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EDITORIAL

Medical imaging for the diagnosis, recurrence and metastasis evaluation of clear cell sarcoma

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Abstract

Clear cell sarcoma (CCS) of soft tissue is extremely rare, accounting for approximately 1% of all soft tissue tumours. It is very difficult to diagnose CCS based on clinical manifestations. Magnetic resonance imaging (MRI) provides high-resolution images of soft tissues and pathological features such as mucus, necrosis, bleeding, and fat through high and low signals on T1 weighted image (T1WI) and T2 weighted image (T2WI). On the other hand, the paramagnetism of melanin in CCS shortens the relaxation time of T1 and T2, and high signal intensity on T1WI and low signal intensity on T2WI can be found. This is different from most other soft tissue sarcomas. At present, the treatment method for CCS is surgical resection. MRI can effectively display the tumour edge, extent of surrounding oedema, and extent of fat involvement, which is highly important for guiding surgical resection and predicting postoperative recurrence. As an invasive sarcoma, CCS has a high risk of metastasis. Regardless of the pathological condition of the resected tumour, MRI or computed tomography (CT) should be performed every 1-2 years to assess recurrence at the primary site and to screen for metastasis in the lungs, liver, and bones. If necessary, PET-CT can be performed to evaluate the overall condition of the patient.

Key Words: Clear cell sarcoma; Recurrence; Metastasis; Computed tomography; Magnetic resonance imaging; Editorial

Core Tip: In this editorial, we comment on a case report by Liu *et al* published in the recent issue. According to the authors of this article, the objective of the study was to investigate the metastasis, surgical treatment and postoperative follow-up of clear cell sarcoma (CCS). In this editorial article, we will focus specifically on the roles of computed tomography and magnetic resonance imaging in the diagnosis, surgical treatment, evaluation of recurrence, and detection of metastasis of CCS.

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INTRODUCTION

In this editorial, we comment on a case report by Liu *et al*[1] published in the recent issue. According to the authors of this article, the objective of the study was to investigate the metastasis, surgical treatment and postoperative follow-up of clear cell sarcoma (CCS). In this editorial article, we will focus specifically on the roles of computed tomography (CT) and magnetic resonance imaging (MRI) in the diagnosis, surgical treatment, evaluation of recurrence, and detection of metastasis of CCS.

CCS

CCS of soft tissue is extremely rare, accounting for approximately 1% of all soft tissue tumours, with an annual incidence of approximately one million[2]. Due to its extremely low incidence, CCS was listed as an ultrarare sarcoma at the consensus meeting held by the Connective Tissue Oncology Society in 2019[3]. The list of ultrarare sarcomas included 56 types of soft tissue sarcomas, all of which are challenging to diagnose and treat.

CCS typically occurs in young people aged 30 to 40 years. Primary CCS typically occurs in the limbs, especially in the feet or ankle joints, followed by the knee joints. The retroperitoneal space, kidneys, gastrointestinal tract, chest cavity, lungs, mediastinum, bones, and head and neck are rarely affected. Tumours that develop in the limbs are often located deep in the soft tissue or under the fascia and are clinically characterized by slow growth without pain[2,4-6]. These tumours can easily be misdiagnosed as benign tumours. Tumours that occur in important areas can cause compression, pain, and functional abnormalities as the tumour grows. If it occurs in the gastrointestinal tract, obstructive symptoms may occur. Some patients have lymph node metastasis and distant metastasis at the initial diagnosis. It is very difficult to diagnose CCS and metastasis based on clinical manifestations.

CT and MRI are important imaging methods. The manifestations of CCS on CT include soft tissue lesions with homogeneous density, and in cases of necrosis, heterogeneous density may be present. Due to insufficient information provided, the diagnostic value of these methods is limited. MRI provides high-resolution images of soft tissue and pathological features such as mucus, necrosis, bleeding, and fat through high and low signals on T1 weighted image (T1WI) and T2 weighted image (T2WI)[7]. According to the latest ESMO guidelines, MRI is the best imaging method for soft tissue sarcomas, except for those in the retroperitoneal and visceral areas[8]. CCS originates from neural crest cells in soft tissues[9,10]. The deposition of melanin within the lesion makes it difficult to distinguish from malignant melanoma through immunohistochemistry[11]. At the same time, the two have similar morphological and clinical features[12], which makes differential diagnosis difficult. The specific signals on MRI that indicate CCS show important clinical value. In cases of CCS, the paramagnetism of melanin shortens the relaxation time of T1 and T2, and there is high signal intensity on T1WI and low signal intensity on T2WI[7]. This is different from most other soft tissue sarcomas, which show low signal intensity on T1WI and high signal intensity on T2WI. However, the MRI signal depends on the amount of melanin in the tumour. A low melanin content may not lead to an increase in signal intensity on T1-weighted images[12]. On the other hand, connective tissue, necrosis, cystic degeneration, and other factors within the lesion can also reduce the signal intensity on T1-weighted images and increase the signal intensity on T2-weighted images. Other components within the lesion, such as fat, haemoglobin, and protein fluid, can also cause changes in MRI signals. However, in most cases, the combination of multiple MRI techniques, including fat suppression, sensitivity-weighted imaging, and enhanced MRI, can accurately diagnose CCS.

At present, the treatment method for CCS is surgical resection[13]. MRI can effectively display the edge of the lesion, the extent of surrounding oedema, and the extent of fat involvement, which is highly important for guiding surgical resection and predicting postoperative recurrence. Tumour cells may be present in the oedematous area surrounding the tumour, which can predict the risk of local recurrence[14,15]. One study[16] revealed that if tumour cells are found in the reactive oedematous area around the tumour, the local recurrence risk may be as high as 40%. The fat gap around the tumour can help clinicians evaluate the involvement of neurovascular bundles and surrounding organs. If the fat gap is eliminated, local invasion may occur.

As an invasive sarcoma, CCS has a high risk of metastasis. Up to 40% of patients experience lymph node metastasis. Remote metastasis mostly occurs in the lungs and bones, and there are also a few reports of metastasis in the gastrointestinal tract, liver, pancreas, and sellar region[17-19]. Insufficient surgical resection, a tumour size > 5 cm, and vascular invasion are important factors for metastasis[20]. Early detection of metastasis during postoperative follow-up is crucial for avoiding a poor prognosis. Regardless of the pathological condition of the resected tumour, MRI or CT should be performed every 1-2 years to assess recurrence at the primary site and to screen for metastasis in the lungs, liver, and bones. If necessary, PET-CT can be performed to evaluate the overall condition of the patient[21].

CONCLUSION

In summary, CCS is an extremely rare sarcoma characterized by invasiveness and high rates of postoperative recurrence and metastasis. The clinical symptoms are often atypical, and early diagnosis is difficult. MRI have high soft tissue resolution, and multiple signals can reflect the internal components of the tumours. It can help with early diagnosis, surgical guidance, and recurrence and metastasis detection during postoperative follow-up, which has important clinical significance.

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FOOTNOTES

Author contributions: Wang WJ and Hui DM designed the case report; Feng JB, Li CM collected the data; Li CM performed data analysis; Wang X revised the manuscript. All authors have read and approved the final manuscript. Wang WJ, Wang X and have contributed equally to this study. Feng JB and Li CM made significant contributions to the research process and the publication of the paper, they were responsible for the selection of the research topic, the paper design, the data analysis, and the writing of the article, as well as for the communication with the journal editors and the other authors, and they were collectively responsible for the content of the paper and the process of submitting the paper for publication, and therefore acted as the co-corresponding authors.

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REFERENCES

1 **Liu YJ**, Zou C, Wu YY. Metastatic clear cell sarcoma of the pancreas: A rare case report. *World J Clin Cases* 2024; **12**: 1448-1453 [PMID: 38576799 DOI: 10.12998/wjcc.v12.i8.1448]
2 **Cornillie J**, van Cann T, Wozniak A, Hompes D, Schöffski P. Biology and management of clear cell sarcoma: state of the art and future perspectives. *Expert Rev Anticancer Ther* 2016; **16**: 839-845 [PMID: 27253849 DOI: 10.1080/14737140.2016.1197122]
3 **Stacchiotti S**, Frezza AM, Blay JY, Baldini EH, Bonvalot S, Bovée JVMG, Callegaro D, Casali PG, Chiang RC, Demetri GD, Demicco EG, Desai J, Eriksson M, Gelderblom H, George S, Gounder MM, Gronchi A, Gupta A, Haas RL, Hayes-Jardon A, Hohenberger P, Jones KB, Jones RL, Kasper B, Kawai A, Kirsch DG, Kleinerman ES, Le Cesne A, Lim J, Chirlaque López MD, Maestro R, Marcos-Gragera R, Martin Broto J, Matsuda T, Mir O, Patel SR, Raut CP, Razak ARA, Reed DR, Rutkowski P, Sanfilippo RG, Sbaraglia M, Schaefer IM, Strauss DC, Sundby Hall K, Tap WD, Thomas DM, van der Graaf WTA, van Houdt WJ, Visser O, von Mehren M, Wagner AJ, Wilky BA, Won YJ, Fletcher CDM, Dei Tos AP, Trama A. Ultra-rare sarcomas: A consensus paper from the Connective Tissue Oncology Society community of experts on the incidence threshold and the list of entities. *Cancer* 2021; **127**: 2934-2942 [PMID: 33910263 DOI: 10.1002/cncr.33618]
4 **Dim DC**, Cooley LD, Miranda RN. Clear cell sarcoma of tendons and aponeuroses: a review. *Arch Pathol Lab Med* 2007; **131**: 152-156 [PMID: 17227118 DOI: 10.5858/2007-131-152-CCSOTA]

5 **Kuiper DR**, Hoekstra HJ, Veth RP, Wobbes T. The management of clear cell sarcoma. *Eur J Surg Oncol* 2003; **29**: 568-570 [PMID: 12943620 DOI: 10.1016/s0748-7983(03)00115-x]

6 **Gonzaga MI**, Grant L, Curtin C, Gootee J, Silberstein P, Voth E. The epidemiology and survivorship of clear cell sarcoma: a National Cancer Database (NCDB) review. *J Cancer Res Clin Oncol* 2018; **144**: 1711-1716 [PMID: 29961184 DOI: 10.1007/s00432-018-2693-6]

7 **Sharma K**, Yadav SK, Valluru B, Liu L. Significance of MRI in the diagnosis and differentiation of clear cell sarcoma of tendon and aponeurosis (CCSTA): A case report. *Medicine (Baltimore)* 2018; **97**: e11012 [PMID: 30075493 DOI: 10.1097/MD.0000000000011012]

8 **Casali PG**, Abecassis N, Aro HT, Bauer S, Biagini R, Bielack S, Bonvalot S, Boukovinas I, Bovee JVMG, Brodowicz T, Broto JM, Buonadonna A, De Álava E, Dei Tos AP, Del Muro XG, Dileo P, Eriksson M, Fedenko A, Ferraresi V, Ferrari A, Ferrari S, Frezza AM, Gasperoni S, Gelderblom H, Gil T, Grignani G, Gronchi A, Haas RL, Hassan B, Hohenberger P, Issels R, Joensuu H, Jones RL, Judson I, Jutte P, Kaal S, Kasper B, Kopeckova K, Krákorová DA, Le Cesne A, Lugowska I, Merimsky O, Montemurro M, Pantaleo MA, Piana R, Picci P, Piperno-Neumann S, Pousa AL, Reichardt P, Robinson MH, Rutkowski P, Safwat AA, Schöffski P, Sleijfer S, Stacchiotti S, Sundby Hall K, Unk M, Van Coevorden F, van der Graaf WTA, Whelan J, Wardelmann E, Zaikova O, Blay JY; ESMO Guidelines Committee and EURACAN. Soft tissue and visceral sarcomas: ESMO-EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol* 2018; **29**: iv51-iv67 [PMID: 29846498 DOI: 10.1093/annonc/mdy096]

9 **Kosemehmetoglu K**, Folpe AL. Clear cell sarcoma of tendons and aponeuroses, and osteoclast-rich tumour of the gastrointestinal tract with features resembling clear cell sarcoma of soft parts: a review and update. *J Clin Pathol* 2010; **63**: 416-423 [PMID: 20418233 DOI: 10.1136/jcp.2008.057471]

10 **Yamada K**, Ohno T, Aoki H, Semi K, Watanabe A, Moritake H, Shiozawa S, Kunisada T, Kobayashi Y, Toguchida J, Shimizu K, Hara A, Yamada Y. EWS/ATF1 expression induces sarcomas from neural crest-derived cells in mice. *J Clin Invest* 2013; **123**: 600-610 [PMID: 23281395 DOI: 10.1172/JCI63572]

11 **Hocar O**, Le Cesne A, Berissi S, Terrier P, Bonvalot S, Vanel D, Auperin A, Le Pechoux C, Bui B, Coindre JM, Robert C. Clear cell sarcoma (malignant melanoma) of soft parts: a clinicopathologic study of 52 cases. *Dermatol Res Pract* 2012; **2012**: 984096 [PMID: 22693489 DOI: 10.1155/2012/984096]

12 **De Beuckeleer LH**, De Schepper AM, Vandevenne JE, Bloem JL, Davies AM, Oudkerk M, Hauben E, Van Marck E, Somville J, Vanel D, Steinbach LS, Guinebretière JM, Hogendoorn PC, Mooi WJ, Verstraete K, Zaloudek C, Jones H. MR imaging of clear cell sarcoma (malignant melanoma of the soft parts): a multicenter correlative MRI-pathology study of 21 cases and literature review. *Skeletal Radiol* 2000; **29**: 187-195 [PMID: 10855466 DOI: 10.1007/s002560050592]

13 **Ikuta K**, Nishida Y, Imagama S, Tanaka K, Ozaki T. The current management of clear cell sarcoma. *Jpn J Clin Oncol* 2023; **53**: 899-904 [PMID: 37451697 DOI: 10.1093/jjco/hyad083]

14 **Gerrand CH**, Wunder JS, Kandel RA, O'Sullivan B, Catton CN, Bell RS, Griffin AM, Davis AM. Classification of positive margins after resection of soft-tissue sarcoma of the limb predicts the risk of local recurrence. *J Bone Joint Surg Br* 2001; **83**: 1149-1155 [PMID: 11764430 DOI: 10.1302/0301-620x.83b8.12028]

15 **White LM**, Wunder JS, Bell RS, O'Sullivan B, Catton C, Ferguson P, Blackstein M, Kandel RA. Histologic assessment of peritumoral edema in soft tissue sarcoma. *Int J Radiat Oncol Biol Phys* 2005; **61**: 1439-1445 [PMID: 15817348 DOI: 10.1016/j.ijrobp.2004.08.036]

16 **Robinson E**, Bleakney RR, Ferguson PC, O'Sullivan B. Oncodiagnosis panel: 2007: multidisciplinary management of soft-tissue sarcoma. *Radiographics* 2008; **28**: 2069-2086 [PMID: 19001660 DOI: 10.1148/rg.287085167]

17 **Insabato L**, Guadagno E, Natella V, Somma A, Bihl M, Pizzolorusso A, Mainenti PP, Apice G, Tornillo L. An unusual association of malignant gastrointestinal neuroectodermal tumor (clear cell sarcoma-like) and Ewing sarcoma. *Pathol Res Pract* 2015; **211**: 688-692 [PMID: 26163185 DOI: 10.1016/j.prp.2015.06.001]

18 **Doglietto F**, Daffini L, Fazzari E, Cominelli M, Pagani F, Poliani PL. Sellar metastasis from clear cell sarcoma: Description of the first case. *Clin Neuropathol* 2022; **41**: 122-127 [PMID: 35200135 DOI: 10.5414/NP301448]

19 **Bai C**, Dong M, Shen W. Rectal clear cell sarcoma-a case report. *Transl Cancer Res* 2020; **9**: 6528-6533 [PMID: 35117261 DOI: 10.21037/tcr-20-1771]

20 **von Konow A**, Ghanei I, Styring E, Vult von Steyern F. Late Local Recurrence and Metastasis in Soft Tissue Sarcoma of the Extremities and Trunk Wall: Better Outcome After Treatment of Late Events Compared with Early. *Ann Surg Oncol* 2021; **28**: 7891-7902 [PMID: 33861406 DOI: 10.1245/s10434-021-09942-8]

21 **Ibrahim RM**, Steenstrup Jensen S, Juel J. Clear cell sarcoma-A review. *J Orthop* 2018; **15**: 963-966 [PMID: 30210202 DOI: 10.1016/j.jor.2018.08.039]

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