Syringocystadenoma Papilliferum
— Unusual Location and Electron Microscopic Study —

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We report a case of syringocystadenoma papilliferum which occurred in the groin since birth as a nodule, without any associated lesions. The direction of differentiation in syringocystadenoma papilliferum was studied through light and electron microscopic study.

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Syringocystadenoma papilliferum occurs as a solitary lesion appearing usually on the scalp but occasionally on the face or elsewhere.1 Usually these lesions are present at birth or appear during early childhood.2 Early lesions may begin as alopecic plaques, and at puberty, verrucous and papilomatous changes occur. Microscopically, there is a cystic invagination of the papillomatous epidermis. Villi lined by two rows of cells project into the resulting spore. Luminal cells are tall and columnar with evidence of secretory activity, and the basally located cells are small and cuboidal.1,2 Origin from either eccrine or apocrine structures has been supported in the literature,3,4 although many such lesions appear to be nearly of apocrine origin, others appear to have an eccrine derivation.

We describe a case of syringocystadenoma papilliferum which had been in the groin since birth, without a preexisting lesion. The origin of this tumor was studied with light and electron microscopic studies.

REPORT OF A CASE

A 76-year-old man was seen on August 10, 1989 in our department for evaluation of an asymptomatic nodule in the groin which had been present since birth. Examination revealed a thumb-tip sized skin-colored nodule which discharged yellowish fluid through a central pore when squeezed (Fig. 1).

Light microscopic study of the tumor showed invagination of the surface epithelium with the formation of projecting villous and papillary structures. These structures were deeply invaginated forming cystic spaces and numerous lumina (Fig. 2). The papilliferous walls were lined by a double cell layer comprising a thin outer layer of flat epithelial cells and an inner columnar or cuboidal cell layer. In other area, multiple layers of cells consisted of walls. The stroma was infiltrated by plasma cells, together with lymphocytes and neutrophils (Fig. 3). Large amounts of PAS-positive, diastase-resistant material throughout the luminal cell were present.

Electron microscopic study shows that the intracytoplasmic cavity was lined by numerous microvilli and filled with amorphous matter (Fig. 4). The cells constituting the intercellular canaliculus were interconnected by desmosomes. Some cells of the luminal wall appeared clear, while others were dark (Fig. 5). There were pinched-off villi with a discharge of small amounts of cell content into the lumen (Fig. 6).

DISCUSSION

Syringocystadenoma papilliferums are more common in women than in men.1,2 They usually appear as solitary ulcerated plaques or nodules. More than half occur on the scalp but they have been

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Fig. 1. Yellowish slightly turbid fluid discharging from nodule when squeezed.

Fig. 2. A cystic invagination extends downward from the epidermis. Numerous papillary projections extend into the lumen of the cystic invagination (H & E stain, x40).

Fig. 3. The papillary projections are lined by two rows or several rows of cells and an inflammatory infiltrate containing plasma cells, lymphocyte and neutrophil in stroma (H & E stain, x200).

Fig. 4. Intracytoplasmic cavity lined by numerous microvilli and filled with amorphous matter (x5000).

Fig. 5. Two densities of cells consist of luminal wall (x4000).

found on the face, neck, shoulder, girdle area, axillae, trunk, thigh, and the genital area. Usually these lesions are present at birth or appear during early childhood, but they often enlarge significantly at puberty, and their surface, previously smooth, may become elevated and verrucose at that time. However approximately 25 percent of syringocystadenoma papilliferum arise on the trunk and
the genital and inguinal regions during adolescence or adult life, without a preexisting lesion.\(^5\) Individual lesions present a variety of clinical appearances. They may be infiltrative plaques, papilomatous or verrucous lesions, or hyperkeratotic nodules with central keratotic plugs. Their color varies from shades of red to brown. The plaques frequently become granulomatous and crusted. The crust is composed of dried blood or a watery to thick mucoid secretion which exudes from the cystic cavities. A verrucous or bulbous elevation occasionally was seen to exude fluid through a pore presumably from a cyst. This fluid varied from watery and clear to thick and sometimes dark brown.\(^1\)

In the present case, a 76-year-old man had thumb-tip sized nodule in the right groin, which were present at birth. Yellowish, slightly turbid fluid was exuded through a pore when squeezed. The arising of this lesion on the groin since birth without a preexisting lesion is rare when we reviewed in the Korean literatures (Tabel 1).

Histologically, syringocystadenoma papilliferum consists of a cystic and papillary growth of epithelial elements projecting downward into the dermis and opening on to the skin surface through one or more orifices of varying width. The structure of the overlying epidermis is quite variable, in some cases consisting of flattened epithelium, while in others assuming the verrucous character sometimes seen in nevus sebaceous of Jadassohn. In any event, the stratified squamous epithelium extends for varying distances down into the orifice of the tumor.\(^9\) At about the level of the papillary dermis, there is an abrupt transition to epithelium resembling that of apocrine glands. Usually this consists of two layers of cells, the peripheral layer being low cuboidal, and the inner, columnar cells showing evidence of decapitation secretion. While two layers of cells are considered typical, areas can be found in most tumors in which the cavity is

<table>
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<tr>
<th>Year</th>
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<th>Age (yrs)</th>
<th>Sex</th>
<th>Age of onset (yrs)</th>
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<td>Birth</td>
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**Figure 6.** Pinched-off villi with discharge of a small amount of cell content into the lumen (×6000).
lined by only one layer of cells, or in other places by multiple layers of cells up to six or eight cells in thickness. In the columnar cells, PAS-positive, diastase resistant material as well as iron containing granules can be demonstrated histochemically.\textsuperscript{20,21}

The connective tissue stroma of the villous projections and of the surrounding dermis is fibrous and vascular. It is marked by a rich infiltration of inflammatory cells, notably a large number of plasma cells. Frequently, there are malformed sebaceous glands and hair structures in the lesions of syringocystadenoma papilliferum.\textsuperscript{2} In about one third of the cases, syringocystadenoma papilliferum is associated with a nevus sebaceous. In about 10% of the cases, a basal cell epithelioma develops, but this is noted only in lesions that also have a nevus sebaceous.\textsuperscript{1}

In our case, the histopathologic features are compatible with syringocystadenoma papilliferum not associated with other anomaly. The luminal cells lining the villi consist of two layers of cells in some areas and multiple layers of cells, up to five cells in thickness, in other places. These cells have given a PAS-positive, diastase resistant reaction.

The histogenesis of syringocystadenoma papilliferum is still debated: Many authors insist that this lesion is an apocrine tumor because of the occasional presence of decapitation secretion in some of the luminal cells of the tumor\textsuperscript{21,22,23} and the frequent presence of tubular glands with large lumina and decapitation secretion beneath the tumor.\textsuperscript{2,22}

Pinkus\textsuperscript{2} has reviewed the dynamics of syringocystadenoma papilliferum and concluded that the histogenesis of syringocystadenoma papilliferum is multiform, i.e. whereas most cases originate from the proliferated, mature, apocrine sweat gland apparatus, some are associated with the eccrine sweat gland or originate from pluripotential cells in the adult epidermis stimulated by trauma or unknown factors.

So far, only three electron microscopic studies have been published on syringocystadenoma papilliferum, with contradictory results. Hashimoto\textsuperscript{3} reported that syringocystadenoma papilliferum is a tumor differentiating towards eccrine structures, and therefore he classified syringocystadenoma papilliferum in his monograph as a tumor of eccrine sweat gland origin. He showed that ductal structures consisted of two types of secretory cells, light and dark, like secretory cells of the eccrine gland. The secretory granules in the dark secretory cells, in contrast to apocrine secretory granules, were small and did not coalesce. However, Niizuma\textsuperscript{4} insisted that none of the histological techniques supplies enough evidence to confirm the difference between apocrine and eccrine sweat gland apparatus, especially in pathologic conditions such as tumors and nevi. His electron microscopic study revealed a non-keratinized intracytoplasmic cavity and an intercellular canaliculi which apparently formed as a continuation of this cavity. Keratinized cells were absent from the glandular and duct epithelium and even from the superficial epithelial cells nor secretory granules were identified in areas showing tubular, glandular, or sinusoidal structures. On the basis of these findings, he concluded that the tumor differed from both the intrafollicular and intradermal duct of the embryonic apocrine sweat gland apparatus. Toribio et al\textsuperscript{14} reported that syringocystadenoma papilliferum shows apocrine secretion.

Histochemical studies have been similarly inconclusive. The luminal cells lining the villi have given a positive Turnbull blue reaction for iron, which suggests an apocrine gland.\textsuperscript{2} The presence of large amounts of PAS-positive, diastase resistant material throughout the luminal cells and of alcian blue positive material in their apical portion favors apocrine differentiation.\textsuperscript{21} The presence of phosphorylase and succinic dehydrogenase activity suggested eccrine differentiation.\textsuperscript{25} However, phosphorylase activity was absent except for a few foci, a factor in favor of apocrine differentiation.\textsuperscript{26}

Also, positive immunoreactivity for gross cystic disease and fluid protein (GCDFP-15) in all eight cases of syringocystadenoma papilliferum thus tested supported an apocrine genesis.\textsuperscript{27}

In our case, the light microscopic study showed that PAS positive diastase-resistant material was found in luminal cells, however, there was no evidence of decapitation secretion which might represent an apocrine tumor. In the electron microscopic study, we were able to find decapitation secretion on luminal cells which had many vil-
lous projections. Luminal cells consisted of two kinds of densities, light and dark cells. Our findings may represent both directions of differentiation.

However, it has been suggested that, under pathologic condition, apparent decapitation secretion in the glands deep in the dermis cannot be taken as unequivocal evidence that the lesion is basically of apocrine origin. Niizuma suggested that the difference in density was attributed to a variation in the electron-density of the cell matrices rather than a difference in kind or quantity of cell organelles such as free ribosomes, rER, glycogen particles, and tonofilaments. Considering above two suggestions, the direction of differentiation in our case is not certain. We think that the view expressed by Pinkus probably is correct, although most lesions of syringocystadenoma papilliferum are apocrine in differentiation, some are eccrine.

REFERENCES