

Synovial Sarcoma of the Anterior Chest Wall: A Case Report

앞가슴벽에 생긴 윤활막육종: 증례 보고

Mingook Kim, MD¹, Seung Eun Lee, MD^{1*} Joon Hyuk Choi, MD²

Departments of ¹Radiology, ²Pathology, Yeungnam University College of Medicine, Daegu, Korea

Synovial sarcoma is a malignant soft tissue tumor that usually involves the extremities, particularly near the knees; a synovial sarcoma originating in the chest wall is extremely rare. We describe a 26-year-old woman diagnosed with a synovial sarcoma originating in the chest wall, based on CT and MRI findings. Contrast-enhanced CT images revealed a small, well-defined enhancing mass with calcification, in the subpleural area. This lesion was initially diagnosed as a benign tumor; however, the patient developed sudden severe pain with spontaneous bleeding and hemothorax, suggesting the possibility of malignancy. MRI revealed a multilobulated mass with a fluid-fluid level, which characterizes a synovial sarcoma.

Index terms Chest Wall; Computed Tomography, X-Ray; Magnetic Resonance Imaging; Synovial Sarcoma

INTRODUCTION

Synovial sarcoma, reported for the first time in 1893, is the third most common sarcoma that accounts for 10% of all soft tissue sarcomas (1). It usually occurs in adolescents and young adults (15–40 years of age), and while it may be misdiagnosed to be benign owing to its slow growth and asymptomatic nature in the early stages, it is actually malignant and has a poor prognosis (2). Synovial sarcomas may occur anywhere near a joint, tendon, or bursa, though approximately 80% of them are commonly found in the lower extremities, especially near the knees (2). The occurrence of these sarcomas in the thorax is extremely rare, with just a few reported cases in the lungs, mediastinum, pleura, and chest wall (3). To the best of our knowledge, there are only 13 reported cases of synovial sarcoma of the chest wall (4-6). Here, we report a 26-year-old woman who was admitted to our hospital with a chief complaint of pain in the left chest wall. Our report focuses on the diagnosis using CT and MRI findings.

Received June 25, 2019 Revised October 10, 2019 Accepted November 25, 2019

*Corresponding author
Seung Eun Lee, MD
Department of Radiology,
Yeungnam University
College of Medicine,
170 Hyeonchung-ro, Nam-gu,
Daegu 42415, Korea.

Tel 82-53-620-4129 Fax 82-53-653-5484 E-mail air1986@naver.com

This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (https://creativecommons.org/licenses/by-nc/4.0) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

ORCID iDs

Mingook Kim (1)
https://
orcid.org/0000-0001-7165-3744
Seung Eun Lee (1)
https://
orcid.org/0000-0001-6693-4752
Joon Hyuk Choi (1)
https://
orcid.org/0000-0002-8638-0360

CASE REPORT

A 26-year-old woman was admitted to our hospital with the chief complaint of pain in the left chest wall, which she had been experiencing since the past 6 months. A CT scan performed at another hospital 1 week earlier showed a well-defined oval-shaped mass, approximately $2.3\,\mathrm{cm}\times1.8\,\mathrm{cm}\times1.0\,\mathrm{cm}$ in size, below the left sixth costal cartilage. The mass was located in the subpleural area and pushing the pleura inward. On pre-contrast CT scan, the mass showed attenuation similar to that of the chest wall muscles with small internal calcification. On contrast-enhanced CT scan, the mass showed heterogeneous enhancement. The CT values of the mass were approximately 45 Hounsfield unit (HU), 95 HU on pre-contrast and enhanced phase, respectively (Fig. 1A).

After admission, the patient suddenly developed severe pain in the chest wall. Additional dynamic CT and MRI scan was performed. Dynamic CT images show active bleeding in the mass (Fig. 1B). MR images revealed a multilobulated soft-tissue mass with fluid-fluid level on T2-weighted images. The mass showed intermediate signal intensity similar to that of the chest wall muscles with small foci of high signal intensity indicating the presence of hemorrhage on T1-weighted images. A significant amount of hemothorax in the left thoracic cavity was newly demonstrated (Fig. 1C).

Based on the belief that the mass had ruptured and the possibility of malignancy, an emergency surgical excision was performed for histological confirmation and radical treatment. Surgical findings revealed a reddish mass attached to the costal cartilage. There was bleeding inside the mass, and a significant amount of fresh blood was seen in the left pleural space. The mass was excised with enough safety margin, together with the costal cartilage and rib.

On gross examination, the excised mass measured approximately $3.0\,\mathrm{cm}\times2.0\,\mathrm{cm}\times1.3\,\mathrm{cm}$ in size. On section, about $0.7\,\mathrm{cm}\times0.7\,\mathrm{cm}$ sized whitish lesion is located at center of the mass with surrounding hemorrhage (Fig. 1D). Histologically, the mass was determined to be a monophasic synovial sarcoma of the spindle-cell type. The sarcoma differentiation was French Federation of Cancer Center grade 3 (poorly differentiated tumors and undefined histologic types), while the mitotic rate was 20 per 10 high power field. Immunohistochemical staining revealed that the cells in the mass were positive for transducin-like enhancer of split 1 (TLE-1), epithelial membrane antigen (EMA), and cytokeratin (AE1/AE3), thereby confirming the diagnosis of synovial sarcoma (Fig. 1E, F).

DISCUSSION

Synovial sarcoma refers to a mesenchymal neoplasm which occurs in the joints, bursae, and tendon sheaths (7, 8). However, unlike what the name suggests, it does not arise in the synovium. Instead, it is known to grow like the synovial tissues by *de novo* differentiation of multipotent stem cells near the joints (7, 8). Consequently, most cases of synovial sarcoma occur commonly in the thighs, knees, ankles, feet, and upper extremities, but in rare cases, as in this one, it may also occur in the chest wall (9).

Image findings of synovial sarcoma arising in the chest wall is similar with those of synovial sarcoma in the extremities. On CT images, synovial sarcoma arising in the chest wall appears

as a well-defined, heterogeneously enhancing mass containing areas of fluid attenuation, compatible with necrosis and hemorrhage). Ipsilateral pleural effusion is often present and cortical bone destruction, tumor calcifications, and infiltration of the chest wall muscles can be also accompanied (2, 4, 5, 7).

Fig. 1. Radiological and histological findings of a synovial sarcoma of the anterior chest wall in a 26-year-old woman.

A. Non-contrast enhanced CT image (left) shows a well-defined, oval-shaped heterogeneous mass with internal calcification (arrow) in the subpleural area of the left anterior chest wall. Contrast-enhanced CT image (right) shows a well-defined oval-shaped mass measuring 2.3 cm \times 1.8 cm \times 1.0 cm with heterogeneous enhancement.

B. Dynamic CT images obtained after sudden onset of severe chest pain show active bleeding (arrow) within the subpleural mass.

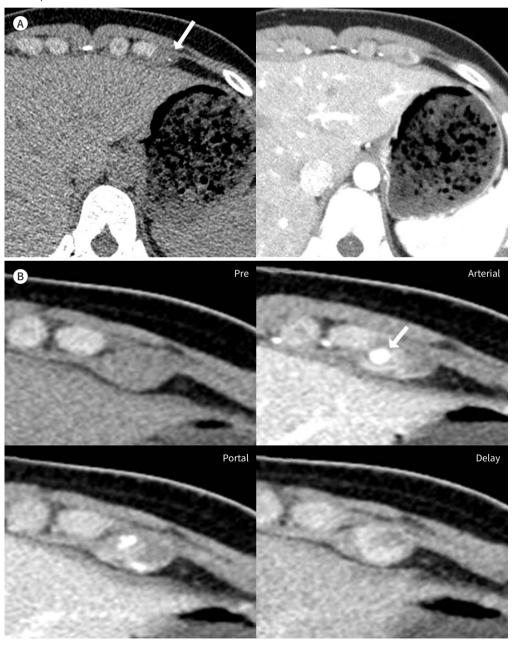
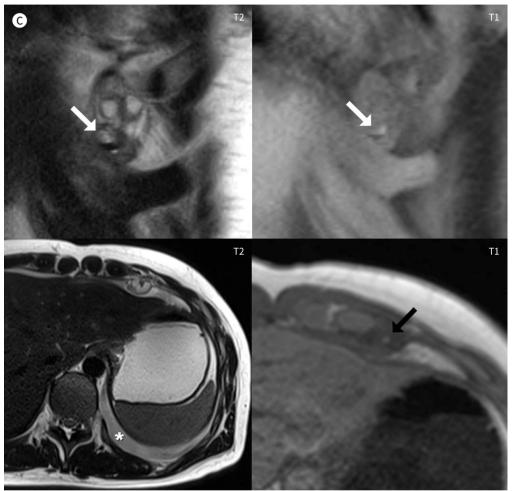


Fig. 1. Radiological and histological findings of a synovial sarcoma of the anterior chest wall in a 26-year-old

C. MR images show a heterogeneous multilobulated mass with a fluid-fluid level (white arrows) and newly developed left-sided hemothorax (asterisk). A small focus of high signal intensity indicating hemorrhage is observed on a T1-weighted image (black arrow).



On MRI, synovial sarcoma arising in the chest wall usually appears as a heterogeneous, multilobulated mass with internal septation and fluid-fluid level (7, 9, 10). It usually demonstrates heterogeneity on both T1- and T2-weighted images. As a result of internal hemorrhage and sedimentary hematoma, the formation of round areas with fluid-fluid level have been reported that are referred to as bowl-of-fruit (7, 10). They usually demonstrate heterogeneous enhancement after the administration of the intravenous contrast material (7). Consistent with previous reports, in our case, a well-defined mass with heterogeneous enhancement and internal calcification was seen on CT images. Internal hemorrhage and ipsilateral hemothorax was accompanied. However, there was no aggressive features such as bone destruction or infiltration of the chest wall muscles. MR images demonstrated a multilobulated mass with fluid-fluid level which is characteristic findings of synovial sarcoma. Our first radiologic diagnosis was benign tumor arising in the chest wall such as peripheral nerve sheath tumor or solitary fibrous tumor based on the image features of small sized, well-defined tumor without aggressive features. However, sudden onset pain with active bleeding and MRI features of multilob-

1230 jksronline.org

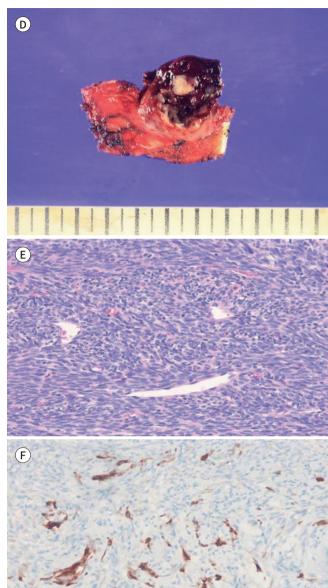


Fig. 1. Radiological and histological findings of a synovial sarcoma of the anterior chest wall in a 26-year-old woman.

- D. On gross appearance, the mass is observed on the rib surface and measures 3.0 cm \times 2.0 cm \times 1.3 cm in size. The section shows a whitish lesion measuring 0.7 cm \times 0.7 cm located at the center of the mass.
- E. Microscopic examination shows spindle-shaped tumor cells with hyperchromatic and monotonous nuclei that are lightly packed together (hematoxylin and eosin stain, × 200). F. Tumor cells showing positive nuclear staining for cytokeratin (immunohistochemical stain, × 200).

ulated mass with fluid-fluid level suggest the possibility of malignancy such as synovial sarcoma and undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma). But there is no reliable imaging findings that help distinguish between these unusual tumors (7).

Histologically, synovial sarcoma can be classified as monophasic, biphasic, or poorly differentiated, consisting of epithelial and/or spindle cell components (2). Among these, the monophasic synovial sarcoma is the most common, accounting for 50–60% of all cases, while spindle cell components predominate (8).

Synovial sarcoma in the chest wall is treated by combination therapy, which includes wide excision, radiotherapy, and adjuvant chemotherapy (7). Because recurrences are common even after treatment, follow-up monitoring is essential.



In summary, we report a case of synovial sarcoma arising in the anterior chest wall in a young woman who complained of long-term pain in the chest wall. CT images demonstrated, a well-defined, heterogeneously enhancing mass with calcification in the subpleural area, which was initially diagnosed as a benign tumor. However sudden onset severe pain with spontaneous bleeding and hemothorax suggest the possibility of malignancy. MR images demonstrated multilobulated appearance with fluid-fluid level which is characteristic findings of synovial sarcoma.

Author Contributions

Conceptualization, K.M., L.S.E.; data curation, K.M., L.S.E.; investigation, K.M.; project administration, L.S.E.; supervision, L.S.E.; visualization, all authors; writing—original draft, K.M., C.J.H.; and writing—review & editing, L.S.E., C.J.H.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

REFERENCES

- 1. Bakri A, Shinagare AB, Krajewski KM, Howard SA, Jagannathan JP, Hornick JL, et al. Synovial sarcoma: imaging features of common and uncommon primary sites, metastatic patterns, and treatment response. AJR Am J Roentgenol 2012;199:W208-W215
- Kawano D, Yoshino I, Shoji F, Morodomi Y, Yano T, Maehara Y. Synovial sarcoma of the chest wall. Gen Thorac Cardiovasc Surg 2010;58:95-97
- **3.** Duran-Mendicuti A, Costello P, Vargas SO. Primary synovial sarcoma of the chest: radiographic and clinicopathologic correlation. *J Thorac Imaging* 2003;18:87-93
- **4.** Braham E, Aloui S, Aouadi S, Drira I, Kilani T, El Mezni F. Synovial sarcoma of the chest wall: a case report and literature review. *Ann Transl Med* 2013;1:9
- Amer HZM, Ren R, Shen R. Biphasic chest wall synovial sarcoma with epithelial pleural effusion: a diagnostic challenge. J Am Soc Cytopathol 2019;8:293-298
- 6. Sudhakar W, Rao Venkata B, Srinivas A, Harshavarsan Majety SR, Madhulika M. Huge synovial sarcoma arising from chest wall: a rare case report. J Evid Based Med Healthc 2015;2:597-601
- 7. Frazier AA, Franks TJ, Pugatch RD, Galvin JR. From the archives of the AFIP: pleuropulmonary synovial sarcoma. *Radiographics* 2006;26:923-940
- 8. Murphey MD, Gibson MS, Jennings BT, Crespo-Rodríguez AM, Fanburg-Smith J, Gajewski DA. From the archives of the AFIP: imaging of synovial sarcoma with radiologic-pathologic correlation. *Radiographics* 2006;26:1543-1565
- Morton MJ, Berquist TH, McLeod RA, Unni KK, Sim FH. MR imaging of synovial sarcoma. AJR Am J Roentgenol 1991;156:337-340
- **10.** Fujimoto K, Hashimoto S, Abe T, Meno S, Terasaki H, Nishimura H, et al. Synovial sarcoma arising from the chest wall: MR imaging findings. *Radiat Med* 1997;15:411-414

1232 jksronline.org

앞가슴벽에 생긴 윤활막육종: 증례 보고

김민국1·이승은1*·최준혁2

윤활막육종은 주로 사지, 특히 무릎 근처에 발생하는 악성 연부조직 종양으로, 가슴벽에 발생한 경우는 매우 드물다. 저자들은 26세 여성에서 앞가슴벽에 발생한 윤활막육종의 증례를 CT 및 MRI 소견을 중심으로 보고하고자 한다. CT에서 흉막하에 위치한, 경계가 좋으며 석회화를 동반한 작은 종괴로 관찰되어, 처음에는 양성 종양으로 생각하였다. 하지만 갑자기 심한 통증과 함께 종괴의 출혈과 혈흉이 동반되어 악성의 가능성을 고려하였다. MRI에서는 다엽성종괴 내부에 액체액체층이 보였고, 이는 윤활막육종의 특징적인 영상 소견이다.

영남대학교 의과대학 1영상의학교실, 2병리학교실