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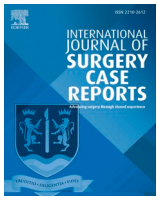

Case report

Extraskeletal mesenchymal chondrosarcoma of the distal thumb: A case report

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ABSTRACT

Introduction and importance: Chondrosarcoma is the most common primary malignant bone tumor of the hand, Mesenchymal chondrosarcoma (MCS) is a rare subtype of chondrosarcoma, extraosseous mesenchymal chondrosarcoma (EMCS) is even rarer.

Case presentation: We presented a case of primary EMCS involving the distal thumb in a 59-year-old man. The patient had abnormally grown nails and a higher skin temperature than the contralateral finger. After admission to our hospital, we found that the tumor has the possibility of malignant transformation, but imageological examinations did not reveal invasive bone lesions and calcification. Finally, due to the patient's strong desire to preserve the thumb, a conservative surgical approach was selected to preserve the patient's finger function, and histological examination indicated MCS.

Conclusion: EMCS is a highly malignant tumor with strong tendency of invasion and metastasis. Imaging is sometimes difficult to diagnose, and pathology is a necessary part of distinguishing between benign and malignant tumors. Operation is the major treatment to EMCS, meanwhile, adjuvant radiotherapy and chemotherapy also be effective. The overall prognosis is poor.

1. Introduction

MCS is a rare subtype of chondrosarcoma and is extremely rare in the soft tissue of the distal thumb. To the best of our knowledge, it is little to the point of nobody report that EMCS occurs in the distal thumb.

We reported a case of primary EMCS involving the distal thumb of the dominant hand in a 59-year-old man [1].

2. Case report

A 59-year-old man was admitted to the hospital due to the gradual enlargement of the right thumb swelling for more than one year. We found that the right distal of thumb has abnormal nail growth and the skin temperature of the right thumb is higher than the opposite side (Fig. 1). The flexion of the interphalangeal joint of the thumb is limited. After an incisional biopsy at the local hospital, the tumor enlarged continuously and the pain worsened. Simultaneously the pathological results of the local hospital suggested chondroma. After admission to our hospital, the patient was scanned by MRI which demonstrated a partial in-defined boundary of the tumor. The tumor involved the periosteum of the distal phalanx and adjacent soft tissues. X-ray and computed tomography (CT) resulted in no invasive bone lesions and calcification (Fig. 2). Accordingly, the tumor has the possibility of malignant transformation. The patient's chest CT scan and laboratory findings were normal, including the alkaline phosphatase and the erythrocyte sedimentation rate. Considering the patient's strong desire to preserve the finger and the great function of the thumb, we finally chose the conservative surgical way to retain the finger function of the patient. During the operation, the tumor could be seen the distal thumb, which was separated along the boundary and completely removed. Meanwhile, we preserved 1/3 of the nail bed and removed 5 mm of the distal phalanx (Fig. 3). A volar digital advancement flap was designed to repair the fingertip defect. The specimen size is about 2.0 cm * 1.6 cm * 1.7 cm, tough in texture and gray in color.

Abbreviations: MCS, mesenchymal chondrosarcoma; EMCS, extraosseous mesenchymal chondrosarcoma; CT, computed tomography; MR, magnetic resonance; MRI, magnetic resonance imaging; T1WI, T1-weighted imaging; T2WI, T2-weighted imaging.

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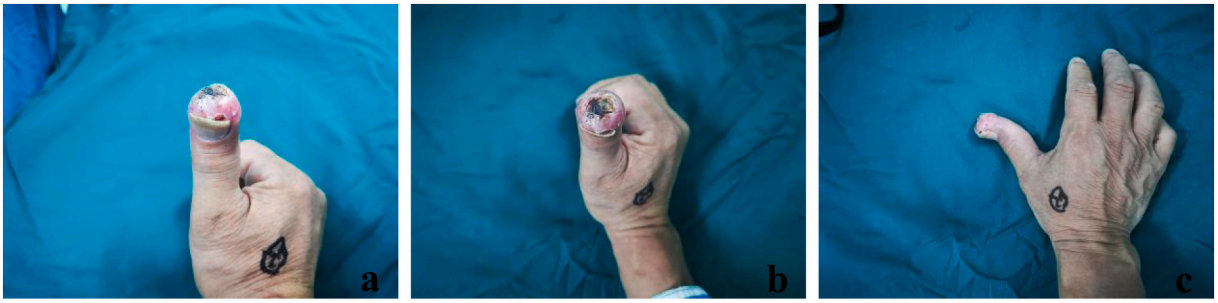

Fig. 1. Preoperative appearance: right thumb mass and abnormal nail (a–c).

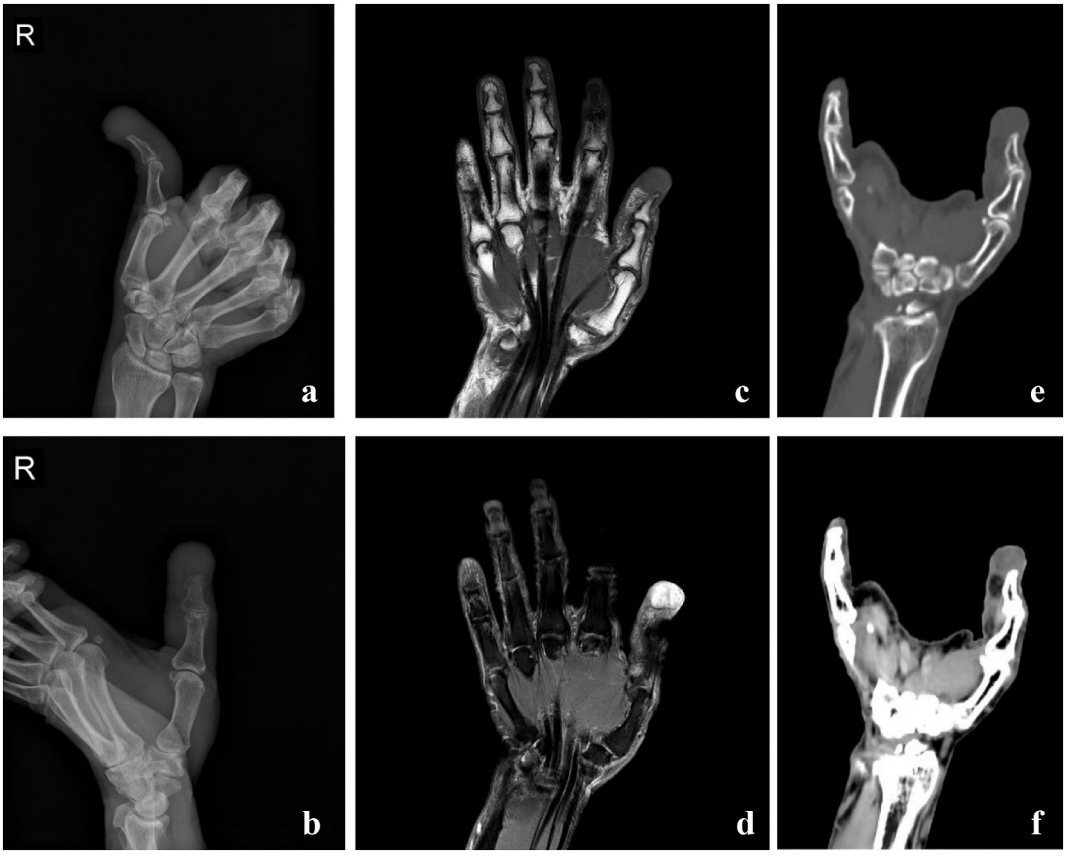

Fig. 2. X-ray and computed tomography (CT) resulted in no invasive bone lesions and calcification(a–d). MRI showing the tumor has a isointensity on T1WI and heterogeneous hyperintensity on T2WI (e–f).

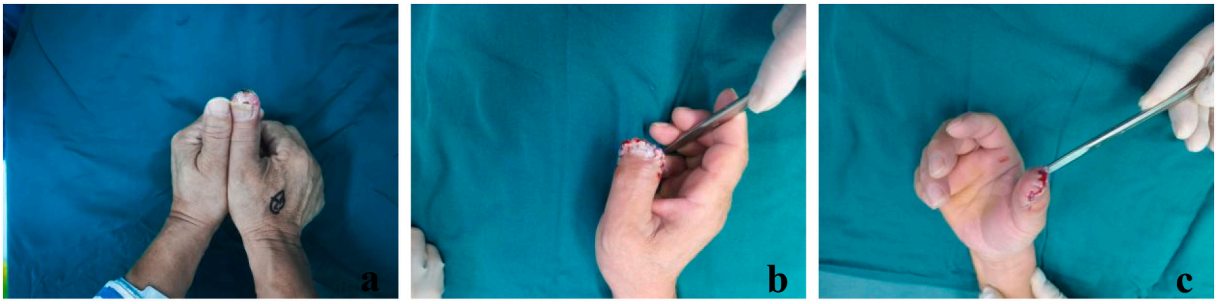

Fig. 3. Postoperative appearance: the tumor has been removed and one-third of the nail bed was preserved (a–c).

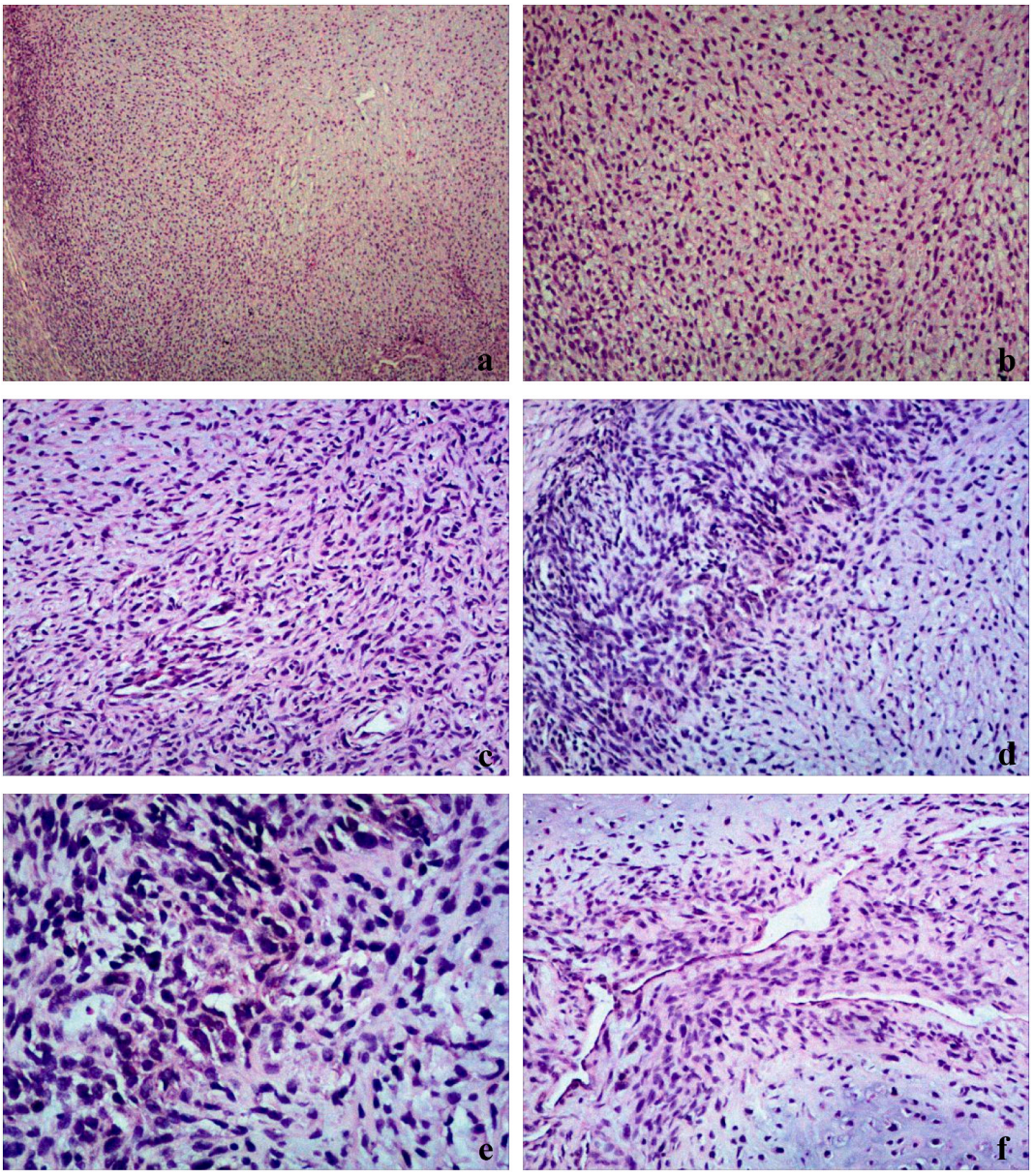

Fig. 4. Photomicrograph showing the tumor originated from the mesenchymal tissue (hematoxylin&eosin, X4&X10) (a–b).
Photomicrograph showing an increase in cellularity can be seen, the neoplastic cells show pleomorphism, with hyperchromatic nuclei (hematoxylin&eosin, X20&X40) (c–e).
Photomicrograph showing the well differentiated hyaline cartilage and undifferentiated small round cells, antler vessels were seen in the interstitium. (hematoxylin&eosin, X40) (f).

The histological examination revealed that chondrocytes were highly differentiated and there were a large number of antlers blood vessels in the interstitium (Fig. 4). Immunohistochemical staining results: 3#, Vim (+), S100 (scattered +), CD99 (+), SMA (−), P53 (−), P16 (+), Ki67 (local +, 45 %). The diagnosis indicated mesenchymal chondrosarcoma.

3. Discussion

Chondrosarcoma is the most common primary malignant bone tumor of the hand, and MCS, a rare subtype of chondrosarcoma, was first reported by Lichtenstein and Bernstein in 1959, account for about 1 % of the total incidence, EMCS is even rarer [2]. The majority of EMCS occurs in the central nervous system and soft tissues, with a slightly female predominance [3–5]. It is easy to be misdiagnosed, because the lack of specificity of the clinical manifestations of EMCS. Some scholars believe that the abnormal growth of nail has a warning effect among the special groups of population [6]. Our case is a 59 year old man with pain in the right thumb, increasing mass, and abnormal fingernail growth. In addition, the skin temperature of the right thumb is higher than the opposite side. The flexion of the interphalangeal joint of the thumb is limited. After an incisional biopsy at the local hospital, the tumor enlarged continuously and the pain worsened.

The imaging features of EMCS are similar to those of various benign and malignant lesions, such as high myxoid tumors, myositis ossificans, etc. It is reported that calcification is common in EMCS (67 %) [7,8]. Scattered calcifications can be found on radiographs or CT scans, typically characterized by “ring and arc calcifications”. Due to the difference in signal intensity between calcified tissue (low signal) and non-mineralized tissue (high signal), MRI typical features of EMCS may appear the “black pepper” sign on T2WI and hypointensity or isointensity on T1-weighted imaging (T1WI). In this case, we did not find

significant calcification in the tumor. X-ray and computed tomography (CT) resulted in no invasive bone lesions and calcification. MR imaging, the tumor has a isointensity on T1WI and heterogeneous hyperintensity on T2WI (Fig. 2). It's hard to differentiate from imaging alone.

Pathology is an essential part of distinguishing between benign and malignant tumors, and histological grading affects patient prognosis [9,10]. EMCS comprisie a number of undifferentiated small round cells with islands of well differentiated hyaline cartilage [10–12]. Immunohistochemistry helps to confirm the diagnosis, such as S-100, CD99, Vimentin, Sox9, etc. In our case, chondrocytes were highly differentiated and antler vessels were seen in the interstitium. Immunohistochemical results were consistent with the diagnosis of EMCS: Vim (+), S100 (scattered +), CD99 (+).

MCS, a highly malignant chondrosarcoma, is mainly treated by extensive excision, finger amputation or ray amputation combined with adjuvant radiotherapy and chemotherapy [11,13]. However, most chondrosarcomas of the hand are low-grade malignant tumors, and the treatment of low-grade malignant chondrosarcomas is controversial. Some researchers believe that chondrosarcoma, especially the finger, is needed to be treated differently from chondrosarcoma of the pelvis or scapula-and that amputation or disabling surgery is rarely necessary. Bovee et al. [14] observed tumors located at the level of the phalanges of the hands and feet, which were locally aggressive and had little likelihood of metastasis. By Mankin's conjecture, chondrosarcoma of the finger may be considered a different benign disease than chondrosarcoma of other skeletal sites, and a finger-sparing approach to surgical management of these tumors is recommended [15]. Roberts and Lee FY et al. [16,17] believed that the temperature of the hands and feet and the location of the tumor limited the ability of the tumor to grow locally, and particularly to metastasize. Secondly, smaller tumors have lower rates of metastasis and mortality than larger tumors. The patient's tumor is an EMCS, which is aggressive and prone to metastasis. Some studies reported that 68.2 % 5-year disease-free survival in children and young adults [18]. Extended resection or finger amputation should be adopted in terms of treatment. Most chondrosarcomas of the hand do not respond to radiation therapy or chemotherapy, except for MCS [13,19]. Because of the patient's strong desire to preserve the thumb, we did not perform another amputation. Long-term follow-up and monitoring is required after surgery.

4. Conclusion

In conclusion, EMCS is a highly malignant tumor with strong tendency of invasion and metastasis. Imaging is sometimes difficult to diagnose, and pathology is a necessary part of distinguishing between benign and malignant tumors. Operation is the major treatment to EMCS, meanwhile, adjuvant radiotherapy and chemotherapy also be effective. The overall prognosis is poor.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Ethical approval is exempt at our institution.

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Declaration of competing interest

No conflicts of interest.

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References

[1] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, International Journal of Surgery 84 (2020) 226–230.
[2] L. Lightenstein, D. Bernstein, Unusual benign and malignant chondroid tumors of bone. A survey of some mesenchymal cartilage tumors and malignant chondroblastic tumors, including a few multicentric ones, as well as many atypical benign chondroblastomas and chondromyxoid fibromas, Cancer 12 (1959) 1142–1157.
[3] N. Ali, D. Vanel, A. Righi, M. Colangeli, M. Manfrini, Imaging features of extraskeletal mesenchymal chondrosarcoma of the hand, Diagn. Interv. Imaging 96 (9) (2015) 965–967.
[4] L.G. Shapeero, D. Vanel, D. Couanet, G. Contesso, L.V. Ackerman, Extraskeletal mesenchymal chondrosarcoma, Radiology 186 (3) (1993) 819–826.
[5] C. Louvet, A. de Gramont, M. Krulik, et al., Extraskeletal mesenchymal chondrosarcoma: case report and review of the literature, J. Clin. Oncol. 3 (6) (1985) 858–863.
[6] S. Au, M.E. Juhl, R. Emmadi, A.L. Krunic, Nail dystrophy as a presenting sign of a chondrosarcoma of the distal phalanx - case report and review of the literature, Acta Derm. Venereol. 95 (8) (2015) 1026–1027.
[7] Y. Chen, X. Wang, L. Guo, et al., Radiological features and pathology of extraskeletal mesenchymal chondrosarcoma, Clin. Imaging 36 (4) (2012) 365–370.
[8] M.D. Murphey, E.A. Walker, A.J. Wilson, M.J. Kransdorf, H.T. Temple, F. H. Gannon, From the archives of the AFIP: imaging of primary chondrosarcoma: radiologic-pathologic correlation, Radiographics 23 (5) (2003) 1245–1278.
[9] D.J. Pritchard, R.J. Lunke, W.F. Taylor, D.C. Dahlin, B.E. Medley, Chondrosarcoma: a clinicopathologic and statistical analysis, Cancer 45 (1) (1980) 149–157.
[10] H.L. Evans, A.G. Ayala, M.M. Romsdahl, Prognostic factors in chondrosarcoma of bone: a clinicopathologic analysis with emphasis on histologic grading, Cancer 40 (2) (1977) 818–831.
[11] K. Arora, N.D. Riddle, Extraskeletal mesenchymal chondrosarcoma, Arch Pathol Lab Med. 142 (11) (2018) 1421–1424.
[12] M.D. Murphey, E.A. Walker, A.J. Wilson, M.J. Kransdorf, H.T. Temple, F. H. Gannon, From the archives of the AFIP: imaging of primary chondrosarcoma: radiologic-pathologic correlation, Radiographics. 23 (5) (2003) 1245–1278.
[13] S. Kawaguchi, I. Weiss, P.P. Lin, W.W. Huh, V.O. Lewis, Radiation therapy is associated with fewer recurrences in mesenchymal chondrosarcoma, Clin. Orthop. Relat. Res. 472 (3) (2014) 856–864.
[14] J.V. Bovée, R.O. van der Heul, A.H. Taminiau, P.C. Hogendoorn, Chondrosarcoma of the phalanx: a locally aggressive lesion with minimal metastatic potential: a report of 35 cases and a review of the literature, Cancer 86 (9) (1999) 1724–1732.
[15] H.J. Mankin, Chondrosarcomas of digits: are they really malignant? Cancer 86 (9) (1999) 1635–1637.
[16] P.H. Roberts, C.H. Price, Chondrosarcoma of the bones of the hand, J. Bone Joint Surg. Br. 59 (2) (1977) 213–221.
[17] F.Y. Lee, H.J. Mankin, G. Fondren, et al., Chondrosarcoma of bone: an assessment of outcome, J. Bone Joint Surg. Am. 81 (3) (1999) 326–338.
[18] M.W. Bishop, J.M. Somerville, A. Bahrami, et al., Mesenchymal chondrosarcoma in children and young adults: a single institution retrospective review, Sarcoma 2015 (2015), 608279.
[19] M. Cesari, F. Bertoni, P. Bacchini, M. Mercuri, E. Palmerini, S. Ferrari, Mesenchymal chondrosarcoma. An analysis of patients treated at a single institution, Tumori 93 (5) (2007) 423–427.