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MR Findings of Trichilemmal Carcinoma:

A Case Report[1]

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A trichilemmal carcinoma is a rare superficial tumor occurring on the sun-exposed regions of older individuals. We examined a 79-year-old female with a large mass located in the right posterolateral aspect of the patient's neck. The MR images showed a central cystic portion with intense peripheral enhancement. The histopathologic examination of the resected specimen revealed a trichilemmal carcinoma. Here, we report the MR findings of the trichilemmal carcinoma in this patient.

Index words : Carcinoma
Skin appendage
Magnetic resonance (MR)

A trichilemmal carcinoma is a rare, cutaneous adnexal neoplasm which can occur on sun-exposed areas of the body. It is the malignant counterpart of trichilemmoma (1). It is however considered to be a low-grade malignant carcinoma due to its rare incidence of recurrence or metastasis (2).

There have been many reported cases of trichilemmal carcinoma with pathologic findings (1-6); however, to the best of our knowledge, the MR findings of trichilemmal carcinoma have not previously been reported. In this case report, we will describe the MR findings of a trichilemmal carcinoma.

Case Report

A 79-year-old female was referred to our hospital with a ten-month history of a large protruding mass in the right posterolateral aspect of her neck. Upon physical examination, the mass was noted to be fixed and hard. The dimensions of the soft tissue mass were approximately $7 \times 11 \times 7$ cm. The patient did not complaint and no tenderness was experienced. Also, the patient had no neurologic symptoms. Discharge was noted from the central portion of the mass and the initial clinical impression was identified as dermatofibrosarcoma protuberans.

On a plain radiography, the soft tissue mass revealed no calcification (Fig. 1A). An ultrasonography was performed using a multi-frequency (5- to 12-MHz) linear-array transducer attached to an iU-22 scanner (Philips Medical Systems). Gray-scale imaging showed an echogenic mass with multifocal hypoechoic and hyperechoic foci (Fig. 1B). Color Doppler imaging revealed a vascular signal in the peripheral portion of the mass (not shown).

MR imaging was performed using a 1.5-T MR system (ACHIEVA, Philips Medical Systems, Netherlands). The mass lesion showed heterogeneously low signal intensity on fast spin echo (FSE) T1-weighted images (Fig. 1C), and heterogeneously high signal intensity on FSE T2-weighted images in relation to the signal intensity of the

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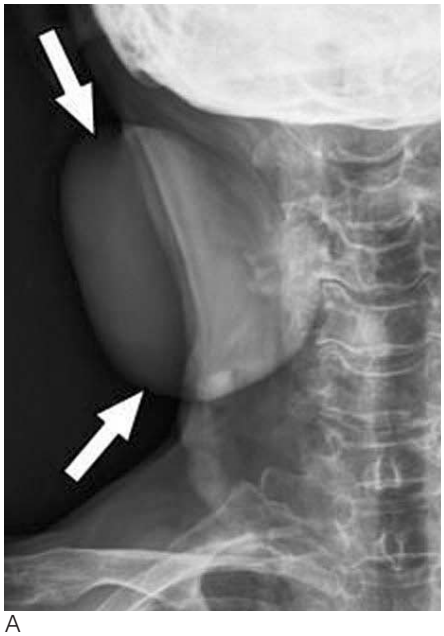
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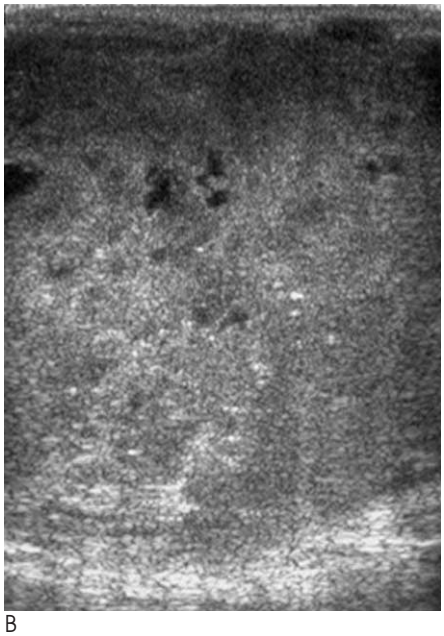
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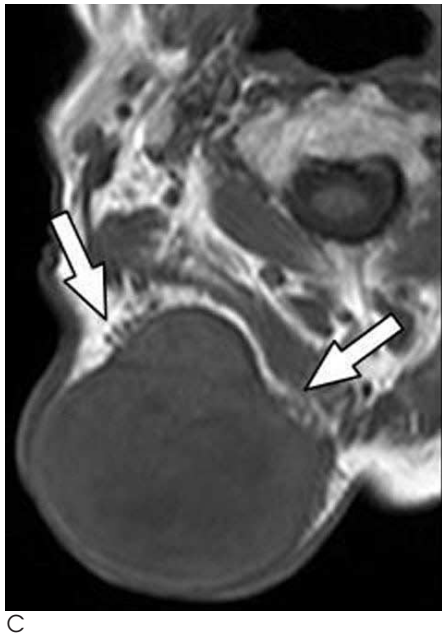
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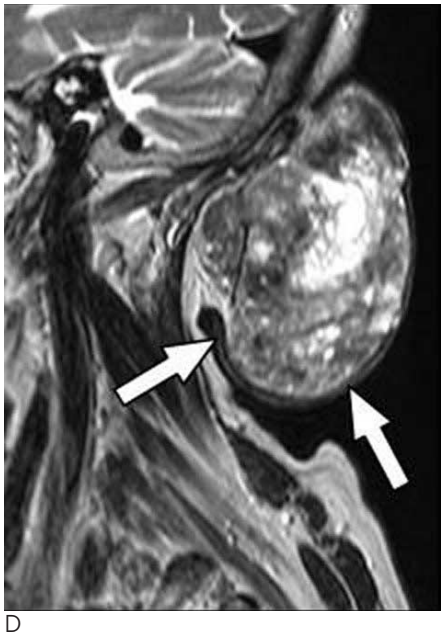
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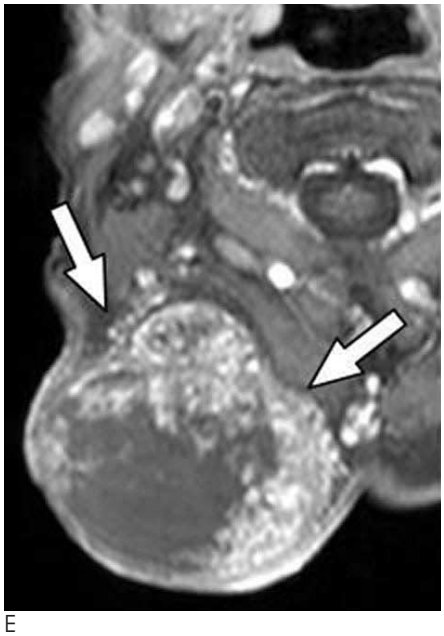
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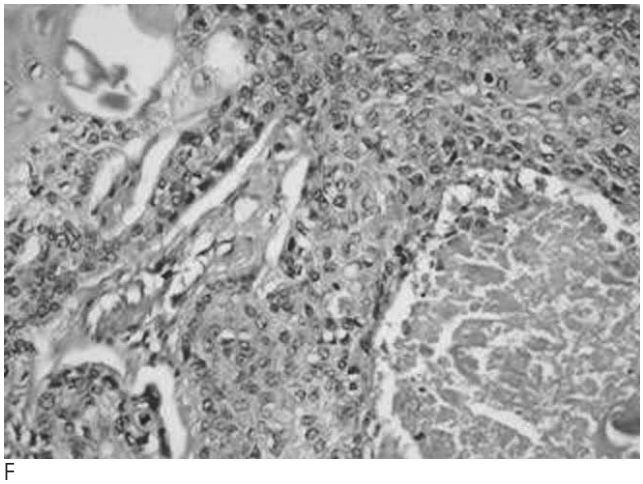
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Fig. 1. A 79-year-old female with a 7×11×7 cm mass in the right posterolateral aspect of the neck.
A. A C-spine AP shows a large mass without calcification on the right side of the patient's neck (white arrows).
B. A gray-scale US image shows an echogenic mass with multifocal hypoechoic and hyperechoic foci.
C. An axial FSE T1-weighted MR image (TR/TE = 531/10) shows a heterogeneously low signal intensity in the mass (white arrows).
D. A sagittal FSE T2-weighted MR image (TR/TE = 3500/120) shows heterogeneously high signal intensity in the mass (white arrows).
E. An axial FSE T1-weighted MR image (TR/TE = 702/10) obtained after MR contrast injection, shows diffusely intense enhancement in the peripheral portion and non-enhancement in the central portion of the mass (white arrows). The mass is located at the superficial portion of the right trapezius muscle with no evidence of invasion.
F. H & E staining (×200) shows proliferating lobules of squamous epithelium with multiple central areas of trichilemmal keratinization and neoplastic cells with moderate nuclear atypia and atypical mitosis.

adjacent muscles (Fig. 1D). Prominent enhancement was observed in the peripheral portion of the mass, whereas non-enhancement was observed in the central portion of the mass following the intravenous injection of MR contrast material (0.5 mmol/ml gadodiamide, Omniscan, GE) (Fig. 1E). No evidence of adjacent muscular invasion was noted. The radiologic impression was a soft tissue sarcoma, such as dermatofibrosarcoma protruberans or malignant fibrous histiocytoma (MFH).

An excisional biopsy was performed on the mass, which was easily and completely removed from the patient's neck. No evidence of invasion to the right trapezius muscle was noted. In addition, no tumor cells were observed at the resected margin on the frozen section.

Upon histopathologic examination, the specimen had fluid with debris and showed proliferating lobules of squamous epithelium with multiple central areas of trichilemmal keratinization and neoplastic cells with moderate nuclear atypia and atypical mitosis. These findings were consistent with the diagnosis of trichilemmal carcinoma (Fig. 1F).

A 12-month follow-up examination found that the patient has had no signs of lesion recurrence and has remained healthy.

Discussion

A trichilemmal carcinoma usually occurs as a solitary, indolent lesion on the sun-exposed areas of older individuals (1). The location of the lesion is most commonly confined to the scalp, face, neck, trunk, or upper extremities. There has been no gender predilection noted with this neoplasm (1). Trichilemmal carcinoma is the malignant counterpart of trichilemmoma and is distinct from the proliferating trichilemmal tumor which has a different origin (2).

A trichilemmal carcinoma usually presents as a mass, ranging from a flesh-colored telangiectatic plaque or papule, to a hyperkeratotic or ulcerated crater (2, 3). The tumor is very indolent, and the incidence of metastasis is extremely rare (4).

The pathogenesis of a trichilemmal carcinoma remains unclear, although the lesion distribution tends to suggest that sunlight plays an important role in the development of this tumor. However, a trichilemmal carcinoma has also been reported to occur on burn scars (5). Moreover, high-dose radiation therapy as well as numerous X-rays obtained for diagnostic purposes, has also been associated with this type of carcinoma (6).

Histologically, a trichilemmal carcinoma is mainly an intraepithelial tumor or is most commonly associated with an invasive component, which may involve the epidermis up to the subcutaneous fat. In general, this lesion appears in continuity with the epidermis and/or follicular epithelium (4). Unlike its benign counterpart, a trichilemmal carcinoma exhibits cytological atypia with a high mitotic index, which is indicative of an aggressive malignancy (1).

To the best of our knowledge, the MR imaging findings of a trichilemmal carcinoma have not been previously reported. Our case shows that a trichilemmal carcinoma can appear on MR imaging as a solid mass with cystic components. Signal intensities of the solid component of the tumor on MR images show slight hyperintensity on FSE T1- and T2-weighted images and substantial enhancement after contrast administration. Signal intensities of the cystic components show hypointensity on T1-weighted images and prominent hyperintensity on T2-weighted images. These findings are similar to the malignant transformation of a proliferating trichilemmal tumor (7) or of a dermatofibrosarcoma protuberans (8). Unfortunately, there are no characteristic MR imaging findings for the differential diagnosis of these soft tissue masses. However, a dermatofibrosarcoma protuberans differs from a trichilemmal carcinoma in terms of the most commonly involved site and the most frequently affected patient age group (8). In addition, malignant skin tumors such as squamous cell carcinomas show relatively ill-defined margins (9), whereas a clear cell squamous cell carcinoma lacks trichilemmal keratinization, lobular growth, and adnexal extension (10).

To our knowledge, this is the first case report of MR imaging findings of a trichilemmal carcinoma. Although a trichilemmal carcinoma is a rare disease, it may be included in the differential diagnosis of a large and superficially located soft tissue mass with heterogeneous enhancement on MR imaging in older patients.

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외모근초암의 자기공명영상 소견: 증례 보고[1]

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외모근초암은, 나이가 많은 사람에게서 햇빛에 노출되는 곳에 발생하는 드문 표재성 종양이다. 저자들은, 우측 후경부의 종괴를 주소로 내원한 79세 여자 환자를 검사하였다. 자기공명영상 소견에서 중심부는 낭성성분을 동반하였으며 이 주변부는 강한 조영증강을 보였다. 종괴는 조직병리학 검사에서 외모근초암으로 확진되었다. 저자들은 외모근초암의 자기공명영상 소견에 대해 보고하고자 한다.