A Case of Periumbilical Perforating Pseudoxanthoma Elasticum

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Periumbilical perforating pseudoxanthoma elasticum is a rare acquired disorder that usually occurs in obese, middle-aged, multiparous women. It is considered as a separate entity from the hereditary pseudoxanthoma elasticum due to lack of hereditary transmission and association with systemic disease. A 70-year-old multiparous woman presented with a 2-year history of a gradually enlarging, yellowish ulcerated plaque in the periumbilical region. The biopsy specimen showed irregularly altered elastic fibers with encrusted calcium salts that underwent transepidermal elimination. We report a rare case of periumbilical perforating pseudoxanthoma elasticum with no signs of hereditary pseudoxanthoma elasticum.

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Key Words: Perforating pseudoxanthoma elasticum, Periumbilical

INTRODUCTION

Periumbilical perforating pseudoxanthoma elasticum (PPPXE), first described by Hicks et al. in 1979, is a rare acquired disorder of connective tissue[1-3]. This localized cutaneous disorder characteristically presents as a well-defined, hyperpigmented periumbilical plaque in multiparous, obese, black women[2,3]. It is suggested that repeated traumas of pregnancy, obesity, or abdominal surgery promote localized degeneration of elastic fibers[2-4]. Herein, we report an uncommon case of PPPXE which occurred in an obese, multiparous woman.

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CASE REPORT

A 70-year-old obese multiparous woman presented with a 2-year history of gradually enlarging ulcerated plaques on her periumbilical region. On physical examination of the skin lesion, there were two discrete, 0.7 × 0.5 cm and 1.0 × 0.8 cm, yellowish to brownish cobblestoned plaques with central ulceration in the supraumbilical area (Fig. 1A). She had not any history of abdominal surgery or trauma on the abdomen but had a history of 5 vaginal deliveries. Her other medical history was unremarkable. On examination of her body, there were no abnormal findings, including in her cardiovascular and ophthalmologic systems. Laboratory evaluations including complete blood count, blood chemistry analysis and urinalysis were normal. A punch biopsy was taken from the yellowish ulcerated plaque on the supraumbilical area. Histopathologically, the epidermis showed a keratin-filled crater. The upper dermis demonstrated irregularly altered elastic fibers with encrusted calcium salts extruded to the surface through the epidermis in a wide channel, that is, transepidermal elimination (Fig. 1B). The Verhoeff-van Gieson stain showed numerous clumped, irregularly thickened elastic fibers in the dermis (Fig. 1C). The von Kossa stain revealed fragmented and

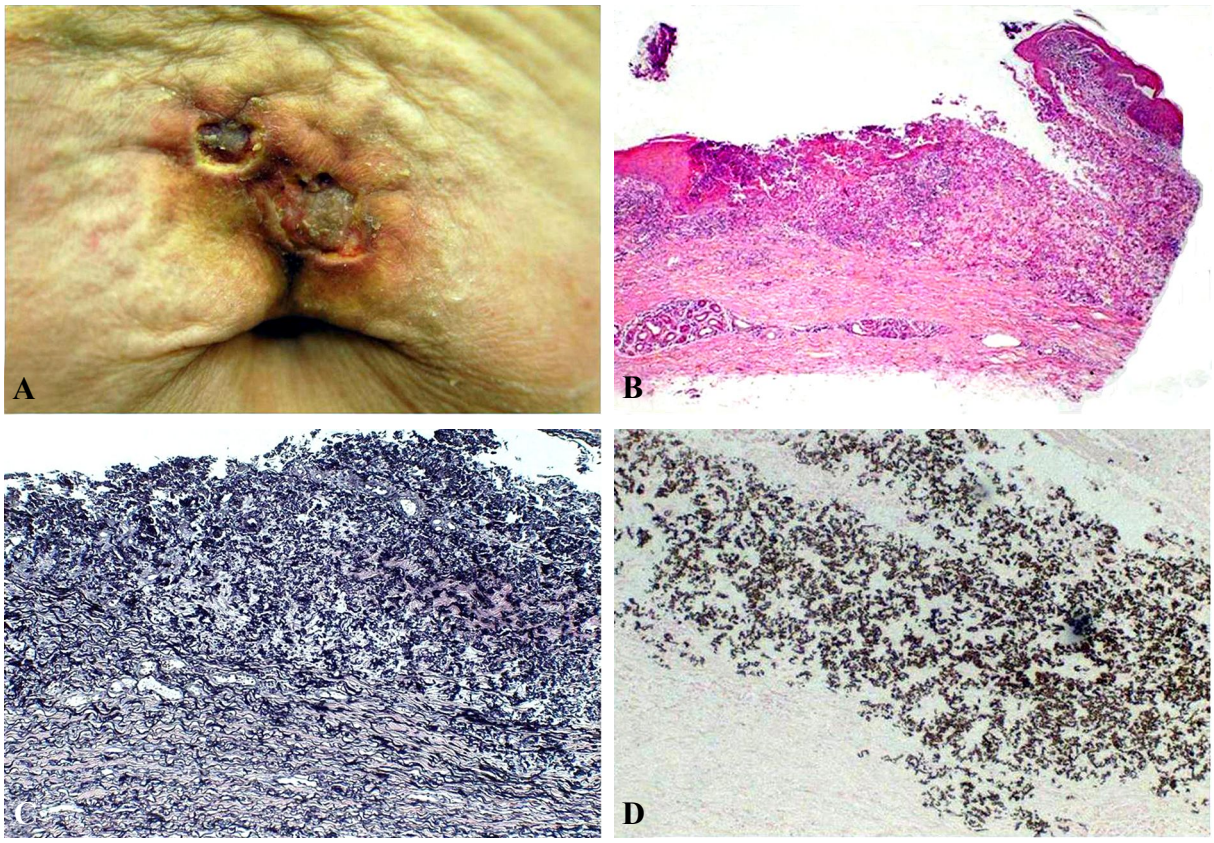

Fig. 1. (A) Ill-defined yellowish to brownish cobblestoned plaques with central ulceration were noted in the supraumbilical region. (B) Transepidermal elimination of altered elastic fibers were seen in the dermis (H & E, × 40). (C) Clumped, irregularly thickened elastic fibers were seen in the dermis (Verhoeff-van Gieson stain, × 100). (D) Calcified and fragmented elastic fibers were demonstrated (von Kossa stain, × 100).

calcified elastic fibers in the dermis (Fig. 1D). These clinical and histopathologic findings were consistent with those of PPPXE. Because there was no known effective therapy, we assured the patient that this was a benign disorder and decided to keep her on close observation. There has not been any progression or change of the lesion during the 8-month follow-up period.

DISCUSSION

PPPXE, also referred as perforating calcific elastosis, is an acquired disorder of connective tissue, characterized by localized degeneration and calcification of elastic fibers. It occurs predominantly in middle-aged, multiparous, obese, black women[1-3]. Patients typically present as an occasionally pruritic, slowly enlarging, hyperpigmented plaque with discrete keratotic papules at the periphery of the lesion. The skin lesion usually locates in the periumbilical region, especially in the supraumbilical area. It is difficult to explain the periumbilical localization of PPPXE, however, it is suggested that the repeated trauma of pregnancy, obesity, abdominal surgery, or massive ascites may act as the initiating factors and result in localized elastic fiber degeneration[5,6].

Clinically, hereditary pseudoxanthoma elasticum (PXE) should be differentiated. Whereas PXE is an inherited disorder of elastic tissue in the ocular, cutaneous and cardiovascular systems as a systemic process, PPPXE is a non-inherited, localized cutaneous disease, which can be distinguished by lack of family history, late onset and absence of other systemic manifestations[2,7,8]. Both PXE and PPPXE are characterized histologically by degeneration of elastic fibers with deposition of calcium in the mid-dermis. However, transepidermal elimination of

these altered elastic fibers is only found in PPPXE and it rarely occurs in PXE[5].

An important histopathological entity in the differential diagnosis is elastosis perforans serpiginosa, which consists of the transepidermal elimination of abnormally large, eosinophilic noncalcified elastic fibers in the upper dermis. In contrast, PPPXE is characterized by the transepidermal elimination of short, fragmented, basophilic calcified elastic fibers located primarily in the mid-dermis. With the von Kossa stain, calcification of elastic tissue in PPPXE is easily demonstrable[2-4,9].

While there are no effective therapeutic options presently available, patients can be reassured that the eruption is a benign process limited to the skin[7]. Up to now, 12 cases of PPPXE have been reported in English literature and 4 cases have been found in Korea[7-8]. In summary, a case of PPPXE with no signs of hereditary pseudoxanthoma elasticum is presented.

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