



Solitary dermal cylindroma: a rare case report

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ABSTRACT

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Dermal cylindromas are rare benign skin appendageal tumors that has two clinical presentation, solitary and multiple form. The diagnosis is unusual. However, it must be considered in clinical practice. A 51-year-old woman had painless nodule on right arm. Skin examination revealed two fragmented nodules, 1x0.5x0.5cm, tan to white, and rubbery. Histopathological findings revealed a well demarcated and an unencapsulated epithelial dermal tumor, composed of numerous oval and polygonal nests molded into a "jig-saw" or "mosaic" appearance at low power. The nests of cells were composed of basaloid cells with scant cytoplasm and hyperchromatic nuclei and paler cells at the center. Nests of epithelial cells were surrounded and penetrated by a thickened band of basement membrane material that was PAS-positive. Immunohistochemistry (IHC) staining with p63 showed positive staining on basaloid cells. Langerhans dendritic cells in the surrounding nests stained positive with CD1a. Solitary dermal cylindroma occurred sporadically in patients without family history of cutaneous cylindromas. Combination of morphologic, histochemistry and immunohistochemistry staining were needed for accurate diagnosis.

ABSTRAK

Silindromadermal adalah tumor jinak adneksa kulit yang jarang ditemukan dan dapat berupa lesi soliter maupun multipel. Kasus silindromadermal jarang ditemukan, akan tetapi diagnosis ini perlu dipertimbangkan dalam praktik klinis. Wanita berusia 51 tahun dengan keluhan nodul pada lengan kanan yang tidak nyeri. Pemeriksaan lesi kulit ditemukan dua buah nodul berukuran masing-masing 1x0,5x0,5cm, berwarna putih kecoklatan dan kenyal. Pemeriksaan histopatologi menunjukkan tumor epitelial pada dermis yang berbatas tegas dan tidak berkapsul yang terdiri dari sarang-sarang tumor yang berbentuk oval dan poligonal yang membentuk gambaran *jigsaw* atau *mosaic*. Bagian tepi terdiri atas sel-sel basaloid dengan sitoplasma sedikit dan inti hiperkromatis, sedangkan sel-sel pada bagian tengah tampak lebih pucat. Sarang-sarang tumor dikelilingi dan dipenetrasi oleh membran basal yang terpulsa positif dengan PAS. Pengecatan IHC dengan p63 menunjukkan hasil positif pada sel-sel basaloid. Sel-sel langerhans disekitar sarang-sarang tumor terpulsa positif dengan CD1a. Kombinasi pemeriksaan morfologi, histokimia dan imunohistokimia diperlukan untuk diagnosis yang akurat.

Keywords:

Dermal cylindroma
solitary
PAS
p63
CD 1a

INTRODUCTION

Dermal cylindroma is a rare tumor in clinical practice, in which the diagnosis is histological surprise because usually diagnosed as another skin lesion.¹ It is a benign cutaneous tumor that have historically been described as sweat gland tumors and have been considered to be an eccrine lineage.¹ However, recent study showed that cylindroma is arising from pluripotent stem cells in the hair follicles.² Dermal cylindroma most commonly occur on the head and neck as solitary or multiple tumors. Solitary cylindroma occur sporadically and typically are not inherited. While multiple cylindromas are observed in an autosomal dominant inherited manner.³ Multiple cylindromas are generally observed as a component of Brooke–Spiegler syndrome (BSS) or as the only skin lesion of familial cylindromatosis (FC).⁴ We here reporting

an unusual case of solitary cylindroma arose in arm of a 51-year-old woman.

CASE

A 51-year-old woman experienced painless right arm nodule that accidentally found on lumpectomy for breast mass. Macroscopic appearance was two fragmented nodules, 1x0.5x0.5cm, tan to white, and rubbery. Histopathological findings revealed a well demarcated and an unencapsulated epithelial dermal tumor, composed of numerous oval and polygonal nests molded into a “jig-saw” or “mosaic” appearance at low power. Some with tubular formation. The nests of cells were composed predominantly of basaloid cells with scant cytoplasm and hyperchromatic nuclei. At the center of the nests, the cells had increased eosinophilic cytoplasm and vesicular nuclei (FIGURE 1 and 2).

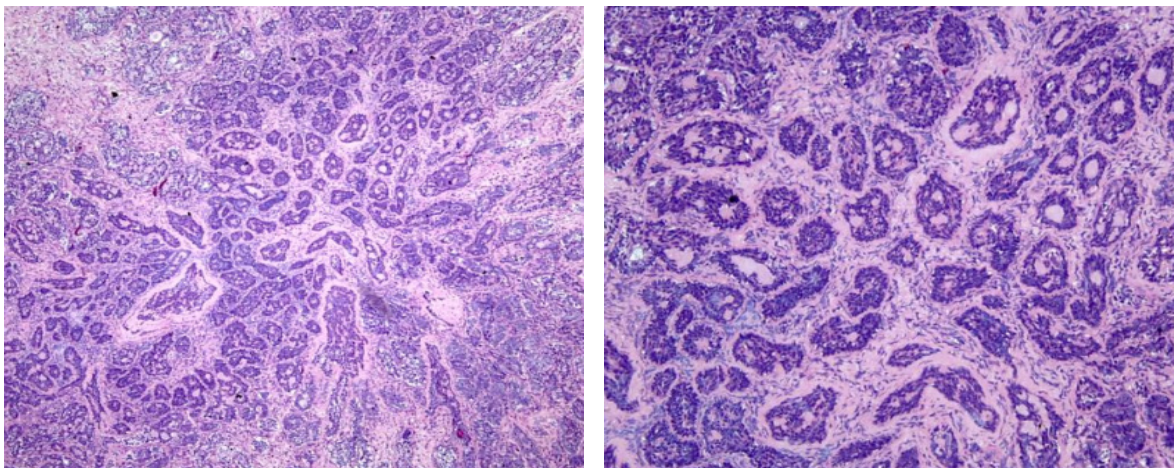


FIGURE 1. Solid, irregular cell nets with “jigsaw patterns”

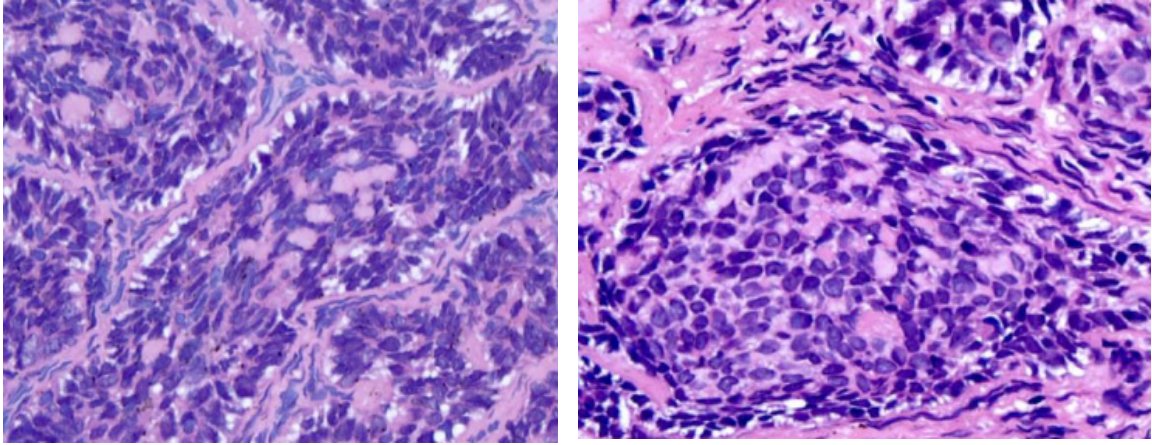


FIGURE 2. The nests surrounded and penetrated by eosinophilic hyalin membrane.

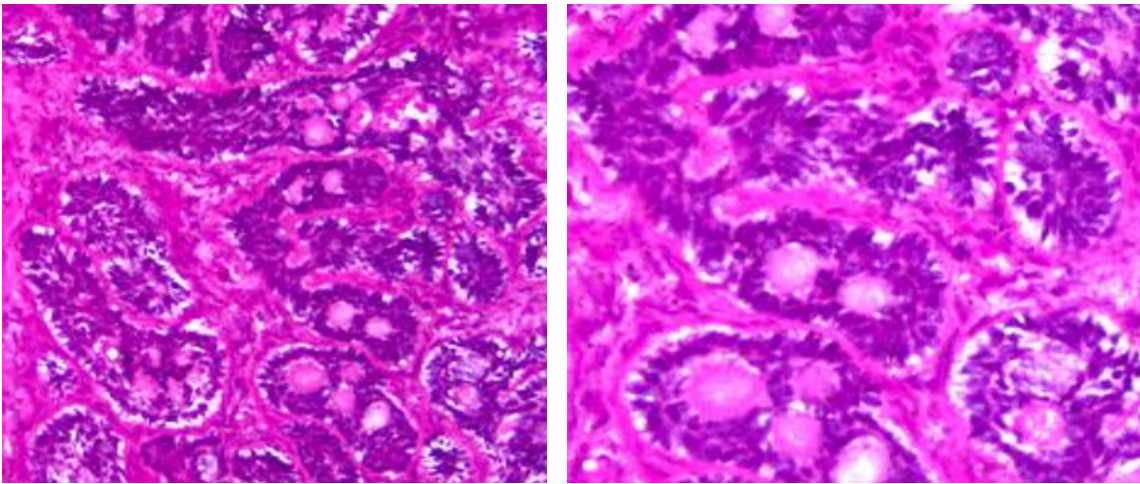


FIGURE 3. The hyalin membrane showed positive

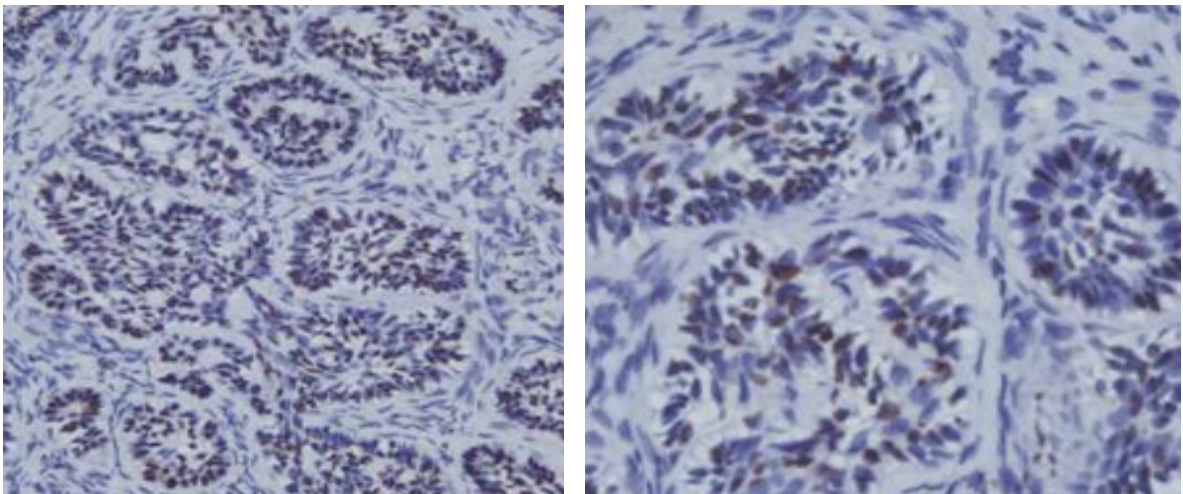


FIGURE 4. The basaloid showed p63 nuclear staining

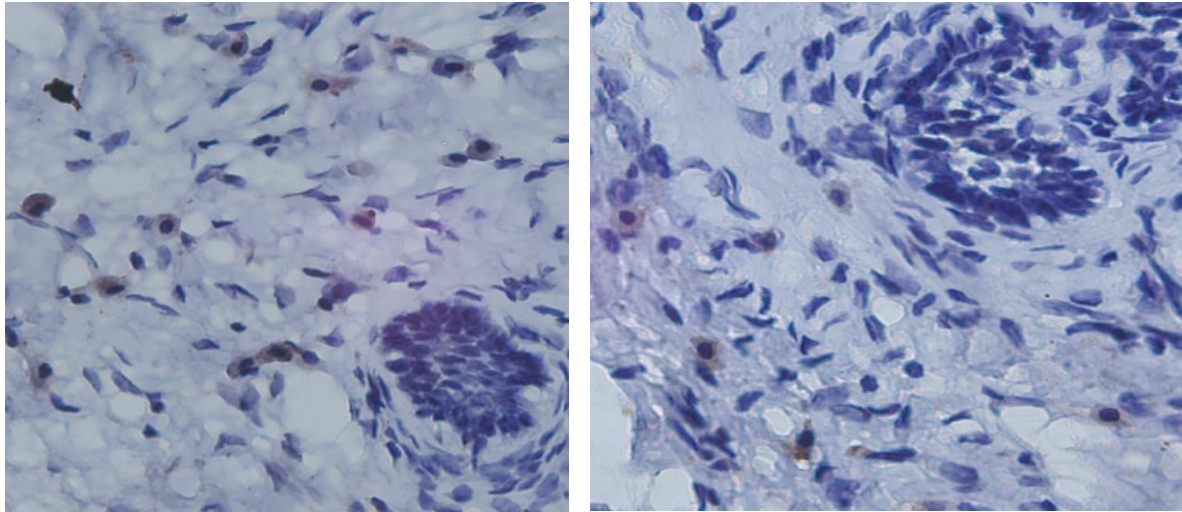


FIGURE 5. Langerhans cells showed membrane and cytoplasmic staining with CD 1a.

Nests of epithelial cells are surrounded and penetrated by a thickened band of basement membrane material that was PAS-positive (FIGURE 3). Immunohistochemistry (IHC) staining with p63 showed positive staining on basaloid cells as shown in FIGURE. 4. Langerhans cells in the surrounding nests stained positive with CD 1a (FIGURE. 5).

DISCUSSION

Dermal cylindromas are benign skin appendage tumors that are most frequently found in scalp and neck skin, with predilection for middle aged and elderly females.⁵ This neoplasm is generally asymptomatic, but some patients may experience pain.⁶ The term cylindroma was first used in 1856 by Billroth to describe an orbital tumor with hyalin and glassy appearance.^{4,7} The term cylindroma originated from its cylindrical shape seen in transversal section.⁸ Currently, three forms of cylindromas are recognized: the benign dermal cylindroma, which may occur as solitary or multiple lesions; the malignant salivary cylindroma (adenoid cystic carcinoma); and the malignant cylindroma.^{4,9} Although the nomenclature was identical, however the clinical behavior and prognosis were

very different.⁴

Cylindroma has two clinical presentation, solitary and multiple form. Solitary form occurs sporadically in patients without family history of cutaneous cylindromas. The solitary form occurs as frequent as the multiple form or even more frequently.¹⁰ Variant with multiple cutaneous cylindromas are inherited in an autosomal dominant manner. The observation of multiple cylindromas was first made by Brooke in 1892 and later by Spiegler in 1899.⁴ Nowadays, the presence of multiple cylindromas is known as the Brooke–Spiegler syndrome. The Brooke–Spiegler syndrome is classified by the presence of cylindromas, trichoepitheliomas, and occasionally, spiradenomas, on the head and neck, beginning in the second decade of life.¹¹

Multiple cylindromas may form a “turban tumors”. The term turban tumor is used to describe the localization of the tumor and its pathognomonic size that can cover the whole scalp.¹² Cylindromas are characterized by irregularly shaped islands of basaloid cells arranged in a “jigsaw puzzle” pattern, which are surrounded by an eosinophilic hyaline sheath.¹³ The tumoral isles typically present two groups of cells: peripheral cells in palisade with a

small dark nucleus, which represent undifferentiated epithelial tumoral cells, and more differentiated centralized cells, with a big pale nucleus resembling ductal or secreting cells.^{4,14}

The nests of cylindroma are commonly surrounded by a rim of densely eosinophilic PAS-positive basement membrane material, and the nests are also punctuated by small round droplets with similar staining qualities.¹⁴ This material was immunoreactive for collagen IV.¹⁵ Diffuse positivity for p63 in the basaloid cells and CK7-positivity in the larger central cells support the morphological impression of two divergent types of cellular differentiation; myoepithelial and ductal. The extent of positive nuclear staining for p63 supports predominant myoepithelial differentiation in this tumor.¹⁶

The histogenesis of cutaneous cylindroma has remained a subject of intense and controversial debate.^{2,13} There are controversies whether its origin is in eccrine, apocrine or follicular epithelium.^{3,4,13,14} The absence of cylindromas on the palms of hands and soles of feet, skin with the highest density of eccrine glands but without hair follicles, support the idea that hair follicles are the cell of origin for these tumours.¹⁷ Immunostaining study by Massoumi *et al.*¹³ in reassures that cylindroma likely originates from the epithelial hair follicle. Another immunohistochemical study with CD200 and other stemcell markers has also indicated the hair follicle to be the origin of cylindroma.²

Dermal cylindroma is usually benign, but malignant transformation of the multiple or solitary dermal cylindroma have been reported. However, it is considered to be rare. The malignant differentiation tendency is higher in the multiple type of cylindroma than in the solid type.¹⁸ The clinical

signs of malignant transformation are ulceration, rapid enlargement, bleeding, and change in the color of the nodules.⁹ The characteristic histologic features of malignant cylindroma are loss of jigsaw pattern, loss of hyaline sheaths, loss of peripheral palisading at the tumor island periphery, loss of bimorphic cell composition, marked nuclear anaplasia, stromal invasion, focal areas of necrosis, pleomorphism of nuclei, and increased and abnormal mitoses.^{8,9}

Differential diagnosis dermal cylindroma werespiradenoma and adenoid cystic carcinoma. Cylindroma may show overlap or even occur together with spiradenoma. Spiradenoma is a benign dermal neoplasm that can show either eccrine or apocrine differentiation. Spiradenoma classically presents in patients between the ages of 20 and 40 years and appears as a solitary, well-circumscribed, firm nodule, measuring usually less than 1 cm, but giant variants and multiple lesions have also been described. The most striking clinical feature of spiradenoma is the presence of pain or tenderness. Spiradenoma is distinguished from cylindroma by the lack of hyaline cylinders. Furthermore, spiradenomas tend to be more vascular and show more likely edematous and cystic stromal changes.¹⁹ Tumor cells with tubular differentiation and lymphocyte infiltration also differentiated spiradenoma from dermal cylindroma.^{14,20} Cylindroma also needs to be distinguished from adenoid cystic carcinoma. The solid variant of adenoid cystic carcinoma differs from cylindroma by its infiltrative growth pattern, cytologic atypia, mitotic figures, and the lack of a thickened basement membrane around the epithelial structures.¹⁵

CONCLUSION

Solitary dermal cylindroma is benign appendageal tumour that

occur sporadically in patients without family history of cutaneous cylindromas. Combination of morphologic, histochemistry and immunohistochemistry staining are needed for accurate diagnosis.

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