

# AN UNUSUAL ADNEXAL TUMOR WITH DIFFERENTIATION TOWARDS HAIR FOLLICLE, WITH CONCOMITANT PRESENCE OF TRICHOFOLLICULOMA, TRICHOEPITHELIOMA AND PILOMATRICOMA IN ONE LESION: A CASE REPORT

GUITI IRAVANLOU, M.D., AND FATEMEH MAHJOUR, M.D.

*From the Cancer Institute, Pathology Department,  
Imam Khomeini Hospital, Tehran, I.R. Iran.*

## ABSTRACT

A case of an interesting previously unreported mixed adnexal tumor of the scalp of a 38 year old man is presented, which was composed of randomly dispersed areas of trichofolliculoma, trichoepithelioma, and pilomatricoma concomitantly. While these lesions are classified under adnexal tumors with differentiation towards hair follicle structures, controversy is present concerning the exact origin and the pathway of their differentiation. So the concomitant presence of them in one lesion may be strong evidence for a common origin or a common direction of differentiation.

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**Keywords:** Trichofolliculoma, Trichoepithelioma, Pilomatricoma, Adnexal tumor, Hair follicle structures.

## INTRODUCTION

Three possibilities exist for the development of benign adnexal tumors; they may develop from primary epithelial germ cells, from pluripotential stem cells, or from cells of pre-existing structures. In some instances primary epithelial germ cells and pluripotential cells differentiate in more than one direction.

Benign adnexal tumors can be divided into four major groups:<sup>1</sup> those differentiating toward: 1-hair follicle structures, 2-sebaceous glands, 3-apocrine glands, and 4-ecrine glands.

These are further divided into three subgroups according to the decreasing degree of differentiation:<sup>1</sup> 1-hyperplasia, 2-adenoma, 3-primordial epitheliomas. Trichofolliculoma, trichoepithelioma and pilomatricoma are classified as benign adnexal tumors with differentiation toward hair follicles. Trichofolliculoma is an adenoma, while trichoepithelioma and pilomatricoma are benign epitheliomas.<sup>1</sup>

There existed some controversies on the mentioned classification,<sup>1,6</sup> for example pilomatricoma was originally described as a calcified sebaceous gland; however, it was rec-

ognized in 1942 that the cells of the tumor differentiated in the direction of hair cortex cells.<sup>1</sup>

Here we have a brief review of different aspects of the three mentioned tumors.<sup>1-5</sup>

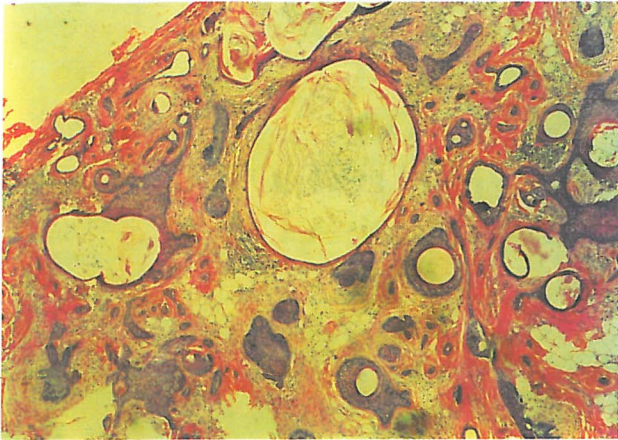
### Trichofolliculoma

It occurs in adults as a solitary, small, skin colored dome shaped nodule, frequently with a central pore, mainly seen on the face but occasionally on the scalp and neck. Histopathological examination of the lesion reveals a large cystic space in the dermis that is lined by squamous epithelium and contains horny material and frequently also fragments of birefringent hair shafts. Sometimes the cystic space is continuous with the surface epithelium. Grouped around the cyst are numerous small secondary well differentiated hair follicles, some of which show in addition to a hair and an outer sheet, an inner root sheet with trichohyaline granules.

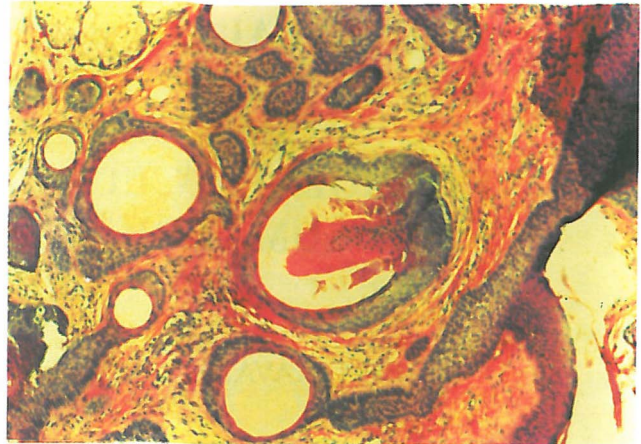
### Trichoepithelioma

It occurs either as multiple or as solitary lesions. The more common solitary form consists of a firm, elevated, flesh colored nodule, usually less than two centimeters in diam-

## Unusual Hair Follicle Tumor



**Fig. 1.** Areas resembling trichofolliculoma; a central cystic structure to which a number of abortive follicles are connected.



**Fig. 2.** Trichofolliculoma. Note the sebaceous glands at the corner of the picture.

eter, most commonly seen on the face, but may occur elsewhere. On histopathological examination, horn cysts represent the most characteristic histologic feature. They consist of a fully keratinized center surrounded by basophilic cells that have the same appearance as the cells in basal cell epithelioma. The keratinization is complete and abrupt. As the second major component, tumor islands of the same basophilic cells are arranged usually in lacelike or adenoid network but occasionally also in solid aggregates, and show peripheral pallisading. Also a foreign body giant cell reaction in the vicinity of ruptured horn cysts and calcium deposits may be seen.

### Pilomatricoma

It usually occurs as a firm, deep seated, solitary nodule mostly on the face and upper extremities. On histopathological examination the tumor is located in the lower dermis and represents irregularly shaped islands of epithelial cells, consisting of basophilic cells and shadow cells. The stroma of the tumor usually shows a considerable foreign body reaction containing many giant cells adjacent to shadow cells and also calcium deposits.

### CASE REPORT

A 38 year old man referred to the Cancer Institute because of a solitary mass lesion of the scalp. After en bloc resection of the lesion, it was sent to our department for pathological examination.

### Gross findings

A firm, gray, well circumscribed nodule 3 centimeters in diameter with a nonhomogenous cut surface consisting of gray white to yellow areas.

### Pathologic examination

A number of slides were prepared from the lesion and

stained with Hematoxylin-Eosin and also S-100 immunostain (DAKO A S-Z 311).

### Histopathology

The H&E stained sections showed a very interesting lesion which was adnexal in nature and showed definite hair follicle differentiation. Part of the lesion resembled a trichofolliculoma, having a central cystic structure to which a number of abortive hair follicles were connected. Other areas consisted of horn cysts with a keratinized center surrounded by basophilic cells, and still other parts showed early developing pilomatricoma with formation of fully keratinized shadow cells. In the S-100 stained slide the structures were all negative for the stain.

### DISCUSSION

Neoplasms of adnexal structures of the skin are of special interest because of the great variety that have been described and the relative rarity of the lesions.<sup>7</sup> Variety is due in part to the presence of several distinct structural units in the skin including pilosebaceous units, eccrine and apocrine glands.<sup>7</sup> Further complexity is created by a subclassification based on the degree of differentiation. As mentioned before benign adnexal tumors can be divided into four major groups and trichofolliculoma, trichoepithelioma and pilomatricoma all belong to the group of those differentiating toward hair follicle structures.

Confirmation of relativeness of each tumor to one of these groups can be accomplished by morphologic, immunohistochemical and electron microscopic studies.

Immunohistochemical study of adnexal tumors includes S-100 stain that indicates origination from eccrine or apocrine glands if positive<sup>1</sup> and involucrine which is claimed to be a good indicator of skin appendage tumors originating from hair follicles<sup>8</sup> but the latter was not performed in our study.



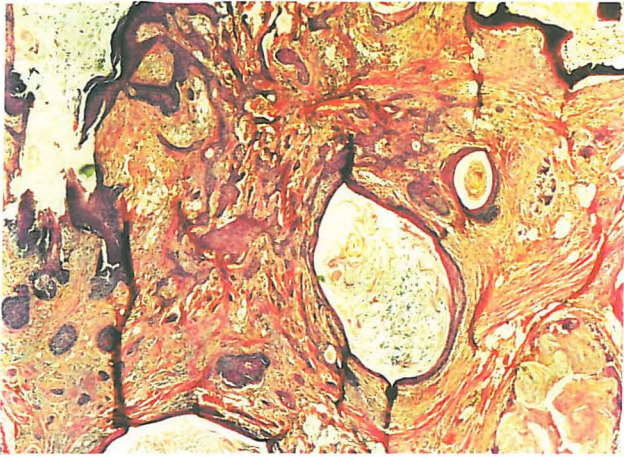


Fig. 3. Areas resembling trichoepithelioma. (predominant feature).



Fig. 4. Trichoepithelioma.

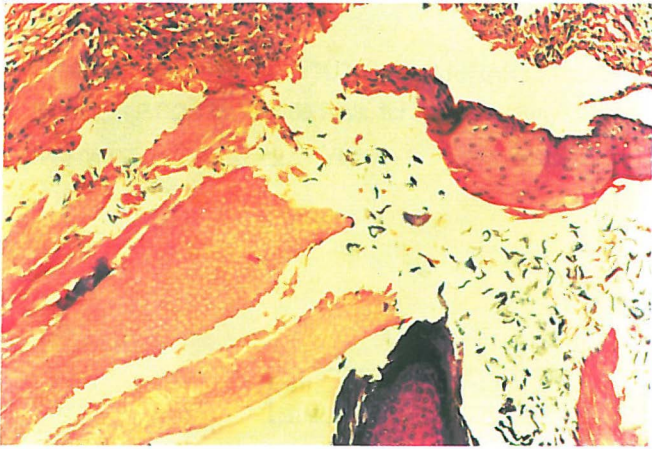


Fig. 5. Pilomatricoma.

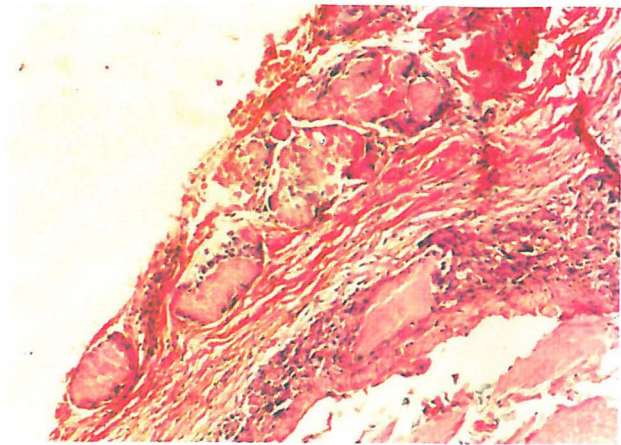


Fig. 6. Pilomatricoma.

The concomitant presence of several different types of tumor in one lesion may denote a common origin and also the same direction of differentiation from primary epithelial germ cells or from pluripotential stem cells which can further be confirmed by the above mentioned methods.

After a complete search of the literature several combinations of variants of benign adnexal tumors were found<sup>9-10</sup> but we did not encounter a combination similar to our case in the literature.

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