a minimum, a full physical examination with emphasis on the cardiovascular system, and a complete ophthalmologic evaluation. Our patient had no angioid streaks, her blood pressure was within the normal range, and other findings from evaluation about cardiovascular system evaluation showed all findings within normal limits.

Lund and Gilbert reviewed the reported histologic findings of all previous cases of PXE associated with elastosis perforans serpiginosa(EPS), and added a report of a case of periumbilical perforating pseudoxanthoma elasticum.[6] They emphasized the histologic differences between elastic fibers in PXE and EPS. Fibers in cases of EPS are generally straight and are primarily located in the upper reticular dermis and in the papillary dermis. In contrast, in cases of perforating PXE fibers are short, fragmented, basophilic, and calcified and are located in the middle portion of the reticular dermis. Calcification of elastic tissue in cases of periumbilical PXE is easily demonstrable with Von Kossa stain. However, elastic tissue calcification is not easily identifiable in cases of EPS.

Perforation and transepidermal elimination of altered elastic fibers, as a secondary phenomenon, has been previously reported in cases of periumbilical PXE. Transepidermal elimination was not present in our case. However, if we had taken a biopsy specimen from the keratotic papules we might have found some evidence of transepidermal elimination.

In conclusion, we report a case of localized periumbilical PXE in a multiparous 73-year-old female. In our opinion repeated cutaneous trauma during multiple pregnancy contributed to the development of periumbilical PXE in this case.

None of our findings were specific for hereditary systemic PXE. Therefore, our case supports the suggestion by Hicks et al and others[5] that this periumbilical localized form of PXE is an acquired lesion.

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