Periumbilical Perforating Pseudoxanthoma Elasticum

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Periumbilical perforating pseudoxanthoma elasticum (PPPXE) is a localized acquired disorder found most frequently in obese, multiparous, middle-aged women. It is characterized clinically by yellowish, lax, well-circumscribed, reticulated or cobblestoned patches or plaques in the periumbilical region. Multiparity, obesity, massive ascites, and abdominal surgery are thought to the initiating factors.

There is controversy about the etiology of PPPXE. Some authors have classified it as a separate entity from hereditary pseudoxanthoma elasticum (PXE), while others speculate that this condition merely represents a variable expression of PXE in which systemic associations are likely.

We report a case of periumbilical perforating pseudoxanthoma elasticum associated with a clinical PXE lesion on the anterior neck. (Ann Dermatol 11(3) 185~188, 1999).

Key Words : Periumbilical perforating pseudoxanthoma elasticum, Pseudoxanthoma elasticum

Periumbilical perforating pseudoxanthoma elasticum (PPPXE) represents a distinct clinicopathological entity that occurs most frequently in obese, middle-aged, multiparous black women. Patients typically presented have a slowly-enlarging hyperpigmented, periumbilical plaque. The lesion is well demarcated with serpiginous borders. It is often flat centrally with discrete and coalescent keratotic papules scattered at the periphery to give a verrucous appearance[1-7].

As far as we know, only 2 cases of periumbilical pseudoxanthoma elasticum have been reported in the Korean dermatological literature[8,9]. We report a case of a patient with periumbilical perforating pseudoxanthoma elasticum occurring in a multiparous, obese woman.

CASE REPORT

A 71-year-old, multiparous, gravida 9, obese woman had a gradually enlarging, hyperpigmented plaque on her abdomen. About 3 years previously, several brownish papules had developed on the upper part of her umbilicus. They had coalesced into a hyperpigmented atrophic plaque and slowly enlarged. The lesion was asymptomatic. She denied a history of trauma to the area or contact with any irritants. No treatment for this lesion was administered.

She had a 20-year history of diabetes mellitus and hypertension, and a 5-year history of congestive heart failure. She had also had hepatitis C, liver cirrhosis, and atrial fibrillation for 5 months. She had undergone a cholecystectomy 15 years previously. She had no family history of similar skin lesions.

A physical examination revealed an obese woman with a well-demarcated, hyperpigmented, atrophic, 3 × 2 cm plaque with an elevated verrucous border on the superior side of her umbilicus(Fig. 1). On the anterior neck, multiple slightly yellowish papules were also found. An ophthalmological examination showed no evidence of angioid streaks.

Laboratory studies revealed the following results: AST, 54 U/L: hemoglobin A1c, 7.7%. A chest radiography revealed cardiomegaly and pleural effusion. Abdominal sonography demonstrated a fatty liver and a small-sized kidney. An EKG showed atrial

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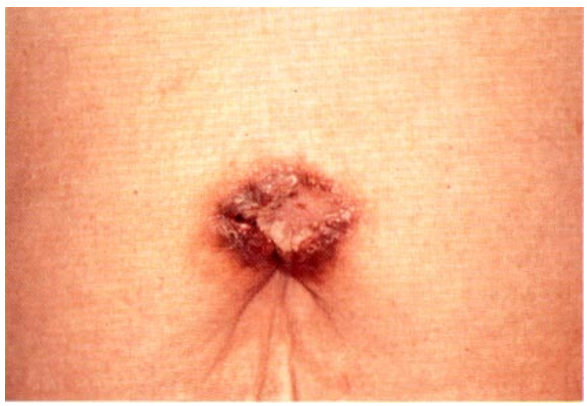

Fig. 1. Well-demarcated hyperpigmented atrophic plaque on the right upper quadrant of the abdomen.

fibrillation. An echocardiogram showed decreased left ventricular function and a small amount of pericardial effusion.

The excisional biopsy specimen revealed altered elastic fibers throughout the dermis that were short, thick, irregularly clumped, and basophilic(Fig. 2). These calcified and curled elastic fibers were being extruded to the skin surface. Verhoeff-van Gieson staining for elastic tissue confirmed this finding. The von Kossa stain for calcium was reactive(Fig. 3).

DISCUSSION

Pseudoxanthoma elasticum (PXE) is an inherited disorder of connective tissue characterized by de-

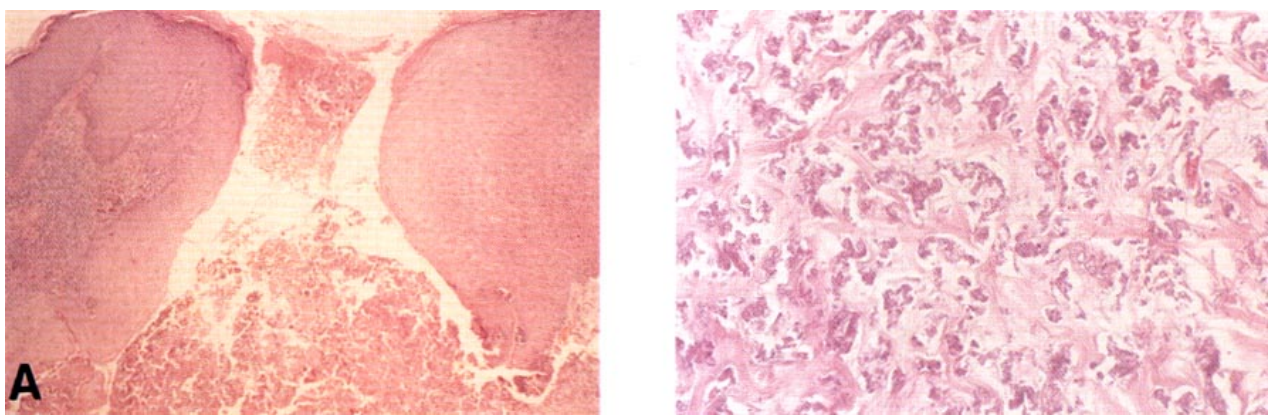

Fig. 2. (A) Transepidermal elimination of altered elastic fibers through the channel lined by acanthotic epidermis (H & E stain ×40). (B) Higher magnification reveals that numerous altered elastic fibers in the reticular dermis are short, thick, irregularly clumped, and basophilic. (H & E stain ×200).

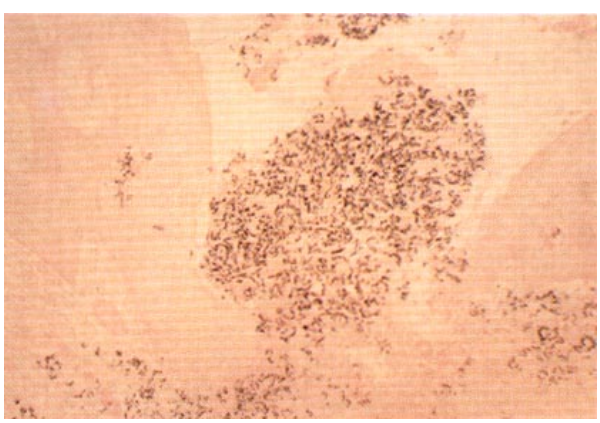

Fig. 3. Calcified elastic fibers extruding to the skin surface (von Kossa stain ×40).

generation and calcification of elastic fibers in the ocular, cutaneous, and cardiovascular systems. Notable complications include angioid streaks, intermittent claudication, angina, myocardial infarction, hypertension, cardiovascular accidents, and hemorrhagic disorders of the gastrointestinal and genitourinary tract[1].

Perforating pseudoxanthoma elasticum is a rare disorder. The localized lesion occurs predominantly in middle-aged, multiparous, obese black women[1,3,5]. Characteristially, it is an abdominal plaque located superior to the umbilicus[1,2,3,5]. The plaque is a well-demarcated and hyperpigmented lesion that may slowly increase in size[1]. Its surface has been described as reticulated, grooved, and atrophic or fissured

and verrucoid[1-3]. Frequently, hyperkeratotic papules are contained at the periphery of the plaque. Compression of the border of a plaque or a papule may produce a purulent discharge[2]. Multiparity, obesity, massive ascites, and abdominal surgery are thought to the initiating factors[7]. In our opinion, repeated cutaneous trauma of pregnancy and obesity may have contributed to the development of PPPXE in this case. PPPXE is distinguished from the usual PXE by a negative family history, localized cutaneous lesions, late onset, and an absence of angioid streaks or other expected systemic manifestations[3].

In the past, many cases of perforating pseudoxanthoma elasticum with perforating lesions were referred to as pseudoxanthoma elasticum with elastosis perforans serpiginosa[4]. Lund and Gilbert[2] analyzed seven of these cases, however, and found that, in fact, only one case clearly established this coexistence. The other cases were considered most consistent with perforating pseudoxanthoma elasticum, showing the distinct clinicopathological features of this entity. Lund and Gilbert[2] emphasize the differentiating histological features of these diverse disorders. Elastosis perforans serpiginosa consists of the transepidermal elimination of abnormally large, abundant eosinophilic (noncalcified) elastic fibers. These fibers are generally straight in configuration and are located primarily in the upper portion of the reticular dermis and in the papillary dermis. In contrast, perforating pseudoxanthoma elasticum is characterized by the transepidermal elimination of altered basophilic calcified elastic fibers, which are short, fragmented, and curled and are located primarily in the middle portion of the reticular dermis. On the basis of these criteria, our case represents a typical example of periumbilical perforating pseudoxanthoma elasticum.

In the Korean dermatological literature, only 2 cases of periumbilical pseudoxanthoma elasticum have been reported[8,9]. Kim et al.[8] reported a 73-year-old multiparous woman who had periumbilical pseudoxanthoma elasticum without definite transepidermal elimination of altered elastic fibers. Chang et al.[9] reported a 72-year-old multiparous woman who had periumbilical perforating pseudoxanthoma elasticum without involvement of other flexural areas and systemic manifestations of pseudoxanthoma elasticum.

There is controversy about the etiology of PPPXE. In some reported cases, perforating PXE is clearly another manifestation of the hereditary systemic form of PXE, with transepidermal elimination occurring secondarily[10-14]. These patients demonstrate many of the other clinical features of hereditary (systemic) PXE, including angioid streaks and flexural lesions. However, in other cases, perforating PXE occurs as a localized lesion without the associated clinical features of hereditary PXE. This localized cutaneous disorder characteristically occurs as a well-defined, hyperpigmented periumbilical plaque in multiparous, obese black women. Some authors suggest that localized PXE be considered a separate entity from hereditary PXE because of its distinct clinical features and its presumed lack of association with systemic disease[5,6]. They hypothesized that localized PXE is an acquired lesion realized locally after cutaneous trauma from obesity, multiple pregnancies, and ascites in predisposed persons. We suggest that PPPXE is a variable expression of the hereditary PXE because our patient has clinical PXE lesions on the anterior neck.

In summary, a case of periumbilical perforating pseudoxanthoma elasticum associated with clinical pseudoxanthoma elasticum lesion on the anterior neck is presented.

REFERENCES

1. Hicks J, Carpenter CL, Reed RJ. Periumbilical perforating pseudoxanthoma elasticum. Arch Dermatol 115:300-303, 1979.
2. Lund HZ, Gilbert CF. Perforating pseudoxanthoma elasticum: Its distinction from elastosis perforans sergiginosa. Arch Pathol Lab Med 100:544-546, 1976.
3. Schwartz RA, Richfield DF. Pseudoxanthoma elasticum with transepidermal elimination. Arch Dermatol 114:279-280, 1978.
4. Kazakis AM, Parish WR. Periumbilical perforating pseudoxanthoma elasticum. J Am Acad Dermatol 19:384-388, 1988.
5. Premalatha S, Yesudian P, Thambiah AS. Periumbilical pseudoxanthoma elasticum with transepidermal elimination. Int J Dermatol 21:604-605, 1982.
6. Somasundaram V, Premalatha S, Rao RN, Abdul Razack EM, Zahra A. Periumbilical perforating pseudoxanthoma elasticum. Int J Dermatol 26:536-537, 1987.
7. Pruzan D, Rabbin PE, Heilman ER. Periumbilical perforating pseudoxanthoma elasticum. J Am Acad

Dermatol 26:642-644, 1992.
8. Kim YH, Yoon JS, Lee JH, Park JK. Periumbilical pseudoxanthoma elasticum. Ann Dermatol 6:49-51, 1994.
9. Chang KY, Whang KK, Myung KB, Hahm JH. A case of perforating pseudoxanthoma elasticum. Kor J Dermatol 36:160-163, 1998.
10. Smith EW, Malak JA, Goodman RM, McKusick VA. Reactive perforating elastosis: a feature of certain genetic disorders. Bull Johns Hopkins Hosp Med J 111:235-251, 1962.
11. Schutt DA. Pseudoxanthoma elasticum and elastosis perforans sepiginosa. Arch Dermatol 91:151-152, 1965.
12. Bos WH. Pseudoxanthoma elasticum associated with perforating elastoma. Dermatologica 136:296-297, 1968.
13. Caro I, Sher MA, Rippey JJ. Pseudoxanthoma elasticum and elastosis perforans serpiginosa. Dermatologica 150:36-42, 1975.
14. Graham JH, Hunter GA. Perforating pseudoxanthoma elasticum. Arch Dermatol 112:1781, 1976.