

A Case of Pilomatricoma in the Upper Arm: Sonographic and MR Imaging Findings¹

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Pilomatricoma is an uncommon benign skin neoplasm arising from the hair matrix. The radiologic features of this uncommon tumor have occasionally been described in the literature however, to the best of our knowledge, there have been no reports in Korea on the sonographic and MR imaging findings of pilomatricoma involving the upper arm. In this article, we describe the imaging findings of a case of pilomatricoma involving the upper arm, with the emphasis being placed on the sonographic and MR imaging findings and a review of the literature.

Index words : Soft tissues, neoplasms

Soft tissues, US

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Pilomatricoma is an uncommon benign skin neoplasm arising from the hair matrix (1 - 3). The preoperative diagnosis of this entity may be difficult because of its multiple manifestations (1), especially if the examining physician is unfamiliar with them. There have been some reports in the radiological literature which included the CT (2, 4, 5) and MR (2, 6 - 8) imaging findings of pilomatricoma however, to the best of our knowledge, there have been no reports in Korea on the sonographic and MR imaging findings of pilomatricoma involving the upper arm. Knowledge of the imaging findings of this uncommon tumor would provide clinicians with useful information and facilitate the correct management of this condition. In this article, we describe the imaging findings of a case of pilomatricoma involving the upper arm, with the emphasis being placed on the

sonographic and MR imaging findings and a review of the literature.

Case Report

A 10-year-old boy presented with a palpable mass in the anterior aspect of the right proximal upper arm, which had been noticed 1 year previously. The mass had remained static in size after an initial period of slow growth. Physical examination revealed a 1.5 × 1.5 cm sized, firm, tender mass in the right upper arm. The mass was freely movable, and the overlying skin was normal in color and texture. The remainder of the physical examination was unremarkable.

Plain radiography showed a smooth, subcutaneous soft tissue lesion without any intralesional calcifications at the anterior aspect of the right proximal upper arm. Ultrasonography revealed a well-defined subcutaneous mass with an echogenic center and a thin hypoechoic rim. There was posterior acoustic shadowing from the echogenic center, indicating the presence of tiny calcifications (Fig. 1). MR imaging showed a well-defined subcutaneous mass in the anterior aspect of the right

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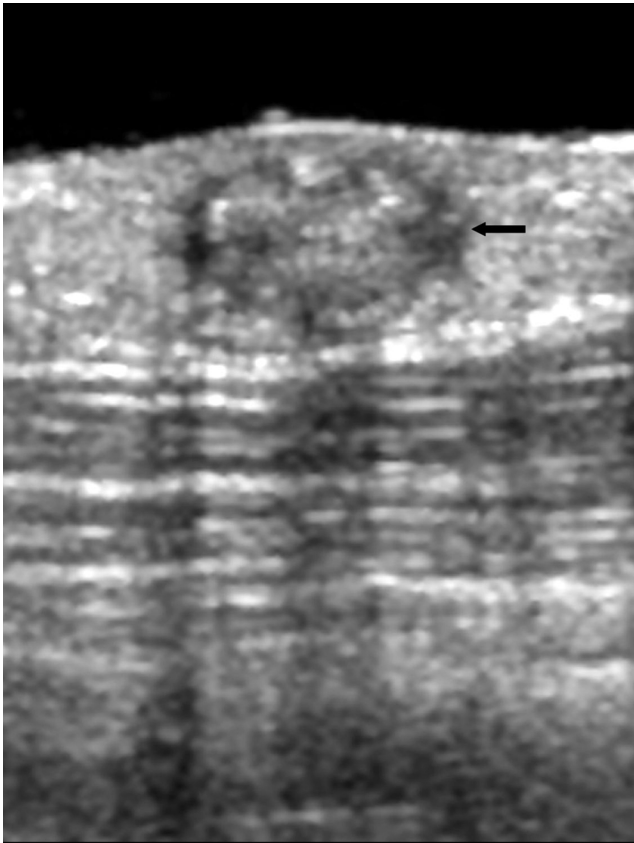


Fig. 1. A 10-year-old boy who had a palpable mass in the anterior aspect of the right proximal upper arm for 1 year. Ultrasonography demonstrates a well-defined subcutaneous mass (arrow) with an echogenic center and a thin hypoechoic rim. There is posterior acoustic shadowing from the echogenic center indicating the presence of tiny calcifications.

proximal upper arm, adjacent to the cephalic vein. The mass was isointense on the spin-echo (SE) T1-weighted image as compared with the muscle, and had a low signal intensity on the fast SE T2-weighted image as compared with the fat. The gadolinium-enhanced T1-weighted image showed homogenous enhancement of the mass without a central necrotic portion (Fig. 2A - C).

Surgical excision was performed. Microscopic examination of the pathologic specimen showed centrally located shadow cells and peripheral basaloid cells. Some areas of scattered small calcific deposits within the nests

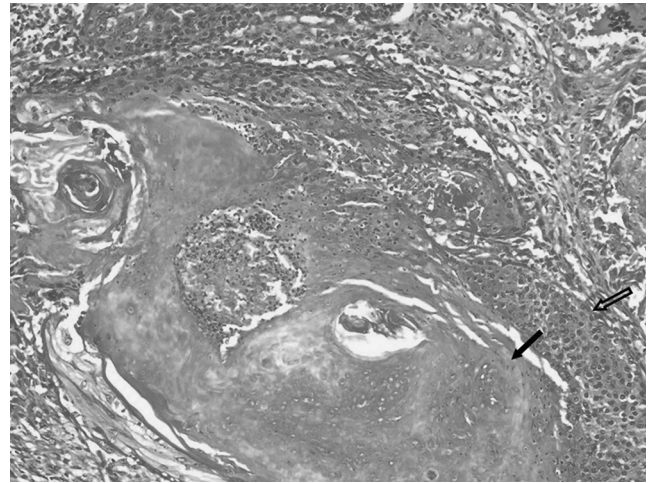


Fig. 3. Photomicrograph shows centrally located shadow cells (arrow) and peripheral basaloid cells (open arrow). Small calcific deposits are present (not shown here). (Hematoxylin-eosin stain; original magnification $\times 100$).

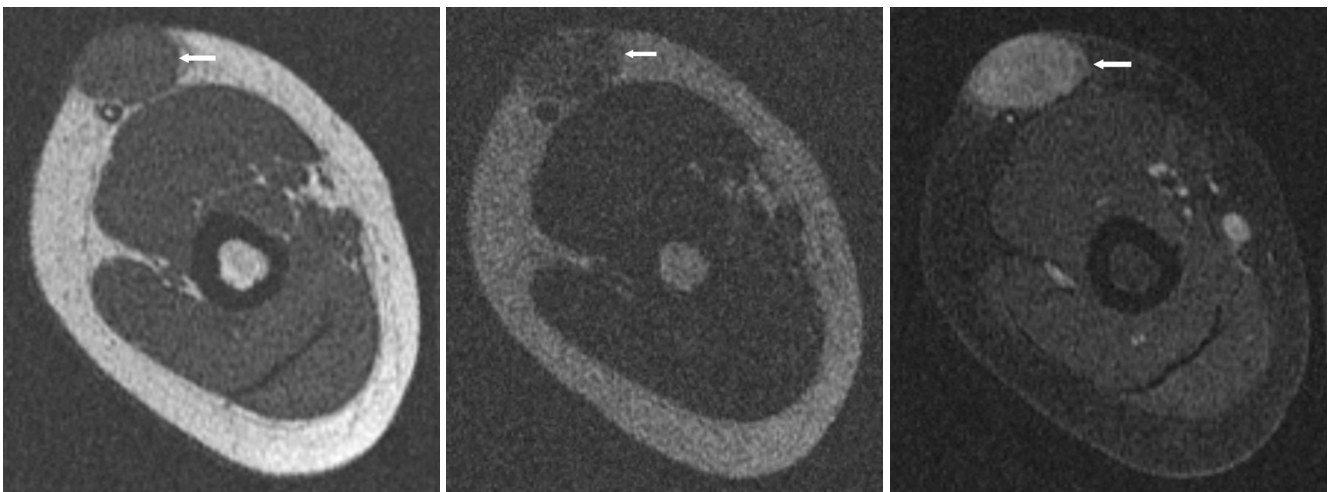


Fig. 2. **A.** Axial T1-weighted (500/10) spin-echo MR image demonstrates a well-defined subcutaneous mass (arrow) in the anterior aspect of the right proximal upper arm, adjacent to the cephalic vein. The mass is isointense as compared with the muscle. **B.** Corresponding axial T2-weighted (3000/96) fast spin-echo MR image shows a homogenous low signal intensity mass (arrow) without any surrounding edema. **C.** Gadolinium-enhanced axial T1-weighted (600/10) spin-echo MR image obtained with fat saturation shows the homogenous enhancement of the mass without a central necrotic portion.

of shadow cells were noted. The microscopic findings of the pathologic specimen were consistent with the diagnosis of pilomatricoma (Fig. 3).

Discussion

Pilomatricoma, which is also called calcifying epitheliomas of Malherbe, is an uncommon benign skin neoplasm that is derived from the hair matrix (1). This tumor is more common in children and young adults, and most cases of this disorder (approximately 60%) occur in the first two decades of life (2). In our case, the age of presentation was similar to that of the majority of other cases described in the literature.

In terms of the overall incidence of this disorder, the female-to-male ratio is 3:2 and this ratio increases to 5:2 when only patients who are less than 20 years old are taken into consideration. Approximately half of all pilomatricomas occur in the head and neck, while other sites of predilection are the upper extremity, trunk and lower extremity, in order of decreasing frequency (1, 2). Clinically, the mass usually manifests as a solitary, slow-growing, firm dermal nodule, which is often accompanied by inflammation or discoloration of the overlying skin (3). In most cases, the diameter of the tumor ranges from 0.5 to 3 cm, but a case of giant pilomatricoma with a maximum diameter as large as 18 cm has been reported (2). Most occurrences are solitary lesions, but multiple tumors occur in 2 - 3.5% of cases. The latter have been associated with Gardner's syndrome, myotonic muscular dystrophy, sarcoidosis, and skull dysostosis (9).

Histopathologically, pilomatricoma is a deep, subcutaneous tumor occurring between the dermis and hypodermis, and is separated from the epidermis by a connective tissue capsule. Epithelial islands are embedded in a cellular stroma. These epithelial cell islands are arranged in a circular configuration, with nucleated basophilic cells being found on the periphery and enucleated shadow or ghost cells in the center (3, 9). Calcification and bone metaplasia are observed in 69-85% and 15 % of cases, respectively (9).

There have been some reports in the radiologic literature which included the CT (2, 4, 5) and MR (2, 6 - 8) imaging findings of pilomatricoma. In these reports, the CT findings of pilomatricoma were described as well-defined subcutaneous masses adherent to the skin with various amounts of calcification (2, 4, 5). De Beuckeleer et al. (7) reported two cases of pilomatricoma in the fore-

arm that were of homogenous intermediate signal intensity on the SE T1-weighted images and predominantly low-to-intermediate signal intensity on the T2-weighted images. In our case, which involved pilomatricoma in the upper arm, the MR imaging findings were similar to those of the previously reported cases. Hoffmann et al. (6) described the MRI findings in a case of pilomatricoma in the neck of a child. The T2-weighted images showed bands of hyperintense signal radiating away from a low signal intensity center towards the periphery. The authors suggested that the hyperintense bands noted on the T2-weighted images might correspond to the basaloid bands present on the histologic specimens. On the other hand, Masih et al. (8) put forward another possible explanation for this observation, suggesting that the high signal intensity reticulations might represent edematous stroma surrounding basaloid cells.

The degree of enhancement of the tumor on the CT scans and MR images varied (2, 4, 6 - 8), probably due to the different degrees of epithelial cell components, and the variable amounts of fibrous tissue stroma and vascular tissues (4). The sonographic findings of a pilomatricoma have also been previously reported (6, 7, 10). Ultrasonography revealed a well-defined subcutaneous mass with an echogenic center and a thin hypoechoic rim. There was posterior acoustic shadowing from the echogenic center, indicating the presence of tiny calcifications (7, 10). The echogenic center corresponds to the central islands of epithelial cells and the hypoechoic rim corresponds to the connective tissue capsule (10). In our case of pilomatricoma in the upper arm, the sonographic findings were similar to those published in these previous reports. The authors believe that ultrasonography, which is a relatively fast and noninvasive technique, allows for the accurate diagnosis of pilomatricoma in childhood through the identification of the tiny calcifications.

In conclusion, pilomatricoma should be included in the differential diagnosis of a well marginated subcutaneous mass containing calcifications in the upper arm, especially if such a mass occurs in a child or young adult.

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