



Ultrasonographic Findings of Pilar Sheath Acanthoma: A Case Report

모극초세포종의 초음파 소견: 증례 보고

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Pilar sheath acanthoma is a rare benign follicular hamartoma that presents with a central sinus containing keratinous material and is lined by epithelium. It typically occurs on the face, especially on the upper lip and forehead. In our case, the ultrasound (US) feature of pilar sheath acanthoma revealed a well-defined, oval hypoechoic nodule with hypoechoic capping within the dermis over the medial aspect of the calf. To the best of our knowledge, despite many reports on the clinicopathological aspects of pilar sheath acanthoma, this entity has not been well described in the radiologic literature, and US findings have not been documented. We report the US findings of a case of pilar sheath acanthoma on the calf.

Index terms

Acanthoma
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INTRODUCTION

Pilar sheath acanthoma is a rare benign follicular hamartoma. Mehregan and Brownstein (1) first reported 9 cases in 1978 that included a spectrum of lesions composed of lobules of follicular epithelium with infundibular maturation. To date, approximately 50 cases have been described worldwide, and only 5 cases have been reported in Korean dermatology journals until 2009 (2). In 1993, Ackerman et al. (3) investigated 21 cases and discovered that in addition to infundibular differentiation, isthmus, and, rarely, sebaceous ducts, apocrine glands, the inner root sheath, and bulb differentiation may be involved. Therefore, Hurt suggested that "pilar sheath acanthoma" should be renamed "lobular infundibuloisthmicoma". To the best of our

knowledge, despite many reports on the clinicopathological aspects of pilar sheath acanthoma, this entity has not been well described in the radiologic literature, and ultrasound (US) findings have not been documented. Here, we report the first US findings of pilar sheath acanthoma located on the medial aspect of the calf.

CASE REPORT

A 56-year-old male patient was hospitalized for treatment of a painful palpable nodule with a central pore-like opening on the medial aspect of the right calf. He had undergone surgery for treatment of varicose veins on the right leg 3 years ago, and then he experienced pain radiating from the operation site to the right

toes. Since the past 10 months, a solitary erythematous hyperpigmented papule measuring approximately 1 cm with pus discharge was visible on the medial aspect of the right calf (Fig. 1A, B). On gross examination, the clinical differential diagnosis included an epidermoid cyst, an open comedo, or a dilated pore of Winer.

US was performed to identify the location and to characterize

the nodule. A well-defined oval hypoechoic nodule, which measured about 0.65×0.35 cm, with hypoechoic capping within the dermis over the medial aspect of the right calf was noted on gray scale US (Fig. 1C). This hypoechoic nodule communicated with hypoechoic capping by a narrow hypoechoic neck. The gray scale US also revealed no posterior acoustic enhancement.

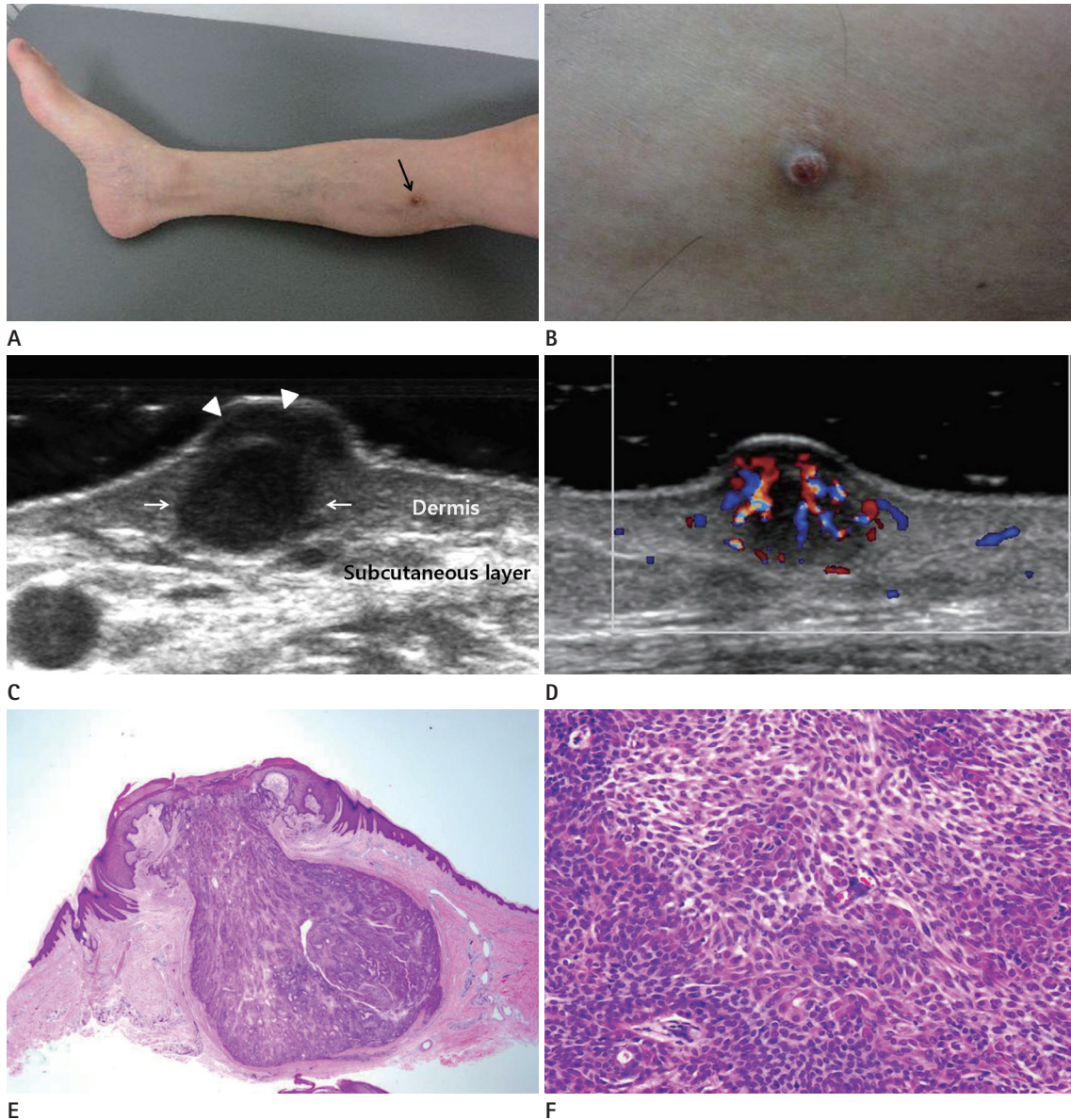


Fig. 1. Pilar sheath acanthoma on the right calf in a 56-year-old man.

A, B. Photograph of the leg. Medial (**A**) and close-up (**B**) views of the papule on the right calf. A solitary, erythematous to hyperpigmented papule with a central punctum (arrow in **A**) is seen on the medial aspect of the right leg.

C. Gray scale ultrasonography shows a 0.65×0.35 cm, well-defined oval hypoechoic nodule (arrows) with hypoechoic capping (arrowheads) within the dermis over the medial aspect of the right calf.

D. A color Doppler image shows internal hypervascularity within the nodule.

E. A photomicrograph (hematoxylin-eosin stain, $\times 10$) shows a lobular patulous tumor that was connected to the epidermis and the dermis.

F. A photomicrograph (hematoxylin-eosin stain, $\times 200$) shows tumor cells consisting of mixed blue-gray (infundibular) and pink (isthmic) corneocytes. The findings are compatible with pilar sheath acanthoma.

This lesion showed internal hypervascularity on color Doppler images (Fig. 1D). According to US findings, the provisional diagnosis was an epidermoid cyst or pilomatricoma.

An excisional biopsy was performed to confirm the diagnosis. Histopathological examination revealed a lobular patulous tumor that was connected to the epithelium, and tumor cells consisting of mixed blue-gray (infundibular) and pink (isthmic) corneocytes were noted in the dermis. The findings were compatible with pilar sheath acanthoma (Fig. 1E, F).

DISCUSSION

Pilar sheath acanthoma is a rare benign follicular hamartoma. Middle-aged and elderly individuals are commonly affected. Pilar sheath acanthoma is characterized by a small (5 to 10 mm in diameter), solitary, painless, skin-colored papule located on the head or neck, particularly around the upper lip (4). A central pore that is occasionally plugged with keratin is often present (4).

Diseases that should be considered in the clinical differential diagnosis include an epidermoid cyst, an open comedo, trichofolliculoma and a dilated pore of Winer. From the pathological viewpoint, it is difficult to differentiate pilar sheath acanthoma from trichofolliculoma and a dilated pore of Winer due to similar histopathological findings. A common histopathological feature among these tumors is the central sinus, which contains keratinous material, and is lined by epithelium that is continuous with the surface epidermis (5). In pilar sheath acanthoma, abortive hair follicle-like structures are present but do not show a high degree of differentiation; hair shafts are absent within the central cavity, prominent fibrovascular stroma is lacking, and several small cysts within the mass of the cyst wall are visible. In trichofolliculoma, small hair follicles radiate from the wall of the central infundibular cyst (6). A dilated pore of Winer is a patulous follicle that contains hair. Numerous small digitate projections radiate from the follicular epithelium into the surrounding connective tissue (4, 5). Mehregan suggested that pilar sheath acanthoma is less mature than a dilated pore of Winer but more mature than a tumor of the follicular infundibulum (1). Treatment consists of surgical excision or electrodesiccation and curettage (4).

In this case, the US findings revealed a non-specific soft tissue nodule that was a well-defined, hypoechoic nodule with hypoechoic capping and had hypervascularity within the dermis.

According to the radiologic-pathologic correlations, a lobular patulous tumor that was connected to the epithelium was noted in the dermis, and it had a central pore-like opening that exhibited a hypoechoic nodule with a narrow hypoechoic neck and erosive or ulcerative epidermis with hypoechoic capping on US. When there are superficial soft-tissue masses, the possible diagnosis is variable. Superficial soft-tissue masses may be classified into one of the following general diagnostic categories: mesenchymal tumors, skin appendage lesions, metastatic tumors, other tumors and tumorlike lesions, and inflammatory lesions (7). Also, it is important to identify the exact location of the mass in the differential diagnosis (7). Skin appendage lesions, which include an epidermoid cyst, pilomatricoma, cystadenoma, cylindroma, and syringoma, originate in the epidermis and dermis (7). In this case, US revealed a well-defined oval hypoechoic nodule with hypoechoic capping within the dermis. Thus, we considered that the possible diagnosis was an epidermoid cyst or pilomatricoma. The epidermoid cyst is filled with loosely packed lamellae of keratin; the walls resemble those of a follicular infundibulum (8). At US, the cyst appears as a circumscribed circular or oval hypoechoic mass, often in association with a hair follicle (7). Lee et al. (8) reported the US findings of an epidermoid cyst, and they stated that 96% of the lesions showed posterior sound enhancement and 83% of the lesions showed no signal on color Doppler image. Pilomatricoma is a benign superficial tumor of the hair follicle (9). It is the most common solid cutaneous tumor in patients aged 20 years and younger. US features of pilomatricoma are a well-defined oval mass with strong posterior acoustic shadowing at the junction of the dermis and subcutaneous fat with focal thinning of the overlying dermis (10). Hwang et al. (9) reported that US revealed a well-defined oval hyperechoic nodule with posterior acoustic shadowing. The reason for posterior acoustic shadowing is probably that pilomatricoma shows frequent calcification. Thus, we can differentiate pilar sheath acanthoma from epidermoid cyst or pilomatricoma by some US features, such as posterior sound enhancement or posterior acoustic shadowing.

Pilar sheath acanthoma can be considered a possible diagnosis when a well-defined dermal nodule with a central pore-like opening is a hypoechoic nodule with hypoechoic capping and it shows hypervascularity on US images.

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모극초세포종의 초음파 소견: 증례 보고

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모극초세포종은 드물게 발생하는 양성인 모낭 과오종으로 내부의 중심 동굴(central sinus)에 각화성 물질을 포함하고 있으며 상피세포로 피복되어 있다. 병변은 얼굴에 대부분 발생하며 특히 상구순과 이마에 호발하는 것으로 알려져 있다. 저자들이 경험한 모극초세포종 증례의 초음파 소견에서 경계가 좋은 타원형의 저에코성의 결절이 저에코성 모자를 쓴 양상으로 오른쪽 종아리의 내측 진피층에서 보였다. 우리가 아는 한, 모극초세포종의 임상 병리학적인 관점에 관한 많은 증례 보고들이 있음에도 영상의학의 문헌에서는 이에 대한 기술이 부족하며 특히 초음파 소견은 아직 보고된 바 없다. 이에 저자들은 하지의 모극초세포종 1예를 경험하고 초음파 소견에 대한 고찰과 함께 보고한다.

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