Skeletal Radiol (2000) 29:187–195

ARTICLE

MR imaging of clear cell sarcoma (malignant melanoma of the soft parts): a multicenter correlative MRI-pathology study of 21 cases and literature review

L.H. De Beuckeleer
A.M. De Schepper
J.E. Vandevenne
J.L. Bloem
A.M. Davies
M. Oudkerk
E. Hauben
E. Van Marck
J. Somville
D. Vanel
L.S. Steinbach
J.M. Guinebretière
P.C.W. Hogendoorn
W.J. Mooi
K. Verstraete
C. Zaloudek
H. Jones

Received: 18 October 1999
Revision requested: 19 November 1999
Revision received: 21 January 2000
Accepted: 25 January 2000

L.H. De Beuckeleer, M.D. (✉)
A.M. De Schepper, M.D., Ph.D.
J.E. Vandevenne, M.D.
Department of Radiology,
University Hospital Antwerp
(University of Antwerp), Wilrijkstraat 10,
2650 Edegem, Belgium

J.L. Bloem, M.D., Ph.D.
Department of Radiology,
Leiden University Medical Center,
Leiden, The Netherlands

A.M. Davies, M.D.
MRI Center, Royal Orthopaedic Hospital,
Birmingham, UK

M. Oudkerk, M.D., Ph.D.
Department of Radiology,
Daniel Den Hoed Kliniek
(Academisch Ziekenhuis Rotterdam),
Rotterdam, The Netherlands

E. Hauben, M.D.
E. Van Marck, M.D., Ph.D.
Department of Pathology,
University Hospital Antwerp
(University of Antwerp),
Edegem, Belgium

J. Somville, M.D.
Department of Orthopaedic Surgery,
University Hospital Antwerp
(University of Antwerp), Edegem, Belgium

D. Vanel, M.D.
Department of Radiology,
Institut Gustave-Roussy, Villejuif, France

L.S. Steinbach, M.D.
Department of Radiology,
University of California San Francisco
(UCSF), San Francisco,
California, USA

J.M. Guinebretière, M.D.
Department of Pathology,
Institut Gustave-Roussy, Villejuif, France

P.C.W. Hogendoorn, M.D., Ph.D.
Department of Pathology,
Leiden University Medical Center,
Leiden, The Netherlands

W.J. Mooi, M.D., Ph.D.
Institute of Pathology,
Erasmus University Rotterdam,
Rotterdam, The Netherlands

K. Verstraete, M.D., Ph.D.
Department of Radiology,
University Hospital Ghent, Ghent,
Belgium

C. Zaloudek, M.D.
Department of Pathology,
University of California San Francisco
(UCSF), San Francisco, California, USA

H. Jones, M.D.
Department of Radiology,
Stanford University, Stanford, California,
USA

Abstract *Objective.* To evaluate MR imaging and pathology findings in order to define the characteristic features of clear cell sarcoma of the soft tissues (malignant melanoma of the soft parts).
Design and patients. MR examinations of 21 patients with histologically proven clear cell sarcoma of the musculoskeletal system were retrospectively reviewed and assessed for shape, homogeneity, delineation, signal intensities on T1- and T2-weighted images, contrast enhancement, relationship with adjacent fascia or tendon, secondary bone involvement, and intratumoral necrosis. In 19 cases the pathology findings were available for review and for a comparative MR-pathology study.
Results. On T1-weighted images, lesions were isointense (*n*=3), hypointense (*n*=7) or slightly hyperintense to muscle (*n*=11). Immunohistochemical examination was performed in 17 patients. All 17 specimens showed positivity for HMB-45 antibody. In nine of 11 lesions with slightly increased signal intensity on T1-weighted images, a correlative MR imaging-pathology study was possible. All nine were positive to HMB-45 antibody.
Conclusions. Clear cell sarcoma of the musculoskeletal system often has a benign-looking appearance on MR images. In up to 52% of patients, this lesion with melanocytic differentiation has slightly increased signal intensity on T1-weighted images compared with muscle. As the presence of this relative higher signal intensity on T1-weighted images is rather specific for tumors displaying melanocytic differentiation, radiologists should familiarize themselves with this rare entity and include it in their differential diagnosis when confronted with a well-defined, homogeneous, strongly enhancing mass with slightly higher signal intensity compared with muscle on native T1-weighted images.

Key words Clear cell sarcoma · Malignant melanoma of soft parts · MRI · Magnetic resonance · Neoplasm

Introduction

Clear cell sarcoma of the soft tissues is a rare sarcoma displaying melanocytic differentiation, accounting for only 0.8–1% of all malignancies of the musculoskeletal system [1–3]. To our knowledge, no large MR imaging series on clear cell sarcomas have been reported.

Since the lesion is mostly well defined and lacks perilesional oedema, bone invasion, satellite nodules, or intratumoral necrosis, it may erroneously be interpreted on MR images as a nonaggressive mass [4–6]. If such a "benign-looking" mass is initially maltreated by marginal excision, tumor dissemination is much more likely and definitely compromises patient outcome and further therapeutic strategy. The purpose of this study was to describe the MR appearance of clear cell sarcoma in a relative large series of patients and to perform an MR-histopathological correlation to determine whether analysis of MR features can provide a basis for predicting gross histological elements. In addition, since the description of MR imaging features of clear cell sarcoma has been limited to some isolated accounts in the surgical, orthopedic, and radiology reports, the literature describing the MR imaging features of clear cell sarcoma was reviewed.

Materials and methods

The records of 26 cases of clear cell sarcoma were retrieved from the medical files and databases of nine institutions. Five patients with histopathologically proven clear cell sarcoma underwent no preoperative MR imaging and were therefore not included in the study population. Our study group consisted of 21 patients. There were 11 male and 10 female patients. Age ranged from 14 to 75 years (mean age 40 years). Eighteen were primary tumors, three were recurrent masses. The radiological and histopathological data of all patients were retrospectively reviewed.

The MR examinations were performed on commercially available MR units of different field strengths, ranging from 0.2 to 1.5 T. Eleven patients were examined on a 1.5-T MR unit, three on a 0.5-T MR unit, six on a 1.0-T and one patient on a 0.2-T MR unit. Twenty patients underwent spin-echo T1-weighted imaging [repetition time (ms)/echo time (ms)=420–800/9–30]. Fast spin-echo T1-weighted imaging (1380/12) was performed in one patient. T2-weighted images were available in 18 cases. Spin-echo T2-weighted imaging (1800–2000/50–120) was performed in seven cases, fast spin-echo T2-weighted imaging (2400–5200/90–108) in seven cases, gradient-echo T2-weighted imaging (600/20/20° flip angle) in one case, fat-suppressed T2-weighted images (5019/17) in one case, and fat-saturated T2-weighted images (1700–4711/100) in two cases. In 13 patients, Gd-enhanced spin-echo T1-weighted images were obtained, with the same imaging parameters as were used at unenhanced T1-weighted imaging. In one patient, Gd-enhanced fat-suppressed images were performed (600/20). In another patient, a dynamic contrast-enhanced study was performed using a T1-weighted gradient-echo sequence (TR/TE/TI/flip angle=9/4/200/8°).

Two reviewers (LHDB, AMDS) analyzed the images together – a process that resulted in a consensus interpretation. The following parameters were assessed: shape, delineation, homogeneity, diameters, signal intensities on both T1- and T2-weighted images, secondary bone involvement, enhancement, and intratumoral necrosis. All patients were treated surgically and clear cell sarcoma was confirmed on pathological examination. In 19 of 21 lesions, histopathological data were available for correlation with the MR findings.

For histopathological review, hematoxylin-eosin-stained specimens were available in 11 cases. Written pathology reports were analyzed in eight other cases. To search for intralesional hemosiderin, iron-stained specimens were available in 13 cases.

Light microscopic examination was performed and the specimens were evaluated for cellularity, mitotic activity (number of mitoses per 10 high-power fields), and nucleo-cytoplasmic index (<1/1, =1, or >1/1). The presence of dense connective tissue, loose connective tissue and myxoid connective tissue, intratumoral necrosis, hemorrhage, hemosiderin, or melanin was noted. Immunohistochemical examination included stains with HMB-45 antibody. A semiquantitative estimation of the HMB-45 positivity of the cells was performed.

Results

The results are summarized in Table 1. Sixteen lesions were located in the lower limb [groin (*n*=1), buttocks (*n*=1), thigh (*n*=3), around the knee (*n*=2), lower leg (*n*=3), foot (*n*=6)], and five in the upper limb [axilla (*n*=1), shoulder (*n*=1), elbow (*n*=1), and hand (*n*=2)]. Nine tumors were oval or fusiform, six were round, four were multilobulated, and two were identified as "spider-like." Fourteen lesions were sharply demarcated. Eighteen of 21 lesions were homogeneous. Mean diameter of the lesions varied from 1.7 to 10 cm. On T1-weighted images, lesions were isointense (*n*=3), hypointense (*n*=7), or slightly hyperintense compared with muscle (*n*=11). On T2-weighted images, low (*n*=3), intermediate (*n*=6) to high (*n*=9) signal intensity was seen. The high signal intensity in three cases of the last group may be attributed to the use of gradient-echo or fat-suppressed sequences. On Gd-enhanced T1-weighted images, intermediate (*n*=4) to strong (*n*=10) enhancement was seen. In one patient, early and fast enhancement was shown on dynamic Gd-enhanced MR imaging. In 14 cases, a relationship with an adjacent tendon or fascia was obvious (Figs. 1, 2). Intratumoral necrosis was observed in one case. Secondary bone destruction was demonstrated in two patients.

On microscopic examination, all lesions were highly cellular (51–75% or 76–100% of the specimen; listed in Table 1 as +++ and ++++, respectively). Mitotic activity ranged from 0 to 15 per 10 high-power fields. Nucleo-cytoplasmic index was <1 in nine, equal to 1 in two, and >1 in three of 14 patients. All except one of the patients with high signal intensity on T2-weighted images had a low nucleo-cytoplasmic index. Iron stains showed hemosiderin deposits in five of 13 patients. Intralesional hemorrhage was seen in four patients. Intratumoral necrosis was seen in nine patients. The presence of intralesional melanin was noted in one specimen. Immunohistochemical examination revealed positive staining against HMB-45 antibody in 17 of 17 patients (Table 2).

Table 1 MR imaging, pathological, and immunohistochemical data (*T1* signal intensity on T1-weighted images, *T2* signal intensity on T2-weighted images, *Gd-T1* signal intensity on Gd-enhanced T1-weighted images, *N/C* nucleo-cytoplasmic index, *Ho* homogeneous, *In* inhomogeneous, *H* high signal intensity, *I* intermediate signal intensity, *L* low signal intensity, *NA* not available)

Case no.	Sex	Age (years)	Field strength (T)	Localisation	Homogeneity	T1	T2	Gd-T1	Cellularity	Mitoses	N/C	Connective tissue			Necrosis	Hemorrhage	Hemosiderin	Melanin	HMB-45
												Dense	Loose	Myxoid					
1	F	14	1.5	Buttock	Ho	L	H	++	++++	2	<1	+	–	–	+	+	–	NA	+
2	M	34	1.0	Achilles tenodn	In	H	I	NA	++++	3	>1	++++	–	–	+	–	–	–	++++
3	M	48	0.5	Thigh	In	L	I	+	+++	12	<1	++	–	–	++	–	–	–	+
4	M	42	1.5	Lower leg	Ho	L	H	++	++++	15	<1	+	–	–	+	–	–	–	+++
5	F	25	0.2	Hand	Ho	H	I	++	++++	0	>1	0	–	–	–	–	–	–	++++
6	F	37	1.5	Foot	Ho	H	H	++	+++	9	<1	+	–	–	–	–	–	–	++++
7	M	47	1.5	Knee	Ho	H	NA	++	+++	2	1	+	–	–	–	–	+	–	NA
8	M	75	1.5	Thigh	Ho	L	NA	++	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA
9	M	52	1.5	Axilla	In	H	I	++	++++	4	<1	+	+	–	++	–	+	–	+
10	F	63	0.5	Knee	Ho	I	H	+	+++	7	<1	+	++	++	++	+	+	–	+
11	M	41	0.5	Hand	Ho	H	H	NA	+++	3	<1	++	++	–	–	–	+	–	+++
12	F	34	1.5	Lower leg	Ho	L	I	++	++++	1	1	+	–	–	++	+	–	NA	+
13	F	10	1.5	Shoulder	Ho	H	H	NA	++++	12	>1	NA	NA	NA	–	NA	NA	–	NA
14	F	28	1.5	Elbow	Ho	H	H	NA	++++	11	<1	+	–	–	+	–	–	+	+
15	M	44	1.5	Foot	Ho	L	L	+	+++	7	<1	++	–	–	–	+	+	–	++++
16	F	38	1.5	Groin	Ho	L	NA	++	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA
17	M	34	1.0	Foot	Ho	H	L	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	+
18	F	33	1.0	Foot	Ho	I	H	NA	NA	9	NA	NA	NA	NA	NA	NA	NA	NA	+
19	M	22	1.0	Foot	Ho	I	I	+	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	+
20	F	66	1.0	Foot	Ho	H	L	NA	NA	3	NA	NA	NA	NA	NA	NA	NA	NA	+
21	M	44	1.0	Thigh	Ho	H	H	++	NA	NA	NA	+	–	–	+	NA	NA	NA	+

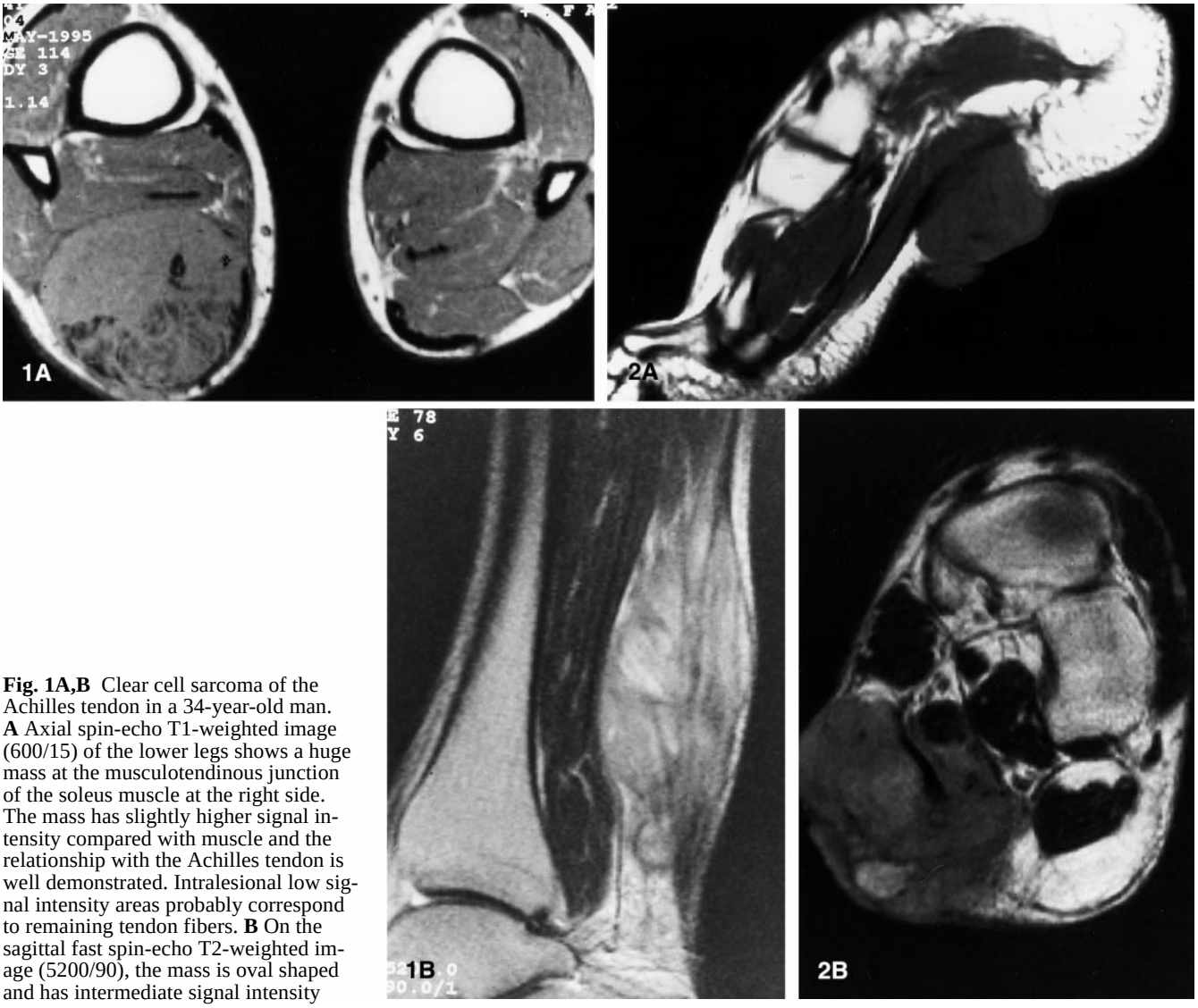


Fig. 1A,B Clear cell sarcoma of the Achilles tendon in a 34-year-old man. **A** Axial spin-echo T1-weighted image (600/15) of the lower legs shows a huge mass at the musculotendinous junction of the soleus muscle at the right side. The mass has slightly higher signal intensity compared with muscle and the relationship with the Achilles tendon is well demonstrated. Intralesional low signal intensity areas probably correspond to remaining tendon fibers. **B** On the sagittal fast spin-echo T2-weighted image (5200/90), the mass is oval shaped and has intermediate signal intensity

Table 2 HMB-45 positivity versus signal intensity on T1-weighted images (*NA* HMB-45 staining not available, *SI* signal intensity, *TI-WI* T1-weighted images)

	HMB-45 positive	NA	Total
Slightly increased SI on T1-WI	9	2	11
Intermediate SI (as muscle) on T1-WI	3	0	3
Low SI on T1-WI	5	2	7
Total	17	4	21

Fig. 2A,B Tumoral lesion of the foot sole in a 34-year-old man who had been operated on for a clear cell sarcoma of the same area at the age of 14 years. **A** Sagittal spin-echo T1-weighted image (480/15) shows a large mass extending within the subcutis/cutis and enveloping the plantar aponeurosis. Signal intensity is slightly higher than that of adjacent muscles. **B** The lesion has low signal intensity on the fast spin-echo T2-weighted image (2720/105). Location, signal intensities, and relationship with the plantar aponeurosis are in favor of a clear cell sarcoma

Discussion

Clear cell sarcoma is an extremely rare, slow-growing malignant tumor showing melanocytic differentiation [3]. It affects predominantly young to middle-aged adults and mostly arises in the deep soft tissues of the limbs, adjacent to tendons, aponeuroses, and fascial structures. Lesions located in the lower limb (foot and ankle, knee, thigh) (Figs. 1–3) outnumber those in the upper limb (Figs. 4–6). Involvement of the head and neck region and the trunk seldom occurs [1–3]. On physical examination, the lesion presents as a firm, ill-defined mass. The overlying skin is not involved, except in bulky lesions ulcerating to the epidermis. Size of the lesion may range from 1 to 14 cm. Pathologically, the tumor consists

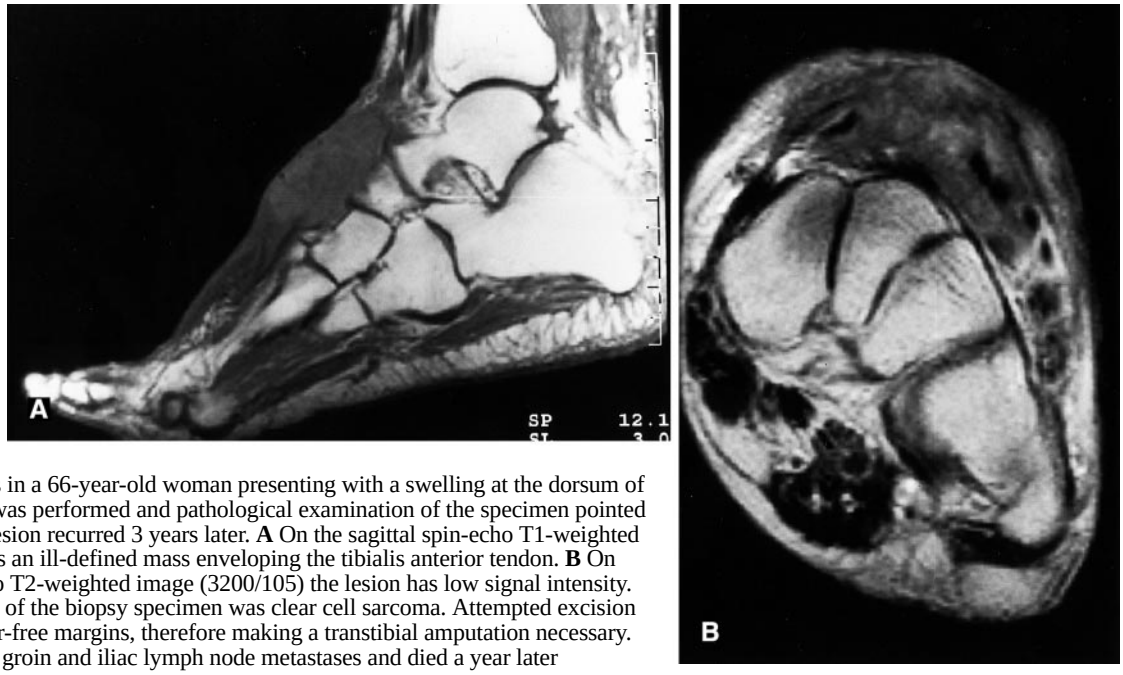


Fig. 3A,B Huge mass in a 66-year-old woman presenting with a swelling at the dorsum of the foot. An excision was performed and pathological examination of the specimen pointed to a synovioma. The lesion recurred 3 years later. **A** On the sagittal spin-echo T1-weighted image (600/15) there is an ill-defined mass enveloping the tibialis anterior tendon. **B** On the axial fast spin-echo T2-weighted image (3200/105) the lesion has low signal intensity. Pathological diagnosis of the biopsy specimen was clear cell sarcoma. Attempted excision failed to achieve tumor-free margins, therefore making a transtibial amputation necessary. The patient developed groin and iliac lymph node metastases and died a year later

of a number of compact nests of uniform, rounded or fusiform cells separated by fibrocollagenous tissue which is frequently contiguous with adjacent tendons, fascial structures, or aponeuroses. The ovoid, vesicular nuclei usually have large, basophilic nucleoli and are surrounded by a clear cytoplasm. Multinucleated giant cells, with 10 to 15 peripherally placed nuclei, can be present throughout the mass. Mitotic figures usually are sparse [3, 7].

Radiologically, the lesion presents as a nonspecific, noncalcified soft tissue mass. Involvement of adjacent bones is an infrequent, but important finding [8] (Fig. 5). To our knowledge, MR imaging findings in clear cell sarcoma of the musculoskeletal system have been reported at least three times in the past. In a series of 14 tumors of the foot, the signal intensities of three clear cell sarcomas were incompletely described. One lesion had low signal intensity on T1-weighted images. On T2-weighted images, low to intermediate signal intensity was seen in all three patients [9]. A small nodular mass at the plantar aponeurosis having intermediate signal intensity on T2-weighted images was described by Prieskorn et al. [5], but signal intensity on T1-weighted images was not reported. A clear cell sarcoma of the elbow, as reported by Schnarkowski et al. [6], had a signal intensity between those of muscle and fat on T1-weighted images, and high signal intensity on STIR images.

In our series, 86% of the masses were homogeneous on T1- and T2-weighted images, 67% of the lesions were well defined, and bone destruction and intratumoral necrosis was seen in only 10% and 5% respectively. As a consequence, clear cell sarcomas are often misdiagnosed radiologically and graded as nonaggressive lesions [4–6] (Figs. 3, 4). Despite the morphology and behavior of the lesion on MR images being in favor of a benign lesion, the prognosis is poor, and this may be caused partly by an inadequate initial surgical approach.

Histologically, the infrequent occurrence of clear cell sarcoma in the general population often leads to an erroneous diagnosis of metastatic melanoma, especially when the tumor is localized in the vicinity of a lymphatic plexus (e.g., axilla, cubital fossa). The immunohistological features may be reflected in the signal intensities of the lesion: on the cut surface, dark pigment is seen in 20% of cases of clear cell sarcoma [10]. On histological examination, using Fontana or Warthin-Starry stains for melanin, up to 78% of clear cell sarcomas contain intracellular melanin [7,10]. Therefore, and because malignant melanoma and clear cell sarcoma share an overlapping immunohistochemical profile, i.e., scattered positivity for keratin, HMB-45, and S-100 protein [11], the term malignant melanoma of the soft parts has been advocated over the purely descriptive term of clear cell sarcoma. However, we believe that this term does not deserve any priority since cytogenetic analysis has demonstrated a reciprocal translocation in clear cell sarcoma between the long arms of chromosomes 12 and 22 [(t(12:22) (q13;q12.2)] – not seen in cutaneous melanoma – supporting the view that they are two complete distinct histopathological entities [3]. Diagnostic difficulties in differentiating clear cell sarcoma and a metastasis of a cutaneous melanoma in the absence of a known primary cutaneous tumor are now resolved by molecular and (cyto)genetic data [12–14].

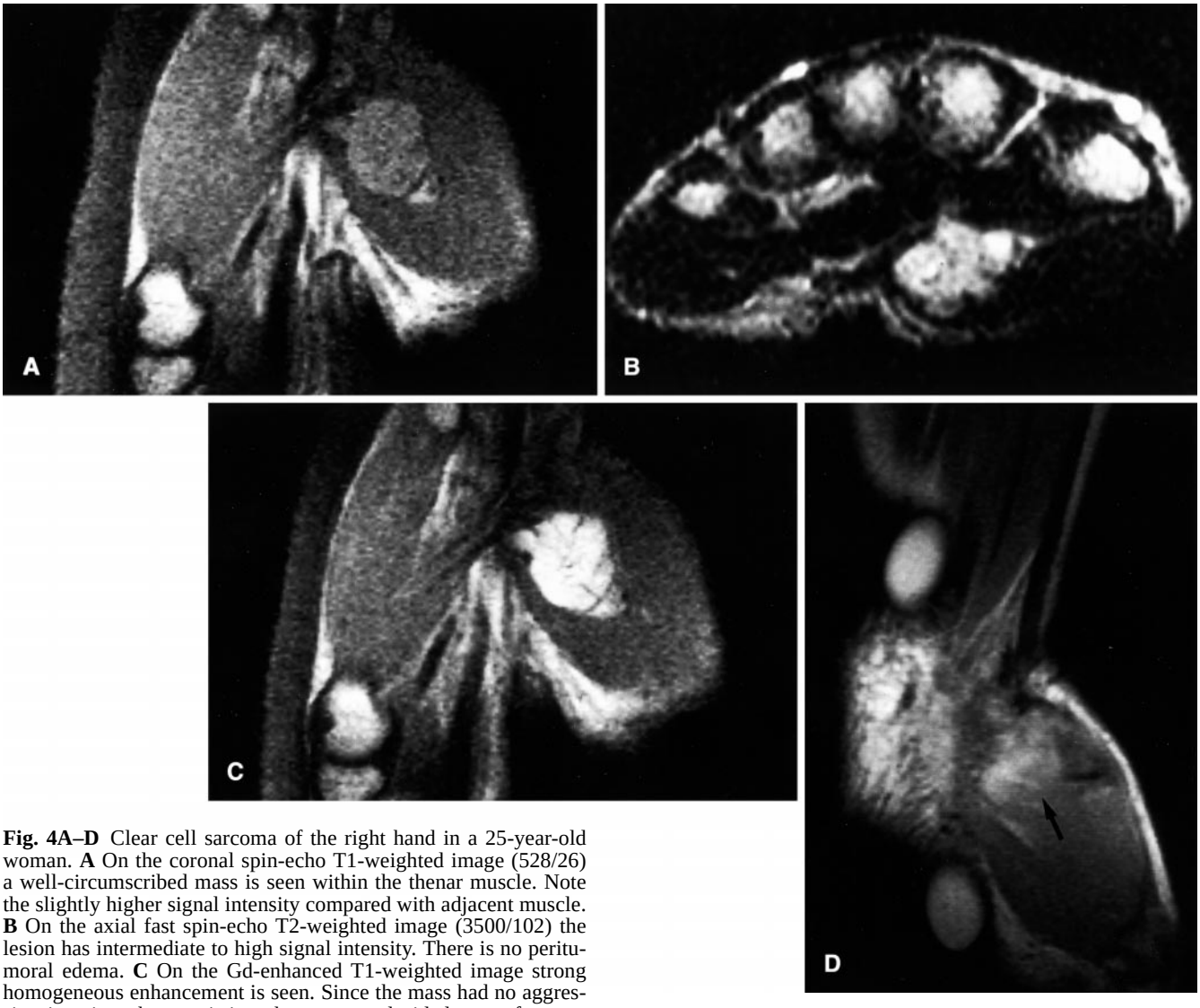


Fig. 4A–D Clear cell sarcoma of the right hand in a 25-year-old woman. **A** On the coronal spin-echo T1-weighted image (528/26) a well-circumscribed mass is seen within the thenar muscle. Note the slightly higher signal intensity compared with adjacent muscle. **B** On the axial fast spin-echo T2-weighted image (3500/102) the lesion has intermediate to high signal intensity. There is no peritumoral edema. **C** On the Gd-enhanced T1-weighted image strong homogeneous enhancement is seen. Since the mass had no aggressive imaging characteristics, the surgeon decided to perform an excisional biopsy. Pathological diagnosis of clear cell sarcoma was made. Section margins were borderline. **D** An MR examination was performed 6 weeks after the initial surgery, showing an indeterminate enhancing area (*arrow*) on the Gd-enhanced spin-echo T1-weighted image. A wide excision of thenar muscle was performed. Pathological examination revealed small nests of malignant cells. Since then a transradial-ulnar amputation has been performed. At 19 months after initial therapy no metastatic disease has been demonstrated

Because of the paramagnetic effects of intralesional melanin, causing T1 and T2 shortening, high signal intensity on T1- and lower signal intensity on T2-weighted images should theoretically be present. The supposedly paramagnetic property of stable free radicals within the melanin pigment results in T1 and T2 shortening. Others adhere to the theory that chelated metal ions in melanin may also contribute to T1 shortening, resulting in hyperintensity on T1- and relative hypointensity on T2-weighted images. The etiology of this phenomenon is still debated. Enochs et al. [15] recently reported that melanin, having a high affinity for metal ions, may contain a wide variety of bound metals having paramagnetic properties. This theory increasingly gains ground.

The slightly increased signal intensity on T1-weighted images (compared with muscle) is rarely seen in soft tissue tumors and therefore is a quite characteristic sign that may narrow down the list of differential diagnoses. In our series, the signal intensity of clear cell sarcomas was isointense (n=3) or slightly hyperintense (n=11) compared with skeletal muscle on T1-weighted images (Figs. 1–6). The latter signal intensity pattern may be attributed to the histological composition (melanocytic differentiation) of the tumor.

On T2-weighted images, nine of 18 lesions showed high signal intensity, six had intermediate signal intensi-

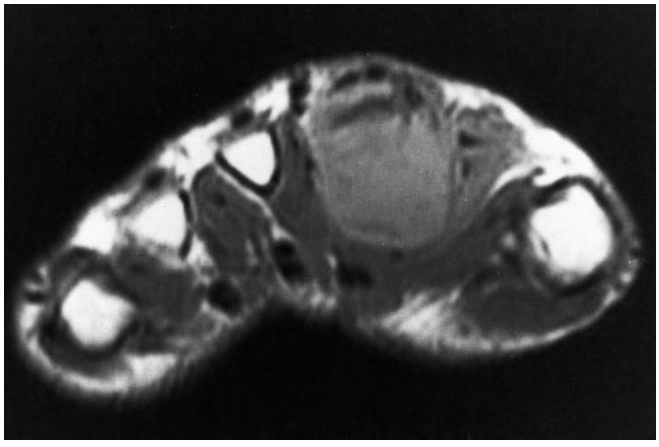


Fig. 5 Clear cell sarcoma of the left hand in a 41-year-old man. On the axial spin-echo T1-weighted image (550/30) a rounded mass with high signal intensity is seen at the second digit. Destruction of the metacarpal bone proves the aggressive behaviour of the lesion

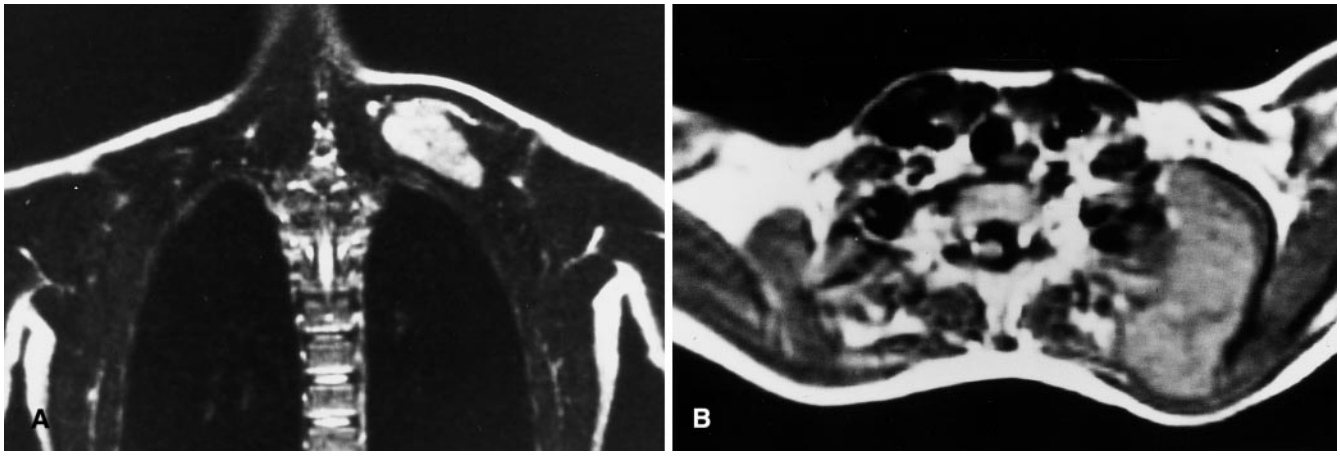
ty (between muscle and fat), and three had low signal intensity. The signal intensity on T2-weighted images correlates with intra- and extracellular water content. Therefore, besides the paramagnetic effects of melanin on T2 shortening, high cellularity and the presence of a nucleo-cytoplasmic index greater than 1 may contribute to the intermediate to low signal intensity on T2-weighted images. In two of six masses with intermediate signal intensity on T2-weighted images, cellularity was high and the nucleo-cytoplasmic index greater than 1. In all but one of the lesions with high signal intensity on T2-weighted images for which pathological findings were available, a low nucleo-cytoplasmic index was found. However, comparative MR-pathology studies on a larger series are mandatory before definite conclusions may be drawn. The high amount of myxoid stroma in one particular case may be responsible for the high signal intensity on T2-weighted images. The presence of abundant loose connective tissue between the cellular nests may be responsible for high signal intensity on T2-weighted images, as seen in two of these cases.

In this series, no correlation was found between signal intensities on T1- or T2-weighted images and other histopathological criteria such as the number of mitoses, focal areas of intratumoral necrosis, hemosiderin, and intralesional hemorrhage. The presence of intralesional hemorrhage, as seen in four cases, was probably due to the manipulation of the mass during resection.

At immunohistochemical examination, all patients were positive to HMB-45 antibody. HMB-45 was initially considered a monoclonal antibody that binds to an antigen common to the cytoplasm of melanoma cells and of premature developing melanocytes [16]. Other researchers tested this antibody on various tissues and neoplasms, and concluded that it very rarely stains nonmelanocytic tumors [17]. Kikuchi et al. [18] demonstrated that the HMB-45 binding site was mainly the stage I, II and III melanosomes during melanogenesis in neoplastic and non-neoplastic melanocytes. Scarce positive staining is seen in stage IV melanosomes in melanoma cells in vivo, regardless of their melanin production [18, 19].

Comparative MR-immunohistochemistry studies revealed that lesions with slightly higher signal intensity than muscle on T1-weighted images were positive to HMB-45 antibody in nine of nine (100%) cases (Table 2). Upon pathological examination, intralesional melanin was detected in only one case. Reasons for the failure to demonstrate intratumoral melanin is first the difficulty of having appropriate stains available for review, a methodological problem encountered in many retrospective studies.

Fig. 6A,B Clear cell sarcoma of the suprascapular region in a 10-year-old girl presenting with dull, aching pain in the left suprascapular area. On clinical examination a well-circumscribed mass was noted. **A** On the coronal spin-echo T2-weighted image (2000/80) a homogeneous mass with high signal intensity is seen. **B** On the axial spin-echo T1-weighted image (800/20) the lesion has higher signal intensity than surrounding muscles



Another hypothetical reason may be the relative paucity of melanin in some clear cell sarcomas. This may also explain the absence of higher signal intensity in three lesions having intermediate signal intensity on T1-weighted images that also showed positive HMB-45 staining. Ferris et al. [20] have quantified the melanin content in uveal melanomas using an image analyzer and reported that low melanin content (0.29–5%) did not result in an increased signal intensity on T1-weighted images. Even lesions containing up to 23% melanin did not have significant higher signal intensity on T1-weighted images [20]. It seems that a certain amount of melanin is needed to shorten the T1 relaxation time significantly in order to result in high signal intensity on T1-weighted images. Another important factor that may explain the unexpected low signal intensity on T1-weighted images for HMB-45 positive lesions may be the low number of stage III or IV melanosomes, the so-called melanin-laden melanosomes. Unfortunately, no fresh tumor material for electron microscopic study was available in the five cases with strong HMB-45 positivity that had low signal intensity on T1-weighted images. Another hypothesis to explain the "atypical" appearance of at least some lesions may be the high field strength (1.5 T) used in 11 cases, six of which lacked slightly increased signal intensity on T1-weighted images. It is well known that the T1 time is dependent upon the external field strength, i.e., it is proportional to (operating frequency ω_0 at the applied field strength)^n, where n is an empirical parameter between 1/3 and 1/2, depending on the tissue under consideration [21]. Given this interdependence, the T1 relaxation times of biological tissues are markedly prolonged at high field strength, resulting in relative lower signal intensity compared with examinations performed on a low-field system.

Furthermore, it is known that MR signal intensities result from a combination of chemical, physiological and biological parameters of tumor cells, subcellular structures, and tumoral components, thereby reflecting gross morphology rather than underlying microscopic histology. The importance of each parameter has yet to be determined.

The radiological differential diagnosis should include other lesions that are characterized by their high signal intensity on T1-weighted images. Lipomatous lesions, such as lipoma, well-differentiated liposarcoma, and lipoblastoma generally have much higher signal intensities on T1-weighted images, and are easily differentiated using fat-saturation techniques or STIR images. Alveolar soft part sarcoma has high signal intensity on T1- and T2-weighted images. Visualization of intralesional flow voids is mandatory to establish this diagnosis [22]. High signal intensity on T1-weighted images is also seen in melanocytic schwannoma, a rare lesion originating from the neural crest and occupying an intermediate position between melanoma and schwannoma. Most cases are reported to originate from spinal nerve roots or the central nervous system [23, 24]. Subacute hematomas have high signal intensity on T1-weighted images, since intralesional methemoglobin also has paramagnetic properties. However, the enhancement pattern differs.

Therapy consists of radical excision or amputation, combined with radiation therapy and chemotherapy depending on the achieved surgical margins and pathological grade. After initial therapy, the clinical course is often characterized by local recurrences and, unlike other soft tissue sarcomas, metastases to regional lymph nodes. Distant metastases are mostly found in the skeleton and the lungs. The overall prognosis is poor. Nevertheless, some patients who are alive 26 to 30 years after the time of initial diagnosis have been reported. Tumor size and intratumoral necrosis are described to be independent predictors of survival. Tumor size larger than 5 cm or the presence of intralesional necrosis is associated with a less protracted disease course and a poor prognosis [25, 26].

In summary, clear cell sarcoma often has a benign-looking appearance on MR imaging studies. Nevertheless, it behaves as a relentless, highly malignant soft tissue sarcoma with a tendency for local recurrence and metastatic spread. Often, this lesion with melanocytic differentiation has slightly higher signal intensity compared with muscle on T1-weighted images. As the presence of this higher signal intensity on T1-weighted images is rather specific for tumors showing melanocytic differentiation, radiologists should familiarize themselves with this rare entity and include it in their differential diagnosis.

Acknowledgement The authors wish to thank Dr. M.J. Kransdorf (Department of Radiology, Commonwealth Radiology, Richmond Virginia, USA) for contributing case material and for his substantial efforts in the preparation of this manuscript.

References

1. Degryse HR. Lesions of uncertain origin. In: De Schepper AM, Parizel PM, Ramon F, De Beuckeleer L, Vandevenne JE, eds. Imaging of soft tissue tumors. Berlin Heidelberg New York: Springer, 1997: 325–344.
2. Kransdorf MJ, Murphey MD. Neurogenic tumors. In: Kransdorf MJ, Murphey M, eds. Imaging of soft tissue tumors. Philadelphia: WB Saunders,1997: 255–273.
3. Graadt van Roggen JF, Mooi WJ, Hogendoorn PCW. Clear cell sarcoma of tendons and aponeuroses (malignant melanoma of soft parts) and cutaneous melanoma: exploring the histogenetic relationship between these two clinicopathological entities. J Pathol 1998; 186: 3–7.

4. Langen RJ, Nyongo AJ, Huntrakoon M, Landry ME. Clear cell sarcoma of tendons and aponeuroses: histogenesis and mode of treatment. J Foot Surg 1989; 28: 112–115.
5. Prieskorn DW, Irwin RB, Hankin R. Clear cell sarcoma presenting as an interdigital neuroma. Orthop Rev 1992; 21: 963–970.
6. Schnarkowski P, Peterfy CG, Johnston JO, Weidner N. Clear cell sarcoma mimicking peripheral nerve sheath tumor. Skeletal Radiol 1996; 25: 197–200.
7. Enzinger FM, Weiss SW. Malignant tumors of the peripheral nerves. In: Enzinger FM, Weiss SW, eds. Soft tissue tumors. St Louis: Mosby, 1995: 889–928.
8. Morishita S, Onomura T, Yamamoto S, Nakashima Y. Clear cell sarcoma of tendons and aponeuroses (malignant melanoma of soft parts) with unusual roentgenologic findings. Clin Orthop 1987; 216: 276–279.
9. Wetzel LH, Levine E. Soft tissue tumors of the foot: value of MR imaging for specific diagnosis. AJR 1990; 155: 1025–1030.
10. Chung EB, Enzinger FM. Malignant melanoma of soft parts: a reassessment of clear cell sarcoma. Am J Surg Pathol 1983; 7: 405–413.
11. Mooi WJ, Deenik W, Peterse JL, Hogendoorn PCW. Keratin immunoreactivity in melanoma of soft parts (clear cell sarcoma). Histopathology 1995; 27: 61–65.
12. Speleman F, Delattre O, Peter M, Hauben E, Van Roy N, Van Marck E. Malignant melanoma of the soft parts (clear cell sarcoma): confirmation of EWS and ATF-1 gene fusion caused by a t(12;22) translocation. Mod Pathol 1997; 10: 496–499.
13. Speleman F, Colpaert C, Goovaerts G, Leroy JF, Van Marck E. Malignant melanoma of soft parts. Further cytogenetic characterization. Cancer Genet Cytogenet 1992; 60: 176–179.
14. Graadt van Roggen JF, Bovée JVMG, Morreau J, Hogendoorn PCW. Diagnostic and prognostic implications of the unfolding molecular biology of bone and soft tissue tumours. J Clin Pathol 1999; 52: 1–8.
15. Enochs WS, Petherick P, Bogdanova A, Mohr U, Weissleder R. Paramagnetic metal scavenging by melanin: MR imaging. Radiology 1997; 204: 417–423.
16. Gown AM, Vogel AM, Hoak D, Gough F, McNutt MA. Monoclonal antibodies specific for melanocytic tumours distinguish subpopulations of melanocytes. Am J Pathol 1986; 123: 195–203.
17. Yates AJ, Banerjee SS, Bishop PW, Graham KE. HMB-45 in non-melanocytic tumours. Histopathology 1993; 23: 477–478.
18. Kikuchi A, Shimizu H, Nishikawa T. Expression and ultrastructural localization of HMB-45 antigen. Br J Dermatol 1996; 135: 400–405.
19. Bleehen SS. Disorders of skin colour. In: Champion RH, Burton JL, Burns DA, Breathnach SM, eds. Rook/Wilkinson/Ebling textbook of dermatology. Oxford: Blackwell Science, 1998: 1753–1815.
20. Ferris JD, Bloom PA, Goddard PR, Collins C. Quantification of melanin and iron content in uveal malignant melanomas and correlation with magnetic resonance image. Br J Ophthalmol 1993; 77: 297–301.
21. Parizel PMRJ. Determinants of MR image quality as a function of field strength: signal-to-noise ratio, spatial resolution and contrast-to-noise ratio. In: Parizel PMRJ. The influence of field strength on magnetic resonance imaging: a comparative study in physicochemical phantoms, isolated brain specimens and clinical applications. Antwerp: University of Antwerp, Belgium, 1994: 23–43.
22. Iwamoto Y, Morimoto N, Chuman H, Shinohara N, Sugioka Y. The role of MR imaging in the diagnosis of alveolar soft part sarcoma: a report of 10 cases. Skeletal Radiol 1995; 24: 267–270.
23. Liessi G, Barbazza R, Sartori F, Sabbadin P, Scapinello A. CT and MR imaging of melanocytic schwannomas; report of three cases. Eur J Radiol 1990; 11: 138–142.
24. Bendszus M, Urbach H, Wolf HK, Schramm J, Solymosi L. Magnetic resonance imaging of intraspinal melanotic schwannoma. Eur Radiol 1998; 8: 1197.
25. Sartoris DJ, Haghighi P, Resnick D. Case report 423. Clear cell sarcoma plantar aspect of right foot. Skeletal Radiol 1987; 16: 325–332.
26. Lucas DR, Nascimento AG, Sim FH. Clear cell sarcoma of soft tissues: Mayo Clinic experience with 35 cases. Am J Surg Pathol 1992; 16: 1197–1204.