

Pilomatricoma: Case Report¹

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Pilomatricoma is a rare benign neoplasm arising from hair follicular matrix cells and exhibiting slow growth. The radiologic features of this neoplasm have rarely been described in the literature; in particular, the ultrasonographic findings have not been published in Korea. We report a case of pilomatricoma presenting as a well-margined soft tissue mass with calcification in the dermis and with overlying bullous skin, a rare clinical variant.

Index words : Skin, neoplasms

Skin, US

Pilomatricoma, neoplasms

Pilomatricoma, or calcifying epithelioma of Malherbe, is a benign, usually asymptomatic tumor arising from hair follicular matrix cells. It is usually a solitary lesion and is most commonly found on the face and upper extremities, appearing as a firm, dermal nodule covered by the normal skin. Although the tumor can occur in patients of any age, about 60% of pilomatricomas are found in those aged less than twenty (1, 2). Most cases involve a single nodule, but multiplicity has been observed (3). Diagnosis is usually based on palpation of a superficial, hard mass and confirmed by histopathologic examination. The authors believe, however, that the ultrasonographic findings of pilomatricoma facilitate accurate diagnosis, and if a calcified soft tissue mass is seen at ultrasonography, pilomatricoma should be included in the differential diagnosis.

Case Report

An otherwise healthy 22-year-old man presented with a two-month history of an expanding mass in the left interscapular area. He had no other associated symptom. At initial physical examination, the tumor appeared as a superficial, bullous soft tissue mass with reddish discoloration, and included a hard, internal nodule (Fig. 1A). Possible clinical diagnoses at that time included dermoid, fibroma, and hemangioma. Plain radiography depicted a well-defined bulging mass with tiny calcifications in its central portion (Fig. 1B), and ultrasonography demonstrated a heterogeneously hypoechoic mass with multiple hyperechoic foci. There was no internal vascularity (Figs. 1C, D). The ultrasonic differential diagnoses were cartilaginous soft tissue mass, dermoid, hemangioma and malignant soft tissue tumor. There was no detectable abnormality in complete blood count, blood chemistry (including serum calcium, phosphorous, and alkaline phosphatase) or urinalysis. The mass, about 3cm in diameter, was successfully excised with 1cm of safety margin. Grossly, its internal texture was wheel-like, a circular pattern of friable tissue was apparent,

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Received June 5, 2002 ; Accepted September 26, 2002

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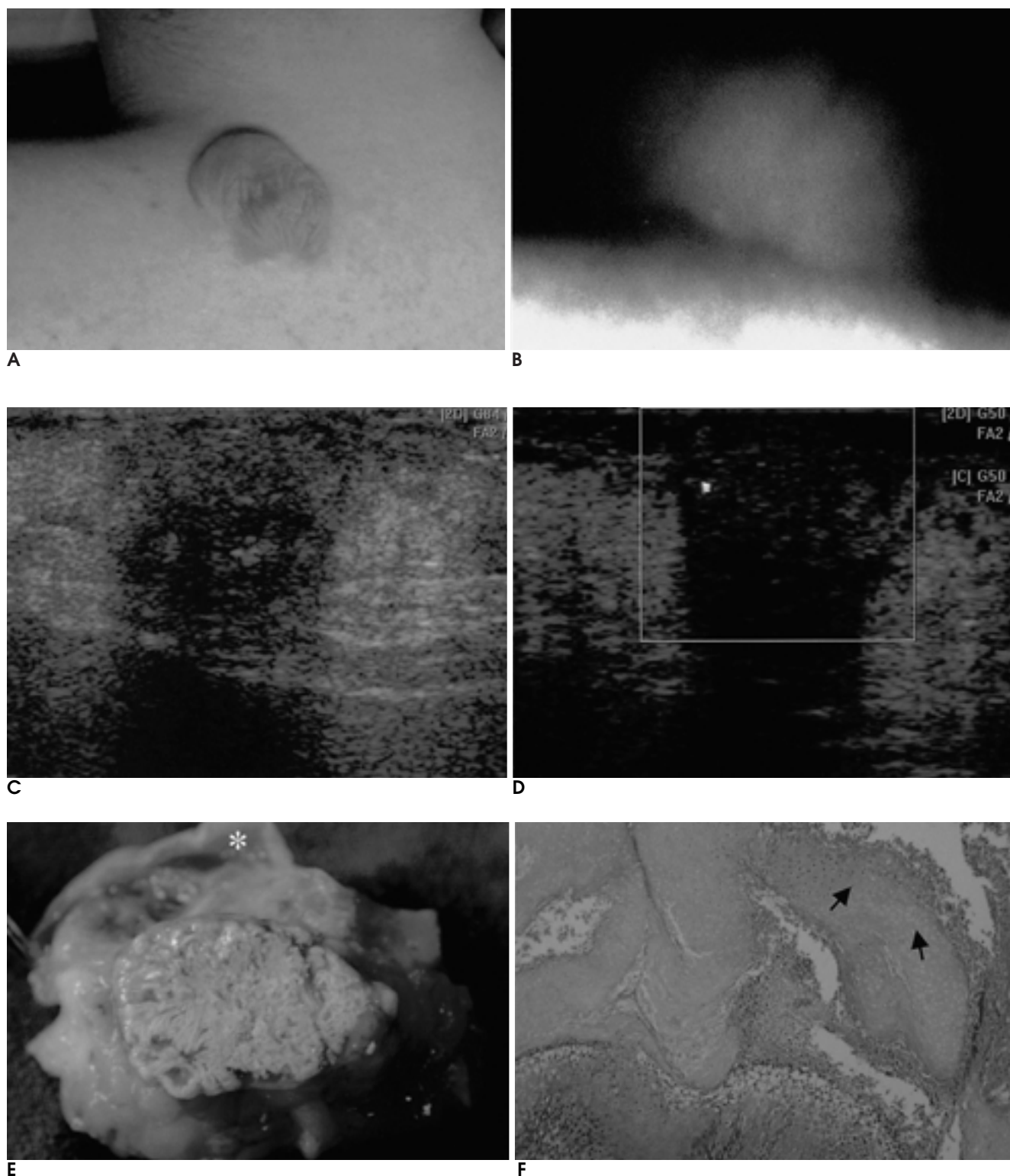


Fig. 1. A 22-year-old man with pilomatricoma.

A. On physical examination, a bullous soft tissue mass with reddish discoloration is in the left interscapular area.

B. Plain radiograph shows the soft tissue mass with tiny calcifications.

C, D. Ultrasonograms demonstrate a heterogeneous hypoechoic mass located in the dermis. There is no vascularity. Multiple echogenic foci show posterior shadowings.

E. Cross section of the specimen shows calcified tissue arranged in a circular pattern. The mass is covered with the bullous skin (asterisk).

F. Photomicrograph of histologic preparation (hematoxylin-eosin, original magnification $\times 100$) shows the characteristic shadow cells (arrows). There is transition from nucleated basophilic cells to enucleated shadow cells.

and the mass was covered with bullous skin (Fig. 1E). Pathologically, it consisted of pale, cornified cells, the nuclei of which had almost disappeared, and on the basis of these and other pathologic findings was confirmed as a typical pilomatricoma (Fig. 1F).

Discussion

In 1880, Malherbe and Chenantais first described a certain benign tumor of the skin, thought to be derived from sebaceous glands and arising in the subcutis, and gave it the name, a calcifying epithelioma, wishing to correctly describe its origin in hair follicle matrix cells, Forbis and Helwig renamed the neoplasm 'pilomatricoma' (4).

Pilomatricoma has a stony consistency, which is its pathognomonic feature. The tumor slides freely over the underlying layer, and the overlying skin may have a reddish or bluish discoloration. It is well-circumscribed, somewhat friable when grasped firmly, and has a grayish-tan appearance. Patients are usually asymptomatic, but certain reports have mentioned that there may be associated pain during episodes of inflammation or ulceration (5).

Histopathologically, the tumor consists of irregularly shaped islands of epithelial cells arranged in a circular pattern. These islands are composed of two cell types: (1) basaloid cells that are nucleated and arranged along the periphery of the tumoral islands, and (2) shadow (ghost) cells that are enucleated and located in the center of the islands. Calcification occurs in 70 - 85% of cases (5, 6). A malignant variety of the neoplasm, with distant metastasis to the lung, bone, brain, skin, and abdominal organs, has been reported (7).

Numerous studies have demonstrated that pilomatricomas most commonly occur in the head and neck, followed by the upper extremity, trunk, and lower extremity (8). A review of the literature has revealed female preponderance in those aged under 21. The neoplasm is mostly solitary, though synchronous multiplicity accounts for 2 - 3.5% of reported cases (3). A pilomatricoma can also be associated with Gardner syndrome, myotonic muscular dystrophy, sarcoidosis, skull dysostosis, Rubinstein-Taybi syndrome, and Turner syndrome.

The tumor can usually be diagnosed solely on the basis of its clinical features. However, various imaging modalities help the diagnostic process. Plain radiographs show nonspecific foci of calcification, while computed tomography depicts a well-marginated subcu-

taneous soft tissue mass adherent to the skin and with or without visible calcification (9). Magnetic resonance imaging demonstrates intermediate intensity at T2WI and slight enhancement at contrast-enhanced T1WI, mottled hypointense components corresponding to areas of calcification (10). Ichikawa et al. (10) reported a case of giant pilomatricoma that was depicted at angiography as a hypervascular mass. Ultrasonography, a relatively fast and noninvasive technique, reveals the presence of a well-defined, round, hyperechoic mass with a posterior dense acoustic shadow (6). A search of the literature has failed to locate a description of the ultrasonographic findings of this uncommon tumor in Korea, and this may well be the first such report to appear here. In our case, ultrasonography demonstrated a well-marginated heterogeneous, hypoechoic mass, unlike those described in the previous literature (6). The observed multiple echogenic foci with posterior acoustic shadowing indicated the presence of calcification. We believe that ultrasonographic findings mimicking a cartilaginous soft tissue mass, as in our case, are nonspecific, but that ultrasonographic findings supported by clinical information will help accurate diagnosis.

Since spontaneous regression of a pilomatricoma has never been observed, surgical excision is mandatory. Occasionally, overlying skin may adhere to the tumor, requiring simultaneous excision of both, as in the present case, where overlying bullous skin was excised along with the tumor. Most recurrences arise due to incompletely excised neoplasms (6), and to minimize this risk, wide resection is recommended.

In conclusion, pilomatricoma is a rare, benign skin tumor derived from the matrix cells of the hair follicle. It has a characteristic clinical presentation, and the clinical diagnosis should thus be suspected at physical examination. We suggest, however, that if ultrasonography reveals a well-marginated, subcutaneous hypoechoic calcified mass adherent to the skin, and its nature has not been clinically proven, pilomatricoma should be included in the differential diagnosis when the tumor mimicks a cartilaginous soft tissue mass.

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