MUSCULOSKELETAL IMAGING
Iran J Radiol. 2023 January; 20(1):e133421. https://doi.org/10.5812/iranjradiol-133421.
Published online 2023 February 15. Case Report

Subcutaneous Myoepithelial Carcinoma of the Ankle

Hyerim Park [1], Yu Sung Yoon [2,*] and Hee Kyung Kim [3]

[1]Department of Radiology, Soonchunhyang University Cheonan Hospital, Soonchunhyang University College of Medicine, Cheonan, Rebuplic of Korea
[2]Department of Radiology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, Bucheon, Rebuplic of Korea
[3]Department of Pathology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, Bucheon, Rebuplic of Korea

[*]*Corresponding author*: Department of Radiology, Soonchunhyang University Bucheon Hospital, Soonchunhyang University College of Medicine, 170 Jomaru-ro, Bucheon 14584, Bucheon, Republic of Korea. Tel: +82-326215848, Email: zick1853@gmail.com

Received 2022 November 27; **Revised** 2023 January 15; **Accepted** 2023 January 17.

Abstract

Introduction: Myoepithelial carcinomas are rare malignant neoplasms, which are known to arise from the salivary glands or other organs.
Case Presentation: Here, we present a case of myoepithelial carcinoma in a 64-year-old woman, presenting to a clinic with a painful ankle mass for the past three years. Ultrasonography showed a 1 cm, iso- to hypoechoic mass on the medial malleolus of the right ankle. Magnetic resonance imaging (MRI) revealed a well-defined mass with iso-signal intensity on T1-weighted images and high signal intensity on T2-weighted images. On the enhanced image, the mass showed homogenous enhancement. In diffusion-weighted images with a high b-value, the lesion showed restriction with a low apparent diffusion coefficient (ADC). Surgical removal was performed, and pathological analysis revealed myoepithelial carcinoma.
Conclusion: Myoepithelial carcinoma of the extrasalivary glands is a very uncommon phenomenon. Despite its rarity, it should be considered as a differential diagnosis when a soft tissue mass of the ankle shows malignant potential in radiological findings.

Keywords: Myoepithelial Carcinoma, Ankle, Magnetic Resonance Imaging

1. Introduction

Myoepithelial carcinoma, also known as malignant myoepithelioma, is a rare tumor, which was first described by Stromeyer et al. in 1975 (1). These tumors have been reported to arise from the salivary glands or other areas, such as cutaneous, subcutaneous, and deep soft tissues of the head and neck regions and upper and lower extremities (2, 3). Myoepithelial carcinomas typically occur in the salivary glands, and most commonly, in the parotid gland (4). Clinically, the majority of patients present with a palpable, usually painless mass (5). The standard treatment involves wide surgical resection with clear margins.

Due to the possibility of recurrence or metastasis following surgery, radiation therapy is recommended (4). Myoepithelial tumors of soft tissue show an equal distribution in males and females, with the highest incidence reported in young to middle-aged adults (5). According to the literature, myoepithelial carcinoma recurs in 40% of patients and metastasizes in 22% of cases (6). The common sites of metastasis include the lungs, liver, bone, and soft tissue (7).

Herein, we report a rare case of myoepithelial carcinoma in a 64-year-old woman with pain in the right ankle. To the best of our knowledge, this is the second report of ankle myoepithelioma in the English literature and the first one in the field of radiology. In this case report, the radiological features, including ultrasonography and magnetic resonance imaging (MRI) findings of the patient, as well as the pathological features of myoepithelial carcinoma of the ankle, were described.

2. Case Presentation

A 64-year-old woman with a history of painful palpable mass in her right ankle over the past three years visited our clinic. Physical examination revealed a bluish colored lump on the medial malleolus (Figure 1). The lesion was also firm and fixed. She complained of an itching sensation with mild pain. Besides, she had high blood pressure and hyperlipidemia and was on medication. She had no trauma or surgical history. The subsequent ultrasonography (Figure 2) indicated a 1 cm hypoechoic mass in the subcutaneous fat layer in the medial malleolar area. The mass exhibited a solid portion and vascularity on color Doppler images.

The ankle MRI (Figure 3) revealed a 1 cm, well-defined nodular lesion on the medial malleolus of the right ankle. The lesion showed hypersignal intensity on T2-

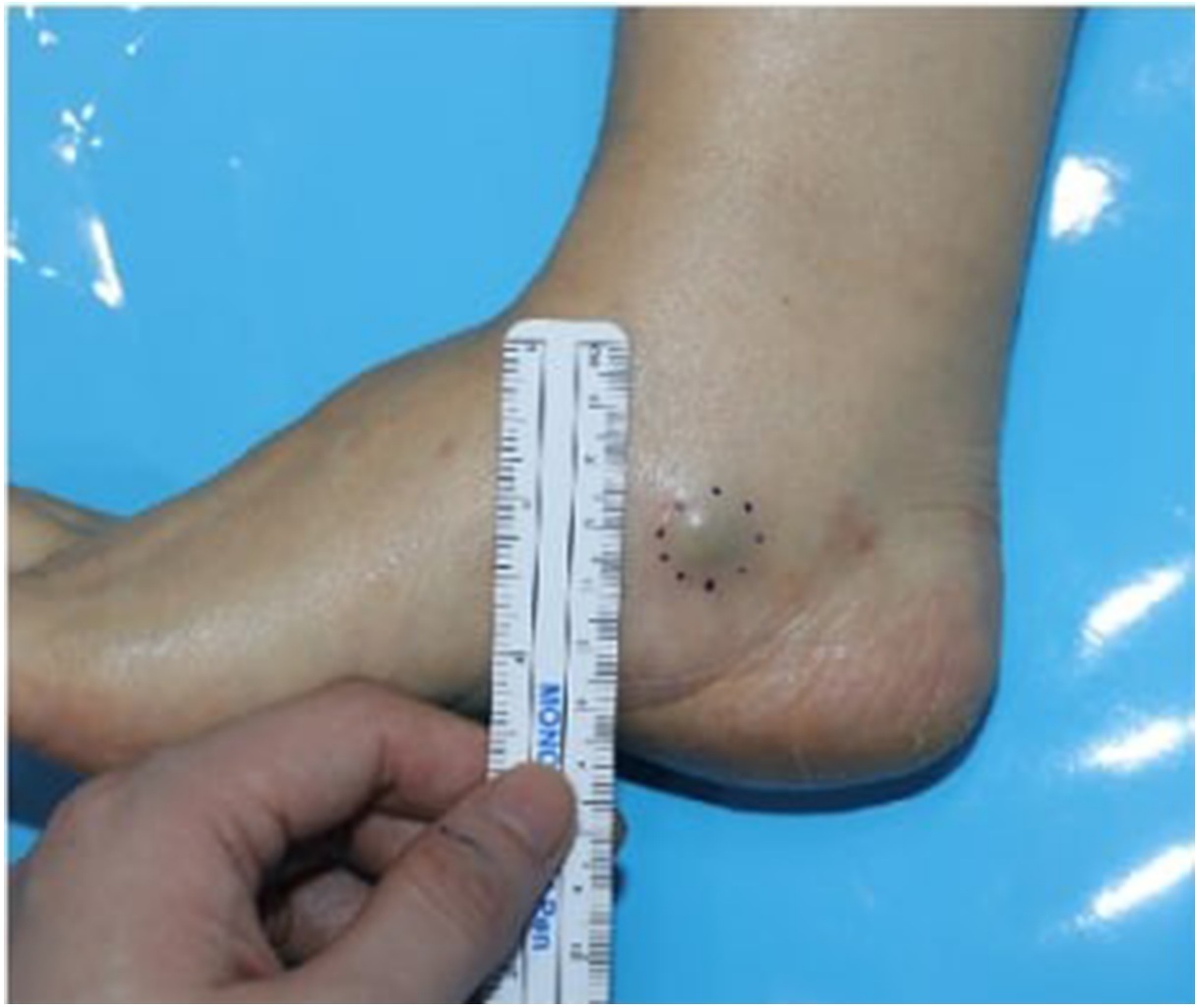

Figure 1. A 64-year-old female with a history of painful palpable mass for three years in her right ankle. In gross examination, the lesion shows a bluish colored protruding mass on the medial ankle.

weighted images and intermediate signal intensity on T1-weighted images. Following gadolinium injection, the lesion showed homogeneous enhancement on fat-suppressed post-contrast T1-weighted images. High b-value (b = 1000) diffusion-weighted imaging (DWI) showed restriction of the lesion with a low apparent diffusion coefficient (ADC). There were no adjacent soft tissue changes or skin involvement.

The patient underwent surgical excisional biopsy. The tumor was arranged in nests or large sheets with epithelioid cells in collagenous stroma. Tumor cells showed high-grade cytologic atypia and frequent mitoses. Based on immunohistochemistry, the tumor cells were strongly positive for S-100 protein (Figure 4). According to the histological findings, the tumor was diagnosed as myoepithelial carcinoma.

3. Discussion

Myoepithelial carcinoma is a rare tumor, which most frequently occurs in the parotid glands (4). The most common site of soft tissue myoepithelial carcinoma is the lower extremity, as reported in 38% of cases (8). The recurrence rate of soft tissue myoepithelial carcinoma is known to be as high as 37 - 42% following treatment (8, 9). Since soft tissue myoepithelial carcinoma commonly presents as distant metastasis and develops in the lungs, kidneys, brain, and skin, chest computed tomography (CT)

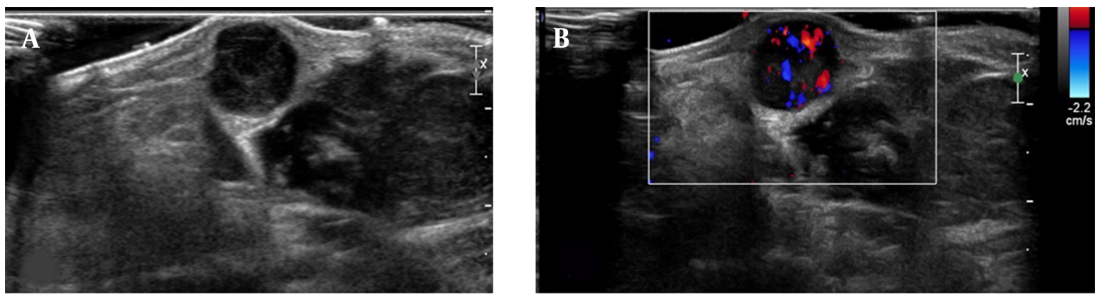

Figure 2. Transverse ultrasonography (A), and color Doppler (B), images show a hypoechoic solid mass in the subcutaneous fat layer with intralesional hypervascularity.

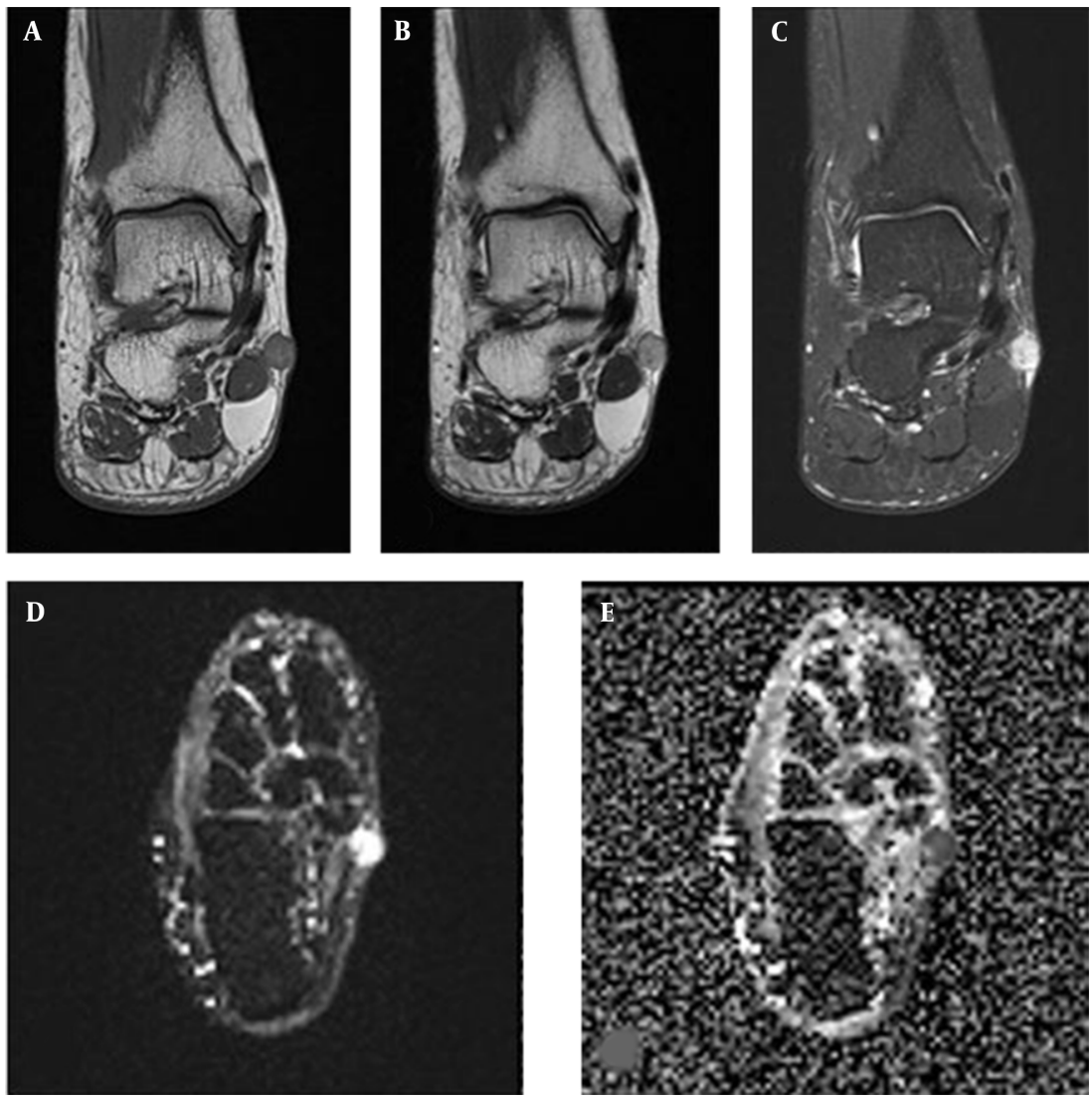

Figure 3. The MRI of the right ankle for further evaluation of the painful soft tissue mass. Coronal T1-weighted image (A), T2-weighted image (B), and contrast-enhanced fat-suppressed T1-weighted image (C), show a 1 cm, well-defined mass with iso-signal intensity on T1-weighted images and high signal intensity on T2-weighted images. After contrast injection, the lesion shows homogenous enhancement. The high b-value (b = 1000) diffusion-weighted image (D), shows restricted diffusion with a low apparent diffusion coefficient (ADC) (E).

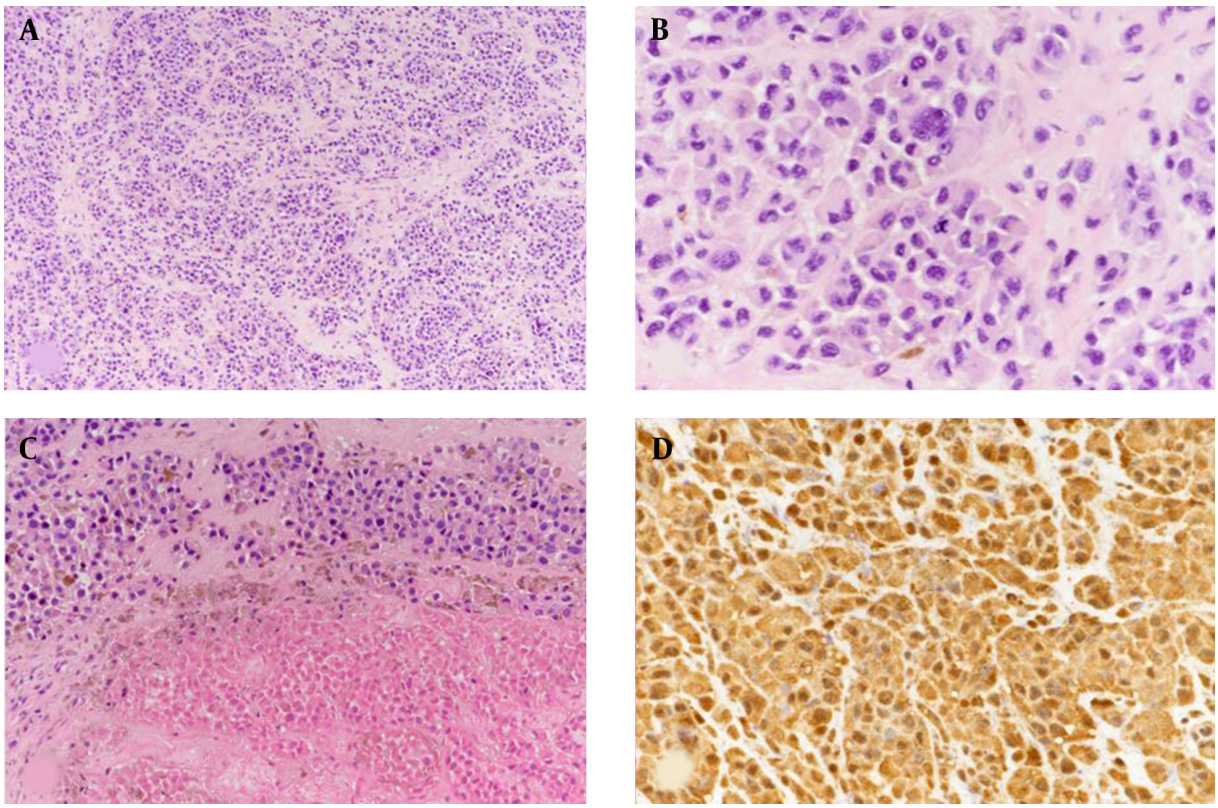

Figure 4. The histological features of the specimens. The tumor is arranged in nests or large sheets with epithelioid cells in collagenous stroma (A) (hematoxylin and eosin [H&E] staining, 100 × magnification). The tumor cells are pleomorphic epithelioid or plasmacytoid cells with high-grade cytological atypia and frequent mitoses (B) (H&E staining, 400 × magnification). Also, tumor necrosis can be found (C) (H&E staining, 200 × magnification). Immunohistochemical staining for S-100 protein is strongly positive (D) (400 × magnification).

and abdominal CT scan or positron emission tomography (PET)/CT can be useful for the initial staging workup (9-11).

The imaging characteristics of myoepithelial tumors in soft tissues are not well-known due to their rarity. Some studies have reported that soft tissue myoepithelial tumors are relatively well-defined lobulated masses with heterogeneous enhancement on contrast-enhanced CT scans. On MRI, tumors show low to intermediate or high signal intensity areas, representing hemorrhage on T1-weighted images and heterogeneous high intensity areas with low-signal-intensity internal septa on T2-weighted images (12, 13). Since there is an overlap between benign and malignant lesions without specific imaging findings, pathological confirmation is necessary for diagnosis.

In our case, the mass showed hypersignal intensity on the T2-weighted image and intermediate signal intensity on the T1-weighted image; these results appear to be similar to a previous report. On DWI, our patient showed diffusion restriction; although little is known, this finding is consistent with the results of a previous study (14). According to the literature, myoepithelial carcinoma of the tibia shows restricted diffusion, reflecting a large cellular microenvironment on DW-MRI with a low ADC. These findings suggest that DWI and ADC values can be used for differentiation of benign and malignant myoepithelial tumors (14).

Regarding treatment, surgical excision is the first option. Chemotherapy may be used for metastatic myoepithelial carcinoma (10). Radiation therapy can be used as the first-line treatment when surgery is contraindicated or as an adjuvant treatment when the risk of recurrence or metastasis is high (4). However, a study reported that radiation therapy is not effective for a recurrent or metastatic disease (15).

In conclusion, myoepithelial carcinoma of the ankle is an uncommon phenomenon, and there are few published reports describing the imaging features of myoepithelial carcinoma. We hope that our case can help radiologists diagnose myoepithelial carcinoma in practice.

Acknowledgments

This study was supported by Soonchunhyang University Research Fund.

Footnotes

Authors' Contributions: Study concept and design: H. P. and Y. S. Y.; acquisition of data: Y. S. Y. and H. P.; analysis and interpretation of data: H. P.; drafting of the manuscript: H. P. and H. K. K.; critical revision of the manuscript for important intellectual content: Y. S. Y.; all authors approved the final manuscript administrative, technical, and material support: H.K.K.; and study supervision: Y. S. Y.

Conflict of Interests: The authors declare that they have no conflict of interest.

Funding/Support: The authors received no funding for the publication of this study.

Informed Consent: Written informed consent was obtained from the patient.

References

1. Stromeyer FW, Haggitt RC, Nelson JF, Hardman JM. Myoepithelioma of minor salivary gland origin. Light and electron microscopical study. *Arch Pathol*. 1975;**99**(5):242–5.
2. Lee JR, Georgi DE, Wang BY. Malignant myoepithelial tumor of soft tissue: a report of two cases of the lower extremity and a review of the literature. *Ann Diagn Pathol*. 2007;**11**(3):190–8. [PubMed ID: 17498593]. https://doi.org/10.1016/j.anndiagpath.2006.04.004.
3. Domingo-Musibay E, Oliveira AM, Okuno SH, Petersen IA, Rose PS, Robinson SI. Myoepithelioma of Soft Tissues: A Single Institution Retrospective Case Series. *Am J Clin Oncol*. 2018;**41**(4):357–61. [PubMed ID: 27139621]. https://doi.org/10.1097/COC.0000000000000292.
4. Xu T, Liao Z, Tang J, Guo L, Qiu H, Gao Y, et al. Myoepithelial carcinoma of the head and neck: A report of 23 cases and literature review. *Cancer Treatment Communications*. 2014;**2**(2-3):24–9. https://doi.org/10.1016/j.ctrc.2014.08.002.
5. WHO Classification of Tumours Editorial Board. *Soft Tissue and Bone Tumours*. 5th ed. Lyon, France: International Agency for Research on Cancer; 2020.
6. Su YX, Roberts DB, Hanna EY, El-Naggar A, Saylam G, Frank SJ, et al. Risk Factors and Prognosis for Myoepithelial Carcinoma of the Major Salivary Glands. *Ann Surg Oncol*. 2015;**22**(11):3701–7. [PubMed ID: 25636455]. https://doi.org/10.1245/s10434-015-4389-3.
7. Skelton E, Jewison AC, Ramesar K, Howlett D. Myoepithelial carcinoma of the parotid: a rare tumour that may provide diagnostic difficulty. *BMJ Case Rep*. 2015;**2015**:bcr2014206163. [PubMed ID: 25721825]. [PubMed Central ID: PMC4342686]. https://doi.org/10.1136/bcr-2014-206163.
8. Gleason BC, Hornick JL. Myoepithelial tumours of skin and soft tissue: an update. *Diagn Histopathol*. 2008;**14**(11):552–62. https://doi.org/10.1016/j.mpdhp.2008.08.005.
9. Hornick JL, Fletcher CD. Myoepithelial tumors of soft tissue: a clinicopathologic and immunohistochemical study of 101 cases with evaluation of prognostic parameters. *Am J Surg Pathol*. 2003;**27**(9):1183–96. [PubMed ID: 12960802]. https://doi.org/10.1097/00000478-200309000-00001.
10. Gleason BC, Fletcher CD. Myoepithelial carcinoma of soft tissue in children: an aggressive neoplasm analyzed in a series of 29 cases. *Am J Surg Pathol*. 2007;**31**(12):1813–24. [PubMed ID: 18043035]. https://doi.org/10.1097/PAS.0b013e31805f6775.
11. Cosentino TB, Brazao-Silva MT, Souza KC, de Faria PR, de Moraes S, Loyola AM, et al. Myoepithelial carcinoma of the submandibular gland: report of a case with multiple cutaneous metastases. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2008;**106**(2):e26–9. [PubMed ID: 18554950]. https://doi.org/10.1016/j.tripleo.2008.04.011.
12. Trevino M, Moorthy C, Kafchinski L, Bustamante D. Foot plantar soft tissue malignant myoepithelioma tumor: Case report and review of the literature. *Clin Imaging*. 2020;**61**:90–4. [PubMed ID: 32000118]. https://doi.org/10.1016/j.clinimag.2019.11.014.
13. Pilavaki M, Givissis P, Tzarou V, Palladas P, Pournaras J. Soft-tissue myoepithelioma of the hypothenar region: a case report. *J Hand Surg Am*. 2007;**32**(5):674–6. [PubMed ID: 17482007]. https://doi.org/10.1016/j.jhsa.2007.02.022.
14. Lin CH, Wu KY, Chen CK, Li CF, Hsieh TJ. Myoepithelial carcinoma of tibia mimic giant cell tumor: a case report with emphasis on MR features. *Skeletal Radiol*. 2019;**48**(10):1637–41. [PubMed ID: 30868231]. https://doi.org/10.1007/s00256-019-03198-w.
15. Bisogno G, Tagarelli A, Schiavetti A, Scarzello G, Ferrari A, Cecchetto G, et al. Myoepithelial carcinoma treatment in children: a report from the TREP project. *Pediatr Blood Cancer*. 2014;**61**(4):643–6. [PubMed ID: 24136896]. https://doi.org/10.1002/pbc.24818.