A Case of Solitary Cylindroma

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A 57-year-old female patient visited our clinic complaining of a single skin lesion on the forehead she had for several years. The lesion was a whitish to yellowish colored, bean-sized nodule. Histologically, the tumor mass is composed of irregularly shaped islands that fit together like pieces of a jigsaw puzzle. The epithelial cell islands are surrounded by a hyaline sheath and a narrow band of collagen which react positively to staining with antibodies against type IV collagen and laminin. These pictures were quite characteristic of cutaneous cylindroma.

We report a rare case of benign adnexal tumor, cylindroma, occuring in a single lesion on the forehead. In our knowledge, this is the first report in Korea.

(Ann Dermatol 6:(2) 208-211, 1994)

Key Words: Cylindroma

Cylindroma is an unusual beingn adnexal tumor, first described in 1842 by Ancell, and named in 1859 by Billroth[1,2]. It occurs more often as a solitary lesion than as multiple lesions which are dominantly inherited. Solitary cylindromas are not inherited. They appear in adulthood and the most common location is the scalp, but they can be seen on the face and elsewhere[2-4]. Clinically they appear as smooth, firm and pink to red-orange dermal tumors that may be pedunculated at times. They usually vary in size from a few millimeters to several centimeters[1]. Their histologic appearance[4] is the characteristic jigsaw puzzle arrangement of epithelial cell clusters encircled by dense hyaline bands which are PAS-positive and diastase resistant. The hyaline membrane contains the basement membrane macromolecules, laminin and collagens IV and VII. The origin of this tumor is controversial, and both eccrine and apocrine derivations have suggested.

We report herein a case of solitary cylindroma together with all the results from the histopathological and immunohistochemical evaluation.

Received August 12, 1993.
Accepted for publication November 15, 1993.
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REPORT OF A CASE

A 57-year-old female visited our out-patient department in February 1993 with a several-year history of a skin-colored nodule on the forehead. The lesion had slowly grown to its present size without prior bleeding, ulceration or crust. No subjective symptom except mild tender sensation was present. There was no history of previous treatment. No family members had a similar lesion. The patinet's past medical history was unremarkable. Routine laboratory tests including complete blood cell count, liver function test, urinalysis and chest roentgenogram were within normal limits or negative.

Cutaneous examination revealed well-demarcated, whitish to yellowish, bean-sized nodule with hair loss and telangiectasia on the forehead (Fig. 1). The specimen obtained from surgical excision of the lesion was used for light microscopic evaluation. Histopathologically, we diagnosed the forehead lesion as a typical cylindroma. In hematoxylin-eosin stain of the lesion, the tumor mass consists of various size of epithelial cell islands with jigsaw

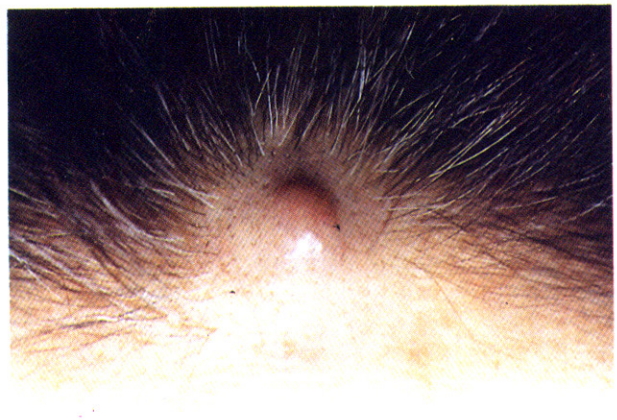

Fig. 1. Well-demarcated, skin-colored, bean-sized nodule with hair loss and telangiectasia on the forehead.

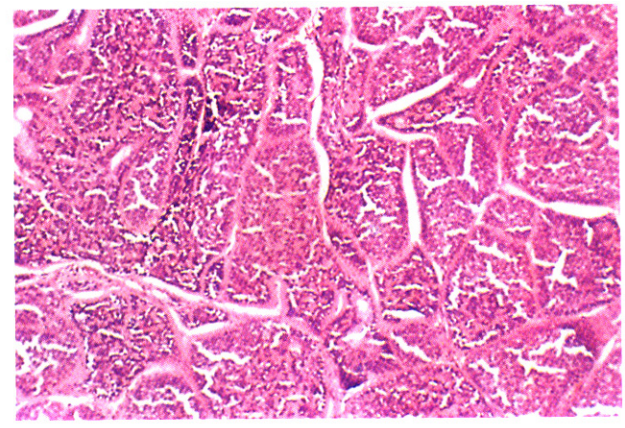

Fig. 2. The tumor is made up of irregularly shaped islands that fit together like pieces of a jigsaw puzzle. Tumor islands are composed of dark and light cells and are surrounded by a hyaline sheath (H & E stain, × 100).

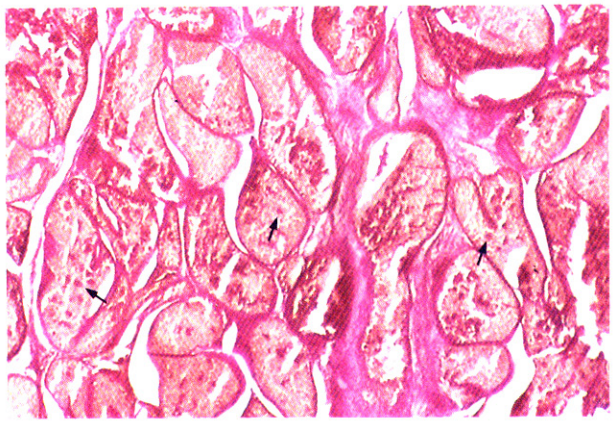

Fig. 3. PAS-staining shows the hyaline bands bordering the tumor cell clusters and the hyaline droplets (arrow indicated) within the cell nests (PAS stain, × 100).

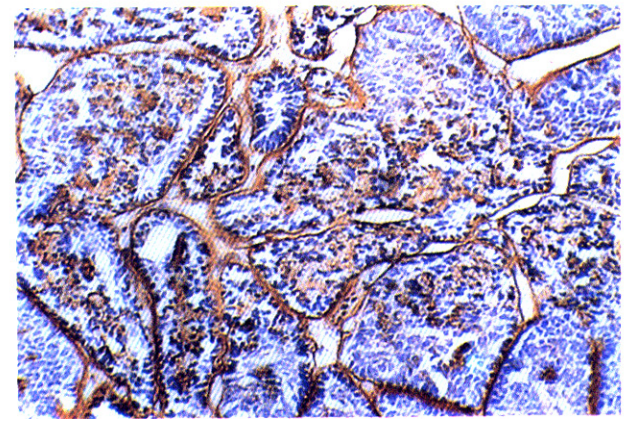

Fig. 4. Positive reactivity with antibodies against type IV collagen in the hyaline membrane and in the particles of hyalin (Immunohistochemical stain, × 200).

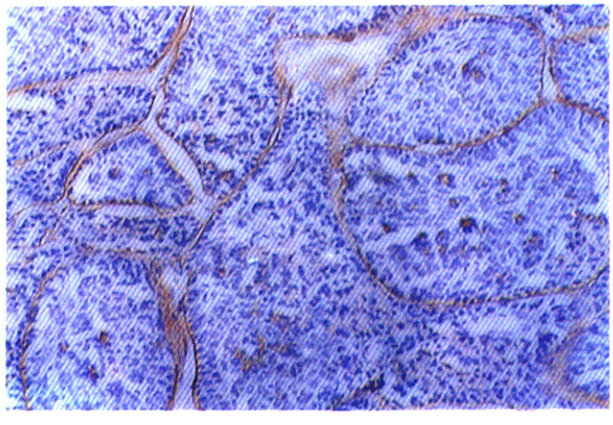

Fig. 5. Positive reaction against laminin (Immunohistochemical stain, × 200).

puzzle arrangement, which are separated often by a hyaline sheath and a narrow band of collagen. Two types of epithelial cells were noted within the tumor lobules: a peripheral cell with a small, dark-staining nucleus and a tendency for palisading, and a larger cell with a pale nucleus more centrally located (Fig. 2). In PAS stain, the thin hyaline band surrounding the tumor islands of cylindroma is PAS-positive and diastase-resistant. Droplets of similar hyaline material are present in the cell nests (Fig. 3). Immunohistochemical stain with antibodies against type IV collagen and laminin showed positive reactivity in the hyaline sheath and also the particles of hyalin present between the cells of tumor islands (Fig. 4, Fig. 5).On the

basis of clinical and histopathological data, a diagnosis of solitary cylindroma was made.

DISCUSSION

Ancell in 1842 is credited with the first description, and Billroth in 1859 coined the term cylindroma[1,2]. Cylindroma is an uncommon benign adnexal tumor of uncertain origin with a characteristic histology and usually manifests as nodules or tumors of the scalp and face[2-4].

The lesion most commonly presents as solitary nodules, but multiple tumors also occur frequently. Their size ranges from a few millimeters to several centimeters[1] and there is a strong predilection for middle-aged and elderly women[5].

Solitary cylindromas are not inherited. They appear in the head and neck in 90% of cases and can affect any skin surface except the soles, palms, and axillace[2,6]. Multiple cylindromas are dominantly inherited[7] and its large varient, usually with multiple coalescing tumors, may arise on the scalp and forehead. These are commonly known as turban tumors. The multiple type has also been associated with the multiple, inherited form of trichoepitheliomas[8] and with eccrine spiradenoma[9].

In our case, a 57-year-old female patient had a single, skin-colored, bean-sized nodule on the forehead for several years. There were no subjective symptom and family history.

The histological features of cutaneous cylindroma are quite characteristic[4]. The intradermal tumor masses are well defined but not encapsulated, and not connected to the epidermis. Commonly, the indivudual nodules are made up of rounded nests of epithelial cells, which sometimes interlock with the adjacent nests to form a jigsawlike pattern. Two cell types are often described: a small dark cell, often located in the periphery of the tumor lobules, and a larger pale cell, which makes up the central portion of the clusters. A characteristic feature of cylindroma is the thick, eosinophilic, hyaline basement membrane material that surrounds the tumor lobules. It is strongly PAS positive, and resistant to diastase. Droplets of similar hyaline material may be present in the cell nests. This thick hyaline membrane and the particles of hyalin present between the cells of the tumor islands react positively to staining with antibodies against type IV collagen and laminin[4,10], analogous to the subepidermal lamina densa. It is also reported that this hyaline membrane contains the type VII collagen[11], the major structural component of the anchoring fibrils. In our patient, the intradermal tumor mass is composed of irregularly shaped islands that fit together like pieces of a jigsaw puzzle. The epithelial cell islands are surrounded by a hyaline sheath or membrane which is PAS-positive and diastase-resistant, and which also reacts positively to staining with antibodies against type IV collagen and laminin. These findings were consitent with cutaneous cylindroma.

Ultrastructural study[4,5,12] shows small dense basal cells, large light indeterminate cells, ductal cells, secretory cells containing secretory granules and some Langerhans cells. In the view of some authors, the differentiation of the secretory cells is eccrine, because they contain secretory vacuoles resembling those seen in the dark mucoid cells of the eccrine gland. Other authors have observed within the secretory cells two types of secretory granules similar to the secretory cells of apocrine glands. Decapitation secretion indicative of apocrine secretory activity has also been observed. Ductal structures are also present. The thick hyaline band is composed of thickened amorphous basal lamina and a fibrous component consisting of anchoring fibrils[11].

So the origin of this tumor has been controversial[1,4,5]. Apocrine, eccrine and pilar differentiation have been claimed by different authors. Mixed apocrine and eccrine features have also been reported[12].

Clinically[1,5], the multiple cylindroma on the scalp is most likely to be confused with a trichilemmal cyst which is, however, usually smoother, firmer and more mobile. Also solitary tumors of a small size must be distinguished from trichoepithelioma, steatocystoma multiplex or basal cell epithelioma. Histopathologically[1,4], eccrine spiradenomas can bear a striking similarity to cylindromas. The tumor nodules of a spiradenoma tend to be rounded and the tumor islands are also composed of two cell types. Larger paler cells lie in the center of the aggregates and may be grouped around lumina, and smaller darker cells form the periphery. In the absence of lumina, the cells with pale nuclei may show a characteristic rosette arrangement. Eccrine spiradenoma[13] is clinically characterized by a solitary, deep-seated nodule occuring most frequently on

the ventral surface of the body, especially over the upper half. A striking symptom is pain appearing in paroxysms. It occurs most frequently between 15 and 35 years of age, equally in both sexes and not familial.

Local aggressive behavior and malignant transformation[3] of cylindroma are uncommon. It is characterized by loss of the hyaline membrane and expanded cellular islands composed predominantly of larger cells, devoid of peripheral palisading. Also it is usually associated with long-standing turban tumors of the scalp. However, metastasis is very rare[5].

Surgery is the treatment of choice[14]. Extensive involvemnt of the scalp may require wide excision and replacement of the whole area by a graft. The patient in the present case was treated with surgical excision.We report a rare case of solitary cylindroma showing typical clinicopathologic findings.

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