Familial Atrophoderma Vermiculata Associated with Epidermal Cysts

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ATrophoderma vermiculata (AV) is a rare disease which normally occurs on the face and cheeks. It consists of numerous, closely-crowded, atrophic pits, with a bilateral distribution, producing a reticulate appearance. Familial occurrence has been noted, as well as in combination with other congenital anomalies.

Two patients, a proband and his maternal aunt, presented with the characteristic changes of facial multiple pits. A clinical diagnosis of AV was determined. In addition, multiple subcutaneous nodules were noted. Four nodules were removed and identified as epidermal cysts. A family history revealed that the proband's mother, maternal grandfather, maternal uncle, two maternal aunts and two older sisters had also suffered from similar facial skin lesions and multiple subcutaneous nodules. An autosomal dominant mode of transmission was indicative from this family tree. We report a rare case of AV associated with multiple epidermal cysts found in eight members of a family. (Ann Dermatol 17(2) 102-105, 2005)

Key Words: Atrophoderma vermiculata (AV), Autosomal dominant, Epidermal cysts

INTRODUCTION

Atrophoderma vermiculata (AV) refers to a form of keratosis pilaris atrophicans, in which the cheeks are predominantly involved. AV was first described by Unna ¹ in 1894 under the name of ulerythema acneiforme. Subsequently, numerous terms have been used for this condition, including acne vermiculante, atrophoderma reticulata symmetrical faciei, folliculitis ulerythematosa reticulata, folliculitis atrophicans reticulata and honeycomb atrophy.² ³ ⁴ AV usually presents as an isolated skin defect with no associated medical conditions. Rarely it may be associated with epidermal cysts, folliculitis decalvans, leukokeratosis oris, Down's syndrome, a congenital heart disorder or neurofibromatosis.⁵ ⁶ To our knowledge, only one case of AV, which occurred in a patient with congenital heart disease, has been reported in the Korean literature.⁶ Herein, we present two cases of AV associated with epidermal cysts in a nephew and a maternal aunt.

CASE REPORT

Case 1
A 26-year-old man presented with irregular, atrophic scars on the face. The lesions had first been noted 10 years earlier, and had progressively worsened over the years. There was no prior history of papules, acne or trauma. During the same period of time, subcutaneous nodules on the chest, arms and groin had appeared. Physical examination showed marked, pitted scarring of the forehead, cheeks, nose and preauricular areas. Some were confluent with a cribiform appearance, while others were solitary (Fig. 1). There was no involvement of
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disorder of the face, which had been present since the age of 15. She denied preceding inflammation. She had no history of hypohidrosis or basal cell carcinoma. Physical examination revealed multiple pits on the face (Fig. 3) and multiple subcutaneous nodules, 1-3 cm in diameter on the trunk, neck, axillae and groin. Two nodules were removed and histologically identified as epidermal cysts.

A family history revealed the proband (case 1), proband's mother, two maternal aunts (including case 2), maternal grandfather, maternal uncle and two older sisters also suffered from similar facial skin lesions and multiple subcutaneous nodules on the trunk and extremities. An autosomal dominant mode of transmission was indicated from this family tree. A pedigree is shown in Fig. 4. No other familial disorders were known.

**DISCUSSION**

AV is a rare skin condition, which is classified as one of the keratosis pilaris atrophicans group of disorders, the others being lerythema oophyrogenes and keratosis pilaris decalvans. Most cases are sporadic, but autosomal dominant inheritance has been described. Generally, AV begins in early childhood with erythematous follicular papules on the cheeks. These papules then go on to develop
We report on two patients, a proband and his maternal aunt, who presented with the characteristic changes of facial multiple pits. They also had multiple, 1-3 cm sized, subcutaneous nodules on the trunk, axillae, groin and extremities. Four nodules were removed and identified as epidermal cysts. In addition, seven members of the proband’s family also had the characteristic facial changes of AV and multiple subcutaneous nodules. In previously reported cases of AV, although epidermal cysts or milia were rarely noted, almost all these cysts were small and limited to the facial lesions of AV.

The main interest of our cases, besides their rarity, is the observation of the association of AV with epidermal cysts. Whether the simultaneous occurrence of these diseases is coincidental or due to related pathogenic mechanisms remains to be seen. The etiology of AV is unknown, but the primary defects appear to be an abnormal keratinization of the pilosebaceous units. It is widely assumed that epidermal cysts are related to the pilosebaceous unit. In addition, eight members of a family all had multiple subcutaneous nodules on the trunk and extremities, as well as the characteristic facial change associated with AV. Although only four of subcutaneous nodules were examined, it is believed that the remaining nodules may have also been epidermal cysts. These cases support a theory that AV and epidermal cysts may have a similar pathogenic mechanism.

In summary, we present two cases of AV associated with epidermal cysts in a nephew and a maternal aunt. Although these findings may be coincidental, we believe these cases highlight the association of AV with epidermal cysts. Further investigations are needed to demonstrate the pathogenesis.

REFERENCES