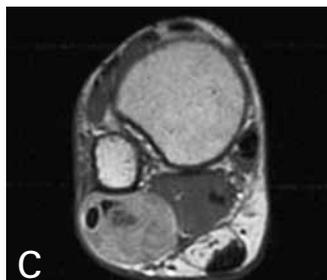


## CLEAR CELL SARCOMA OF THE ANKLE

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**Key-word:** Soft tissues, neoplasms

**Background:** A 48-year-old woman was referred to our hospital for evaluation of a soft tissue mass at her right ankle. At initial presentation, 5 years earlier, her general practitioner thought of a benign lesion and suggested to remove the mass. At the time, no additional imaging was requested. Over the years, the swelling was slowly growing but remained painless. Now, on clinical examination, a mass lesion was seen, located posterior of the lateral malleolus, consisting of a firm mass fixed to the surface underneath. The estimated size of the tumor was 2 x 6 cm, which gave rise to suspicion of a malignancy.



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### Work-up

Conventional radiograph of the right ankle (AP and profile views) (Fig. 1) shows normal appearance of bones and joints.

MRI of the right ankle and calf (Fig. 2) shows on coronal T1-weighted MR-image (A) a well-circumscribed soft tissue mass posterolateral of the fibula. The mass presents with homogeneous signal intensity, iso-intense to the surrounding flexor hallucis longus muscle. The lesion shows a direct relationship to the peroneal tendon. Coronal contrast-enhanced T1-weighted MR-image (B) demonstrates heterogeneous intense enhancement of the lesion is seen. On axial contrast-enhanced T1-weighted MR-image (C) the close relationship of the mass with the peroneal tendons is better demonstrated.

### Radiological diagnosis

The MRI findings are highly suggestive of a soft tissue sarcoma. Tru-Cut biopsy was performed. The histopathological diagnosis was *clear cell sarcoma*.

### Discussion

Masses around the foot and ankle are uncommon and the benign variants account for 75% of the cases, e.g. ganglion cysts, neurofibroma and lipoma. Clear cell sarcoma (CCS) of tendons and aponeuroses, formerly known as malignant melanoma of the soft tissue, is a rare entity with mass composed of cells of displaying melanocytic differentiation and accounts for 1% of all soft tissue sarcomas. CCS is mainly found in the lower extremities (95%) and especially around the foot and ankle. This tumor presents as a deep-seated mass lesion, with close relationship to tendons or aponeuroses. Most CCS lesions are slowly growing

and seldomly present with pain or tenderness. The etiology of CCS remains unknown. MRI is the examination of choice in the assessment of soft tissue tumors. On MRI, CCS presents with a slightly increased signal compared to muscle tissue on T1-weighted images and a low signal intensity on T2-weighted images. These short T1 and T2 relaxation times are said to be caused by the paramagnetic effect of the melanin present in the tumor cells. A certain amount of melanin is needed to give this particular higher signal on T1-weighted images. However, this intracellular melanin is only seldom found in the tumor. If the melanin is absent or present in too small amounts, the tumor can show a homogeneous appearance on T1 and T2-weighted images, thereby mimicking a benign tumor. Therefore, it is important to realize that, when a soft tissue mass is seen on a rare localization or reveals an unusual presentation, and the lesion shows a homogeneous appearance on MRI, CCS is a possible diagnosis.

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