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Cornoid lamellae associated with follicular infundibulum and acrosyringium
in porokeratosis
(毛包漏斗部と表皮内汗管に関連した汗孔角化症のcornoid lamellae)

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ORIGINAL ARTICLE

Cornoid lamellae associated with follicular infundibulum and acrosyringium in porokeratosis.

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Abstract

Skin lesions of porokeratosis consist of an atrophic center bordered by a peripheral grooved keratotic ridge that corresponds histopathologically to the cornoid lamella. Originally porokeratosis was named based on the assumption that the columns of parakeratosis emerge from the ostia of eccrine ducts. Despite this, it is generally accepted that the cornoid lamellae are rarely related to the acrosyringium. We recently encountered two cases of porokeratosis, where the cornoid lamellae were related to the follicular infundibulums. Therefore we analyzed the location of the cornoid lamellae in 86 lesions of porokeratosis from 73 patients from our archives. We found that many cornoid lamellae are also detected at follicular infundibulum and acrosyringium. The existence of so many cornoid lamellae at follicular infundibulum and acrosyringium inside the plaques cannot be explained by a pure coincidence and may be more than fortuitous.

Key words: porokeratosis, cornoid lamella, follicular infundibulum, acrosyringium

Introduction

Porokeratosis is inherited in an autosomal dominant pattern, but many of them are sporadic. Six types of porokeratosis are distinguished, which are classic porokeratosis (Mibelli), linear porokeratosis, localized porokeratosis, superficial disseminated porokeratosis, disseminated superficial actinic porokeratosis (DSAP) and porokeratosis palmaris et plantaris. It is characterized by a distinct peripheral keratotic ridge that corresponds histopathologically to the cornoid lamella. Originally porokeratosis was named based on the assumption that the columns of parakeratosis emerge from the ostia of eccrine ducts. Despite this, it is generally accepted that the cornoid lamellae are rarely related to the acrosyringium. We recently encountered with two cases of porokeratosis, where the cornoid lamellae were related to the follicular infundibulums. Besides the detailed case reports of these two cases we investigated the exact location of the cornoid lamellae in 86 cases of porokeratosis from 73 patients which were diagnosed histopathologically at the Department of Dermatology, Asahikawa Medical College.

Case1

An 85-year-old Japanese man noted an annular hyperkeratotic plaque on his right cheek approximately 1 year ago. Since the plaque gradually enlarged, he visited our clinic in August 2005 (Fig.1a). The histopathology of an incisional biopsy disclosed the typical cornoid lamella with a

thin column of parakeratotic cells, decreased underlying granular zone, vacuolated and dyskeratotic cells in the spinous layer of the epidermis of the ridge (Fig.1 b,c). In addition, there were parakeratotic columns associated with two follicular infundibulums in the central part of the plaque (Fig.1 b,d). We diagnosed this case as the classic porokeratosis (Mibelli) with follicular cornoid lamellae.

Case2

A 70-year-old Japanese man noted itchy light brownish hyperkeratotic plaques on the four extremities approximately 4 years ago. The first histopathological examination of an excisional biopsy in June 2000 showed two typical cornoid lamellae which correspond clinically to the peripheral grooved keratotic ridge (Fig.2 a,b,c). There were parakeratotic columns associated with two follicular infundibulums in the center (Fig.2 a,d). We diagnosed this case as the superficial disseminated type porokeratosis with cornoid lamellae associated with follicular infundibulums. The number of plaques had increased, and he visited us again (Fig.3a,b). The second histopathological examination of an incisional biopsy in June 2006 showed typical cornoid lamella on the ridge (Fig.3c,d), and other cornoid lamellae associated with follicular infundibulum in the central part of the plaque (Fig.3e).

Because we encountered two cases of porokeratosis with the cornoid lamellae related to the

follicular infundibulums, we investigated the location of the cornoid lamella in porokeratosis.

Methods

A total of 86 lesions of porokeratosis from 73 patients were retrospectively collected from the archives of Department of Dermatology, Asahikawa-Medical College from 1978 to 2006. All cases had been previously fixed in formalin and embedded in paraffin. Hematoxylin and eosin stained sections were reviewed from each case. Besides the ordinary 2 μ m- section cut at the maximal long length-wise, we prepared further up to 50 consecutive sections for lesions we could not find cornoid lamellae associated with hair follicles or sweat ducts. The definition of the cornoid lamella is a thin column of parakeratotic cells with an absent or decreased underlying granular zone and vacuolated or dyskeratotic cells in the spinous layer. We excluded the follicular pluggings which have hyperkeratotic invaginations of epidermis without parakeratotic columns and dyskeratotic cells in the spinous layer.

Results

Out of 86 lesions we found 12 lesions of porokeratosis from 11 patients related to follicular infundibulums, including Case1 and Case2 (Fig.4,1~6), and 5 lesions of porokeratosis from 5 patients associated with acrosyringiums in the first section (Fig.5,1~3). In the consecutive sections,

21 lesions of porokeratosis from 21 patients related to follicular infundibulums (Fig.6,7~12), 16 lesions of porokeratosis from 16 patients associated with acrosyringiums were detected (Fig.7,4~9). Thus hair follicle- or acrosyringium-related cornoid lamellae were shown to be observed much more frequently than our expectation. The location of cornoid lamellae were constantly the lower parts of follicular infundibulum and acrosyringium. Although the clinical subtypes were examined (Table 1 and 2), we could not find a clear correlation between the clinical subtypes and the association of cornoid lamellae with cutaneous appendages.

Discussion

In 1893, Mibelli¹⁾ named porokeratosis based on the assumption that the columns of parakeratosis emerge from the ostia of eccrine ducts. In the same year, Respighi²⁾ emphasized the plugging of the follicular infundibulum and acrosyringium of porokeratosis. Despite this, it is generally believed that the cornoid lamellae are rarely related to the follicular infundibulum or acrosyringium³⁾.

Cornoid lamellae are typically formed at the peripheral ridge of the porokeratotic plaques. In our study, we found many of the cornoid lamellae not only at the peripheral ridges, but also at the follicular infundibulum and acrosyringium inside the plaques. Koyama and Matunaka⁴⁾ reported a 80-year-old man with generalized confluent annular and circinate eruptions, porokeratosis Mibelli with multiple Bowen disease. Light and electron microscopic examination showed follicular cornoid

lamellae and examination using a videomicroscope revealed follicular spinous vellus hair-like processes. This is the first report which detected cornoid lamella associated with follicular infundibulum. The reason for the frequent occurrence of cornoid lamellae at the follicular infundibulum and acrosyringium is not yet clear, however, it can not be explained by a pure coincidence. Abnormal epidermal clones are located at the base of the parakeratotic columns of the porokeratosis⁵⁾. Cornoid lamellae are usually formed at the boundary between normal epidermal cells and abnormal epidermal cell clones at the ridge of the plaque. Because the location of putative stem cells of epidermal appendages is assumed to be distinct from interfollicular epidermal stem cells, abnormal epidermal cell clones of porokeratosis might form parakeratotic columns at the boundary of epidermal appendages in the presence of intact stem cells of hair follicles and sweat ducts in a similar manner. The location of cornoid lamellae, i.e. the lower parts of follicular infundibulum and acrosyringium are consistent with this assumption.

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Figure legends

Fig1 (a) A 1cm sized annular plaque with peripheral keratotic ridge on the right cheek.

(b) Histopathological examination showed the typical cornoid lamella on the left side (arrow)

(Hematoxylin-eosin stain;40x) .

(c) A closer view of cornoid lamella on the left portion of Fig (b). A thin columns of parakeratotic cells with decreased underlying granular zone and vacuolated and dyskeratotic cells in the spinous layer are shown. (Hematoxylin-eosin stain;400x) .

(d) A closer view of the right hair follicle of Fig (b).Parakeratotic columns are associated with follicular infundibulums (arrows) (Hematoxylin-eosin stain;200x) .

Fig2 (a) Histopathological finding of the excisional biopsy (Hematoxylin-eosin stain;12.5x) .

(b)A closer view of cornoid lamella on the left portion (Hematoxylin-eosin stain;40x) .

(c) A closer view of cornoid lamella on the right portion (Hematoxylin-eosin stain;100x) .

(d) A closer view of two columns of parakeratotic cornoid lamella in the center of the plaque.

Cornoid lamellae are seen at the follicular infundibulums (arrows) (Hematoxylin-eosin stain;400x) .

Fig3 (a) The clinical feature of superficial disseminated porokeratosis on the leg.

(b) A closer view. Up to 1cm sized well-demarcated brownish plaques with follicular plugging.

(c) Histopathological finding of the incisional biopsy (Hematoxylin-eosin stain;20x) .

(d) A closer view of a cornoid lamella on the left portion (Hematoxylin-eosin stain;200x) .

(e) A closer view of cornoid lamella associated with follicular infundibulum on the right portion

(arrow) (Hematoxylin-eosin stain;400x) .

Fig4 (1~6) Cornoid lamellae associated with follicular infundibulums in the first sections.

Fig5 (1~3) Cornoid lamellae related with acrosyringiums in the first sections.

Fig6 (7~12) Cornoid lamellae associated with follicular infundibulums in the consecutive sections.

Fig7 (4~9) Cornoid lamellae related with acrosyringiums in the consecutive sections.

Table1 Clinical subtypes of porokeratosis and occurrence of cornoid lamellae at the follicular infundibulum.

Classic porokeratosis (Mibelli)	4/7	
Linear porokeratosis	2/3	
Localized porokeratosis	8/18	
Superficial disseminated porokeratosis	10 / 16	
Disseminated superficial actinic porokeratosis	8 / 25	
Porokeratosis palmaris et plantaris	0 / 1	
Unknown		2 / 2

Table2 Clinical subtypes of porokeratosis and occurrence of cornoid lamella at the acrosyringium.

Classic porokeratosis (Mibelli)	1 / 7	
Linear porokeratosis	1 / 3	
Localized porokeratosis	0 / 18	
Superficial disseminated porokeratosis	8 / 16	
Disseminated superficial actinic porokeratosis	11 / 25	
Porokeratosis palmaris et plantaris	0 / 1	
Unknown		0 / 2













