Pilomatricoma Combined with Epidermoid Cyst

Soyun Cho, M.D., Kyu-Kwang Whang, M.D., Jeong-Hee Hahm, M.D.

Department of Dermatology, Ewha Womans University, Seoul, Korea

A 22-year-old woman with no history of trauma or other diseases presented with a slowly enlarging firm nodule with a central pointed opening on the right chest. An excisional biopsy revealed a pilomatricoma associated with an epidermoid cyst. Histopathologically, an epidermoid cyst located in the deep dermis was lined mostly by keratin-forming stratified squamous epithelium but focally had columns of shadow cells that projected from the epithelial lining of the cyst into the lumen. The lumen also contained masses of shadow cells, unattached to the epithelial lining. In the stroma surrounding the cyst, numerous masses of shadow cells with giant cell reaction and focal calcification were seen, which were in turn surrounded by a connective tissue capsule. These features are similar to those seen in infundibular cysts associated with Gardner's syndrome. Since epidermoid cysts and pilomatricomas originate in common from the pilosebaceous unit and pilomatricomas begin as infundibular matrix cysts, we suggest that they can occur simultaneously, although rarely, in a healthy person.

(Ann Dermatol 12(3) 155~159, 2000).

Key Words : Pilomatricoma, Epidermoid cyst

Pilomatricoma is a benign tumor with differentiation towards the hair matrix. Pilomatricoma-like changes have been described in epidermoid cysts together with a variety of bone lesions and colonic polyposis in Gardner's syndrome.

We present the case of a patient without Gardner's syndrome who had a pilomatricoma combined with an epidermoid cyst within.

CASE REPORT

A 22-year-old woman presented with a slowly enlarging firm nodule with a central pointed opening on the right chest. The skin lesion had been present for 18 months. At presentation, the bean-sized, movable, erythematous lesion was exuding cheesy material from the central opening (Fig. 1). The patient had no history of trauma or insect bite, and she was in otherwise good health. She had no evidence of multiple osteomas, fibromas, desmoid tumors, lipomas, fibrosarcomas, leiomyomas, familial polyposis of the colon, or any other signs of Gardner's syndrome. An excisional biopsy was performed, and subsequently the diagnosis of pilomatricoma combined with an epidermoid cyst was established. One-year follow-up showed no indication of recurrence.

Histopathologically, a unilocular epidermoid cyst and pericystic masses of shadow cells were surrounded by a connective tissue capsule and located in the deep dermis (Fig. 2A). The cyst was lined mostly by keratin-forming stratified squamous epithelium identical to that found in ordinary epidermoid cysts. There were columns of squamous cells that projected from the epithelial lining of the cyst for variable distances into the lumen (Fig. 2B). The cells had faint, thready outlines, light eosinophilic, strongly birefringent cytoplasm, and central, pale or empty appearing oval spaces that approximated the position and shape of nuclei. The cells were, thus, indistinguishable from the shadow cells of pilomatricoma. The epithelium at the base of the columns of shadow cells were not dis-

Received November 15, 1999.
Accepted for publication April 7, 2000.
Reprint request to : Soyun Cho, M.D., Department of Dermatology Ewha Womans University Tongdaemun Hospital 70 Chongro-6-ka, Chongro-ku Seoul, Korea 110-126
Tel: 82-2-760-5140, Fax: 82-2-743-0825
E-mail: vickycho@hanmail.net

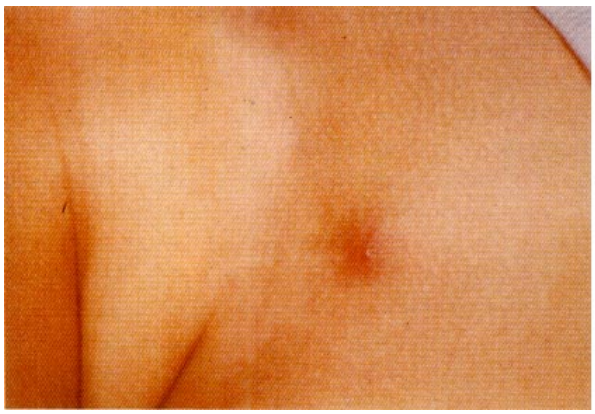

Fig. 1. Firm erythematous movable nodule with a central pointed opening on the right chest.

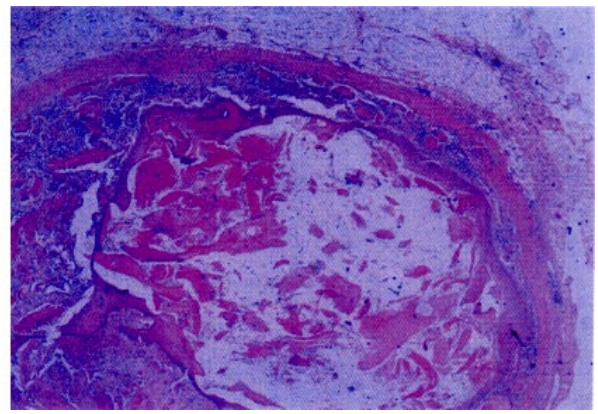

Fig. 2A. An epidermoid cyst and pericystic masses of shadow cells are in turn surrounded by a connective tissue capsule (H & E, ×20).

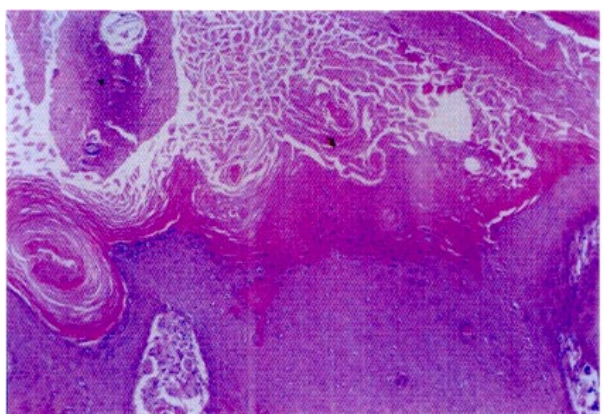

Fig. 2B. Epidermoid cyst with a column of shadow cells projecting into the cyst lumen. Note sharp transition in the cyst wall from infundibular to pilomatricoma-like areas as well as sharp delimitation in the cyst contents between laminated basophilic keratin of the epidermoid cyst in the left and shadow cells of the pilomatricoma-like column in the right. There are also masses of shadow cells floating in the lumen of the cyst, unattached to the epithelial lining (H & E, ×100).

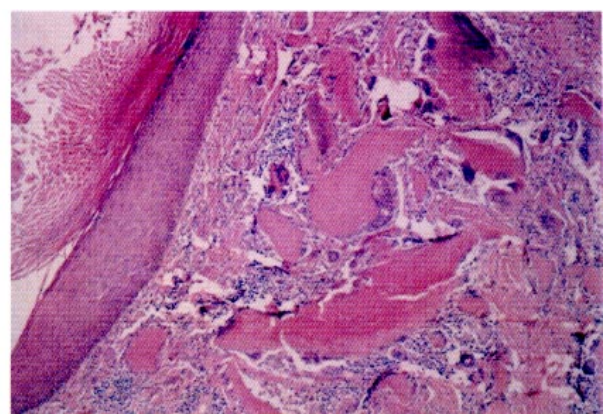

Fig. 2C. Pericystic deposits of shadow cells with giant cell reaction and focal calcification. The cyst lining in this field is that of an epidermoid cyst (H & E, ×40).

tinctly different in appearance from that lining the remainder of the cyst. The transition between the cells at the base of a column and shadow cells within a column was either sharp, with abrupt loss of nuclei, or more gradual, with the persistence of shrunken, pale, shadowy nuclei for variable distances into the column. No keratohyaline or trichohyaline granules were seen in the zone of transition between the lining epithelium and shadow cells. There were four such columns in the cyst. The lumen also contained masses of shadow cells, unattached to the epithelial lining, in addition to loose, flaky keratin characteristic of epidermoid cysts. The pericystic deposits of numerous masses of shadow cells were associated with a giant cell reaction and focal calcification (Fig. 2C). Calcification of luminal contents was not seen, nor was ossification noted.

DISCUSSION

Shadow cells are pale eosinophilic cells with distinct borders and poorly stained round zones in their centers, i.e., shadows or ghosts of nuclei (karyolysis). The presence of shadow cells signifying matrical differentiation is not unique to pilomatricomas but has been reported in several benign and

malignant neoplasms, including matricomas, pilomatrical carcinomas, basal cell carcinomas with matrical differentiation, follicular cysts, adnexal tumors, keratoacanthomas, and squamous cell carcinomas[1-10]. The presence of shadow cells within epithelial neoplasms is a subtle but reliable clue to the follicular nature of those neoplasms: shadow cells in pathological processes represent faulty attempts at formation of hair shafts[9].

The pilomatricoma-like changes known to be present in two-thirds(63%) of epidermoid cysts of Gardner's syndrome are not regarded as true pilomatricomas[6]. Among the cysts and apart from this association, there are only two articles on pilomatricoma associated with them. One is in association with a pilar cyst in the eyelid[11]; this report, however, is lacking in detailed histological documentation. The other one[12] reported 4 cases in which a pilomatricoma was situated in the immediate vicinity of an epidermoid cyst; in one of them the epidermoid cyst was within the tumor itself, just as in our case. In that series, the pilomatricomas were composed mostly of shadow cells with giant cells in stroma, and only 1 case showed a small amount of basophilic cells. The case with epidermoid cyst within the pilomatricoma occurred on the forehead of a 24-year-old male, and it had no basophilic cells. Recently Kaddu et al.[13] identified 4 distinctive histopathologic stages in the evolution of pilomatricoma: the natural course of this neoplasm is a chronological process in which the lesion begins as an infundibular matrix cyst and ends up as a calcified and ossified nodule with no viable epithelial component. Hence in stage 3 or 4, few or no basophilic cells are visible. In a separate article being prepared for publication by us, most pilomatricomas present for longer than 6 months were already in stage 3. Since the lesion in this case had been present for 18 months, the lack of basophilic cells is an expected finding and not an exclusion criteria in diagnosing pilomatricomas. In addition, our case had another evidence of pilomatricoma, a connective tissue capsule surrounding the tumor; thus we believe this case is a true pilomatricoma having an epidermoid cyst within it, distinct from pilomatricoma-like changes seen in the epidermal cysts of Gardner's syndrome.

There appears to be distinct differences in the histogenesis of pilomatricomas and epidermoid cysts. In the latter, the keratin in the keratinized cells consists of relatively electron-lucent tonofilaments embedded in an electron-dense substance, derived from the keratohyaline granules[14]. In the transition zone to shadow cells within the pilomatricoma, however, the keratin forms without the appearance of keratohyaline granules[15]. If the histogenesis of these lesions is indeed different, then some additional factor is necessary to explain their occurrence at the same site. However, since pilomatricomas and epidermoid cysts originate in common from the pilosebaceous unit, albeit from different portions of it, and since pilomatricomas begin as infundibular matrix cysts, we believe that simultaneous occurrence of the two lesions is possible.

Although we could demonstrate a zone of transition between an epidermoid cyst and a pilomatricoma, we chose not to call the association a hybrid cyst, since this was valid for a minority of cases only. A true hybrid cyst from pilomatricoma and infundibular cyst is a single cyst with both types of keratinization in its wall, infundibular in the upper areas connected with surface epidermis and hair matrix-like cells in deeper zones of the cyst wall[16]. The transition between both zones is also sharp. This event should not be confused with pilomatricoma associated with infundibular cyst[12], which consists of the presence of a pilomatricoma adjacent to infundibular cyst as two different neighboring cysts.

In Gardner's syndrome, the cysts are infundibular cysts but in some areas of the cyst wall there are columns of shadow cells, similar to those of pilomatricoma, that project into the lumina[6]. The cells, at the point of attachment of the columns to the cyst lining, are indistinguishable from the hair matrix-like, basophilic cells of pilomatricoma. Some authors suggested that granulomatous reaction had in some manner altered the sequence of keratinization so that shadow cells were formed[5]. These features suggest that pilomatricoma-like changes in the cyst wall and into the lumina may be a characteristic of the infundibular cysts in patients with Gardner's syndrome[16]. In Gardner's syndrome, the pericystic deposits of shadow cells probably result from extrusion of columns and intraluminal masses[6]. Although our patient has no evidence of Gardner's syndrome, the histopathological findings are very similar to those seen in Gardner's syndrome. There are too few reports to differentiate pilomatricoma-like changes in epider-

moid cysts of Gardner's syndrome and true combined pilomatricoma and epidermoid cyst. However, the general architecture of the lesion in our case, a pilomatricoma surrounded by a connective tissue capsule and containing within it an epidermoid cyst, would favor the latter. Although the patient has no recall of trauma, traumatic inoculation of the epidermis might have caused the later development of the epidermoid cyst.

REFERENCES

1. Marrogi AJ, Wick MR, Dehner LP: Pilomatrical neoplasms in children and young adults. Am J Dermatopathol 14:87-94, 1992.
2. Lopansri S, Mihm MC Jr. Pilomatrix carcinoma or calcifying epitheliocarcinoma of Malherbe. Cancer 45:2368-2373, 1980.
3. Weedon D, Bell J, Maize J. Matrical carcinoma of the skin. J Cutan Pathol 7:39-42, 1980.
4. Aloi FG, Molinero A, Pippione M. Basal cell carcinoma with matrical differentiation. Am J Dermatopathol 10:509-513, 1988.
5. Leppard BJ, Bussey HJR. Gardner's syndrome with epidermoid cysts showing features of pilomatrixomas. Clin Exp Dermatol 1:75-82, 1976.
6. Cooper PH, Fechner RE. Pilomatrixoma-like changes in the epidermal cysts of Gardner's syndrome. J Am Acad Dermatol 8:639-644, 1983.
7. Tsoitis G, Brisou B, Destombes P. Mummified cutaneous mixed tumor. Arch Dermatol 111:194-196, 1975.
8. Rapini RP, Kennedy JS, Golitz LE. Hair matrix differentiation in chondroid syringoma. J Cutan Pathol 11:318-321, 1984.
9. Jacobson M, Ackerman AB. Shadow cells as clues to follicular differentiation. Am J Dermatopathol 9:51-57, 1987.
10. Korting GW, Hassenpelug KH. Zur Histogenesis des Epithelioma Malherbe und von seiner topischen Beziehung zum Keratoakanthom. Arch Klin Exp Dermatol 22:11-22, 1965.
11. Ni C, Kimball GP, Graft JL, Wauf WJI, Chong CS, Albert DM. Calcifying epithelioma: a clinicopathological analysis of 67 cases. Int Ophthalmol Clin 22:63-86, 1982.
12. Benharroch D, Sacks MI. Pilomatricoma associated with epidermoid cyst. J Cutan Pathol 16:40-43, 1989.
13. Kaddu S, Soyer HP, H dl S, Kerl H: Morphological stages of pilomatricoma. Am J Dermatopathol 18:333-338, 1996.
14. McGavran MH, Binnington B. Keratinous cysts of the skin. Identification and differentiation of pilar cysts from epidermal cysts. Arch Dermatol 94:499-508, 1966.
15. Hashimoto K, Nelson RG, Lever WF. Calcifying epithelioma of Malherbe. Histochemical and electron microscopic studies. J Invest Dermatol 46:391-408, 1966.
16. Requena L, Sanchez Yus E. Follicular cysts. An expanded spectrum. Am J Dermatopathol 13:228-233, 1991.