








Merkel Cell Carcinoma of the Trunk: Two Case Reports and Imaging Review

몸통에 생긴 메르켈 세포암종: 2예 증례 보고 및 영상 소견 고찰

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Merkel cell carcinoma (MCC) is a rare malignant cutaneous tumor primarily located in the head and neck. We report the imaging features of pathologically confirmed MCC in the trunk. On US, MCC showed heterogeneous echogenicity with perpendicular hypoechoic linear bands that resembled “columns of smoke” in the skin and subcutaneous layers as well as prominent vascularity. On MRI, the tumor showed hypointensity on T1-weighted images and hyperintensity on proton density and T2-weighted images with linear low-signal bands in the skin and subcutaneous layers as well as intense enhancement on T1-enhanced images. Although MCC has nonspecific imaging features, these characteristics may be helpful for the early diagnosis of this disease.

Index terms Merkel Cell Carcinoma; Skin; Ultrasonography; Magnetic Resonance Imaging

INTRODUCTION

Merkel cell carcinoma (MCC) is an uncommon, highly malignant cutaneous tumor, primarily located in the head and neck. It is characterized by rapid growth, regional nodal or distant metastases, and a high recurrence rate. MCC has a nonspecific clinical presentation, and its diagnosis is confirmed by histopathological and immunohistochemical studies (1-4). Owing to the rarity of the neoplasm and insufficient awareness, imaging features have been reported only in case reports and a few case series (4-9). Herein, we report the imaging fea-

tures of MCC located on the back and buttock. We also included a literature review of the imaging features.

CASE REPORT

CASE 1

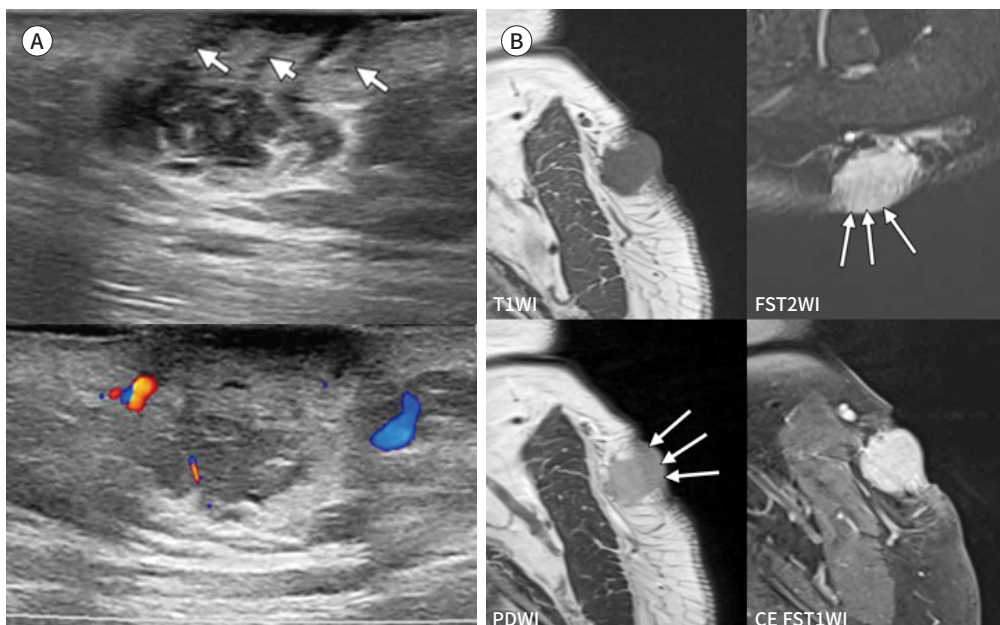
A 79-year-old female presented with a growing mass on the left upper back. The mass measured 2.0 cm × 1.5 cm. It was cherry red in color, firm, minimally fixed, and non-tender. On US, the mass showed heterogeneous echogenicity with perpendicular hypoechoic linear bands that resembled “columns of smoke” in the skin and subcutaneous layers. Color Doppler US revealed mild vascularity, with arterial flow evident in the mass (Fig. 1A). On MRI, the mass was hypointense on T1-weighted images and hyperintense with linear bands in the skin and subcutaneous layers on proton density and T2-weighted images. It also showed intense heterogeneous enhancement after contrast injection. Peritumoral enhancement with skin thickening was also observed (Fig. 1B). Preoperative radiological differential diagnoses included malignant superficial soft tissue or vascular tumors. The patient underwent wide excision of the mass, with a clear resection margin. The final pathological diagnosis was MCC (Fig. 1C). There was no residual tumor or metastasis on 18F-fluorodeoxyglucose PET-CT.

Fig. 1. Merkel cell carcinoma of the left upper back in a 79-year-old female.

A. US (upper panel) shows a heterogeneous echogenicity mass with perpendicular hypoechoic linear bands (arrows) that resemble “columns of smoke” in the skin and subcutaneous layers. In the color Doppler mode (lower panel), there is mild intratumoral and peritumoral vascularization.

B. This mass is hypointense on sagittal T1WI and hyperintense on sagittal proton density and axial FST2WI. It contains linear low-signal bands (arrows) in the skin and superficial subcutaneous layers, and shows intense heterogeneous enhancement on T1-enhanced image; peritumoral enhancement and skin thickening are also observed.

CE = contrast-enhanced, FS = fat-suppressed, PDWI = proton density-weighted image, T1WI = T1-weighted image, T2WI = T2-weighted image



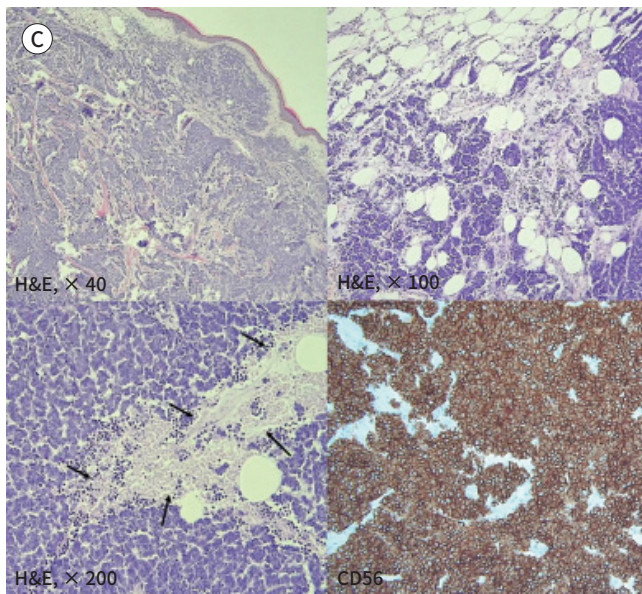


Fig. 1. Merkel cell carcinoma of the left upper back in a 79-year-old female.

C. Photomicrograph of the histological section of a tumor shows diffusely infiltrating tumor nests under the epidermis (upper left image) and infiltration of the tumor cells into the subcutaneous fat tissue (upper right image) as well as hyperchromatic small nuclei and necrosis in the tumor cells (arrows) (lower left image); furthermore, the tumor cells are positive for neuroendocrine markers such as CD56 (lower right image) and chromogranin (not shown). H&E = hematoxylin and eosin

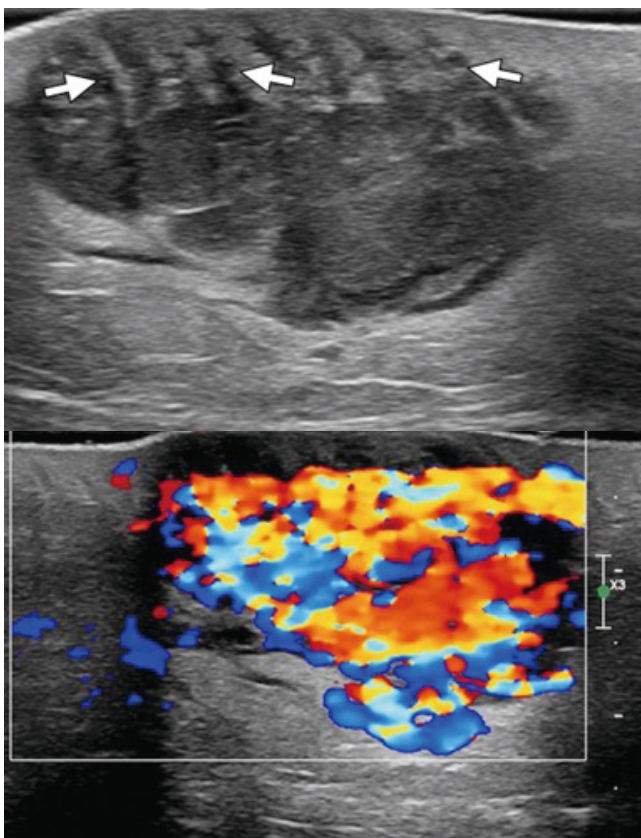


Fig. 2. Merkel cell carcinoma of the right buttock in a 78-year-old female.

US (upper panel) shows a heterogeneous echogenicity mass with perpendicular hypoechoic linear bands (arrows) that resemble “columns of smoke” in the skin and subcutaneous layers. Color Doppler mode (lower panel) shows prominent intratumoral vascularization.

CASE 2

A 78-year-old female presented with a palpable mass on the right buttock. The mass measured 3.3 cm × 1.9 cm. The mass was oval, firm, fixed, and non-tender. On US, the mass showed heterogeneous echogenicity, with hypoechoic linear bands and prominent hypervascularity (Fig. 2). A vascular tumor or lymphoma was considered as differential diagnoses.

The patient underwent wide excision of the mass. The final pathological diagnosis was MCC. No metastasis was observed on chest or abdominal CT. After the surgery, the patient underwent local radiotherapy.

This study was approved by the Institutional Review Boards of the Eulji University Nowon Eulji Medical Center (IRB No. EMCS 2022-10-018) and Daegu Fatima Hospital (IRB No. DFH 2022-10-003).

DISCUSSION

MCC is an aggressive neuroendocrine carcinoma of the skin that originates from Merkel cells located at the base of the epidermis, close to the epidermal-dermal interface, and frequently extends into subcutaneous fat tissue. MCC predominantly involves the head and neck (43%–50%), followed by the extremities (39%–40%) and trunk (9%–11%). MCC tends to affect men more than women in a ratio of 2:1. Most affected individuals are older than 50 years. The typical clinical presentation is a 2- to 4-cm, firm, non-tender, red to violaceous nodule, located in areas of the body that are often exposed to sunlight. The most clinically relevant prognostic factors include tumor size and the presence of regional lymph node metastasis or hematogenous metastasis to other organs including the liver, bone, lung, and brain (1-3).

Standardized imaging guidelines for the diagnosis of MCC have not yet been established. Common imaging features of primary MCCs include cutaneous or subcutaneous nodules and focal or diffuse skin thickening. MCCs can appear as perifascial muscular, or intramuscular metastases. Necrosis is common. In contrast, calcifications are generally invisible (2-4).

On US, MCC appears as a predominantly hypoechoic solid mass with variable degrees of heterogeneity that extends to the subcutaneous fat layer with different levels of posterior acoustic enhancement and peritumoral skin thickening. Recently, it was reported that hypoechoic linear bands shaped like “columns of smoke” are typical features of MCC (6). Histologically, the hypoechoic linear bands perpendicular to the skin in our cases appeared to correspond to the remaining thick fibrous collagen bands in the dermis and superficial subcutaneous layer. MCC may also show prominent and chaotic hypervascularity in the arterial vessels (6-8). Hypoechoic linear bands with vascularity were evident in both of our cases.

On CT, cutaneous fat stranding adjacent to the primary site of MCC may suggest engorgement and edema from lymphatic invasions (2). On MRI, MCC shows hypo- to iso-intensity on T1-weighted images and iso- to hyper-intensity on T2-weighted images, reflecting the high cellularity of small, round cell tumors. The tumor may also demonstrate low apparent diffusion coefficient values owing to its high cellular density. It typically shows rich vascularization on pathological analysis, which can be due to hypertrophic peritumoral and intratumoral vessels. Furthermore, the tumor can show strong contrast enhancement on MRI. Peritumoral focal skin thickening and cutaneous or subcutaneous nodules are also often observed. These findings correspond to the histological findings of increased vascularity in the stromal tissues of the tumor and between the tumor cells. Radiographic, CT, and MRI images of MCC are similar to those of other small cell carcinomas (1, 2, 4, 9).

The differential diagnosis should include lymphoma, owing to the increased cellularity, as well as squamous cell carcinoma, non-pigmented melanoma, and dermatofibrosarcoma pro-

tuberans. Due to its rarity, radiologists are unaware of evocative imaging features and usually do not consider MCC in the differential diagnosis of soft tissue tumors (2-4, 9). Although the definitive diagnosis of MCC is based on characteristic pathological findings, imaging features of a mass in the dermal and subcutaneous layers with linear bands that resemble “columns of smoke” and marked vascularity may be suggestive of MCC. Awareness of these features may be helpful for the early diagnosis of MCC.

Author Contributions

Conceptualization, O.H.Y., C.Y.S.; data curation, all authors; investigation, O.H.Y., K.D.; resources, all authors; supervision, C.Y.S.; visualization, O.H.Y., K.D.; writing—original draft, O.H.Y., C.Y.S.; and writing—review & editing, all authors.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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메르켈 세포암종은 드문 악성 피부종양으로 주로 두경부에 발생한다. 저자들은 몸통에 발생한 병리학적으로 확진된 메르켈 세포암종의 그레이스케일 및 색 도플러 초음파 소견과 자기공명영상 소견을 보고하고자 한다. 메르켈 세포암종은 초음파상 비균질 에코를 보이고 피부 및 피하층에 수직방향으로 저에코의 '연기기둥' 같은 선형 띠를 가지고 있었으며 내부에 혈류가 약간 증가되어 보였다. 한편 T1강조 자기공명영상에서 저신호강도, T2 및 양자밀도강조 영상에서 같은 층에 저신호강도의 선형띠와 함께 고신호강도로 보였으며, 강한 조영증강을 보였다. 메르켈 세포암종의 영상 소견이 비특이적으로 알려져 있지만, 이러한 특징적인 소견들은 이 질환을 조기 진단하는데 도움이 될 수 있을 것이다.

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