Proliferating Trichilemmal Tumor In The Chest Wall: Report Of A Rare Case

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Abstract

Proliferating trichilemmal tumor (PTT) is an uncommon, usually benign lesion that usually arises on the scalp of elderly women. The lesion bears close morphologic resemblance to squamous cell carcinoma at the microscopic level, making a correct histologic diagnosis extremely important. We present a case of PTT occurring on the chest wall of a 75-years-old man to highlight the significance of recognizing this unusual tumor at an uncommon location and discuss points of differentiation from squamous cell carcinoma.

Key words

Trichilemmal cyst, proliferating trichilemmal tumor, pilar tumor, squamous cell carcinoma.

Introduction

Proliferating trichilemmal tumor (PTT), also known as proliferating trichilemmal cyst or pilar tumor is an uncommon but usually benign skin appendageal tumor with distinctive morphologic features. Ninety percent of the cases occur on the scalp, though there are infrequent reports of its occurrence at various other sites such as the forehead, nose, back, chest, abdomen, buttocks, elbow, wrist, mons pubis and vulva. [1,2] The lesion assumes importance because of its close morphologic resemblance to squamous cell carcinoma. Here we report a case of proliferting trichilemmal tumor occurring on the chest wall of a 75-year-old man and discuss the differentiating features between the two entities.

Case Report

A 75-year-old Kuwaiti male presented with history of a painless gradually enlarging nodule on the anterior chest wall for the last six months. Local examination revealed a 1x1 cm soft, freely mobile, lobulated, skin covered mass on the left anterior chest wall. The overlying skin was unremarkable. The clinical diagnosis was skin papilloma. The lesion was completely excised. On gross examination, it revealed a skin covered globular mass measuring 1x1cm. Cut surface showed a well-circumscribed greyish brown firm tumor in the dermis with cystic spaces filled with yellowish cheesy material. Microscopically, the lesion was well demarcated from the epidermis and demonstrated coalescing masses of squamous epithelium showing abrupt trichilemmal type of keratinization with formation of horn pearls focally (Fig. 1 and 2). There were foci of cystic degeneration filled with keratinous debris along with areas of hemorrhage. There was no atypical mitosis or cellular atypia. The excision margins were free of tumor. Based on the presence of proliferating lobules of squamous epithelium showing trichilemmal type keratinization and absence of cellular atypia, the diagnosis of proliferating trichilemmal tumor was entertained.

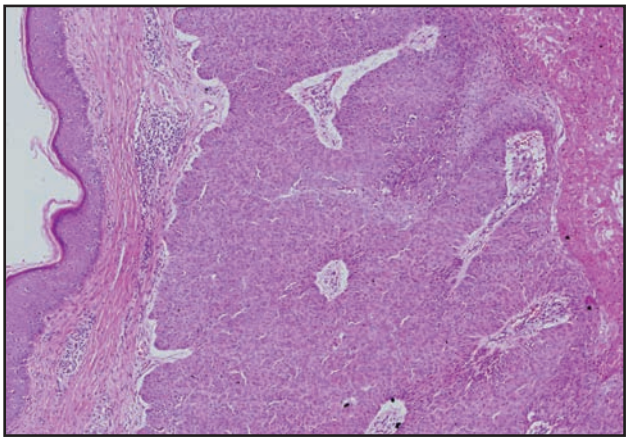

Fig. 1: Photomicrograph showing proliferating lobules of squamous epithelium in the dermis (H&E x 200).

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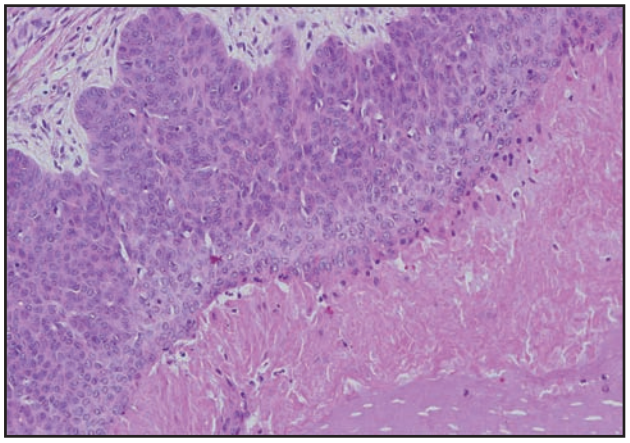

Fig. 2: Squamous islands showing abrupt trichilemmal keratinization (H&E x 200)

Discussion

Proliferating trichilemmal tumor, also known as pilar tumor or proliferating trichilemmal cyst, is an uncommon but usually benign lesion, which may be misdiagnosed as squamous cell carcinoma as both bear close histologic similarities.[1]In almost ninety percent of the cases the tumor is confined to the scalp, though they have also been found in various other locations such as the forehead, nose, back, chest, abdomen, buttocks, elbow, wrist, mons pubis, and vulva.[2,3] Most patients are women (84%) in the age group of 27 to 83 years. [2] Typically, it appears as a slowly enlarging, painful subcutaneous scalp nodule with a tendency to bleed, and produce purulent discharge. Clinically, those are often mistaken for sebaceous cyst, pilar cyst, and even dermatofibroma.[2] Ulcerated lesions often masquerade as squamous cell carcinoma .The tumor may occur in association with one or even several trichilemmal cysts of the scalp. There is also evidence suggesting that a PTT may develop from an ordinary trichilemmal cyst.[2, 4] These tumors originate from the outer sheath of the hair follicle .[3] Grossly, the tumors are sharply demarcated and lobulated, ranging from 0.4 to 10 cm in diameter. However, a tumor as large as 25 cm is also reported.[5] Cut surface reveals solid and cystic areas, sometimes with honeycomb configuration.[2] The tumor exhibits distinctive histologic appearance characterized by sharply circumscribed convoluted lobules of squamous epithelium. Characteristically, the epithelium in the center of the lobules abruptly changes into eosinophilic amorphous keratin without formation of granular layer.[3, 5,6] Foci of calcification, although generally small, are often present in the areas of amorphous keratin.

The morphologic resemblance of PTT with well-differentiated squamous cell carcinoma often poses diagnostic challenge to the histopathologists. The tumor cells in an otherwise classical PTT may show focal areas of nuclear atypia as well as individual cell keratinization, which at first glance suggest a squamous cell carcinoma. The presence of abrupt keratinization, minimal pleomorphism, low mitotic activity, sharp circumscription, foci indistinguishable from a trichilemmal cyst, calcification, and absence of a pre-malignant lesion such as actinic keratosis help to differentiate proliferating trichilemmal tumor from squamous cell carcinoma.[2,6,7] In rare instances, PTT can undergo malignant trans-formation, indicated by rapid enlargement of the nodule. Histologically, malignant proliferating trichilemmal tumors show severe nuclear atypia, marked cellular pleomorphism with atypical mitoses, dyskeratotic cells, and infiltrating margins.[3, 8] A combination of various features such as non-scalp location, recent rapid growth, size greater than 5 cm, infiltrative growth, and significant cytologic atypia with mitotic activity favor a diagnosis of malignant PTT. [9] Mutations of *p53* gene have been reported, [10] although others failed to detect over-expression of p53 protein.[8] Lymph node and distant metastases have been reported in malignant proliferating trichilemmal tumors.[7, 8] Surgical excision with wide margins is the treatment of choice for both benign and malignant PTT.However, both forms are known to recur even after adequate resection. Chemotherapy and radiotherapy have also been used for malignant tumors.[8]

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