Case Report

J Korean Soc Radiol 2022;83(6):1366-1372
https://doi.org/10.3348/jksr.2022.0005
eISSN 2951-0805

JOURNAL of THE KOREAN SOCIETY of RADIOLOGY

MRI Findings in a Rare Case of Myxofibrosarcoma in the Anterior Mediastinum

전종격동에 발생한 매우 드문 점액섬유육종의 자기공명영상 소견

Mi Jin Kim, MD, Se Ri Kang, MD*, Ji Young Rho, MD

Department of Radiology, Wonkwang University Hospital, Wonkwang University School of Medicine, Iksan, Korea

ORCID iDs
Mi Jin Kim https://orcid.org/0000-0001-5568-0187
Se Ri Kang https://orcid.org/0000-0001-8684-3778
Ji Young Rho https://orcid.org/0000-0001-6645-3983

Received January 10, 2022
Revised February 22, 2022
Accepted June 1, 2022

***Corresponding author**
Se Ri Kang, MD
Department of Radiology,
Wonkwang University Hospital,
Wonkwang University
School of Medicine,
895 Muwang-ro, Iksan 54538,
Korea.

Tel 82-63-859-1920
Fax 82-63-851-4749
E-mail kangseli21@naver.com

Myxofibrosarcoma is one of the most common soft tissue sarcomas in elderly patients. It often occurs in the extremities, trunk, and retroperitoneum. However, it is rarely observed in the mediastinum, and only a few cases have been reported in the literature. Herein, we present the imaging findings, with an emphasis on the MRI results, of a surgically confirmed anterior mediastinal myxofibrosarcoma in a 66-year-old male.

Index terms Computed Tomography, X-Ray; Fibrosarcoma; Magnetic Resonance Imaging; Mediastinum

INTRODUCTION

Myxofibrosarcoma, a malignant fibroblastic lesion with variably myxoid stroma and a distinctively curvilinear vascular pattern, is one of the most common soft tissue sarcomas in elderly patients (1). It is most commonly found in the extremities, followed by the trunk, retroperitoneum, and head and neck (1-3). However, it rarely occurs in the mediastinum. To the best of our knowledge, there are only six cases of mediastinal myxofibrosarcoma reported in the literature (4-6). Of these, only three cases described MRI findings, and no diffusion-weighted imaging (DWI) with MRI was performed. Herein, we present the CT and MRI findings, including DWI, of a surgically confirmed anterior mediastinal myxofibrosarcoma in a 66-year-old male.

CASE REPORT

A 66-year-old male visited our emergency department complaining of chest discomfort and dyspnea for two months. On physical examination, his breath sounds were clear, and his heartbeat was regular, without any murmur. Electrocardiography and hematological examination showed nonspecific findings. Chest radiography showed a markedly enlarged cardiac silhouette with mediastinal widening (Fig. 1A). A non-contrast-enhanced axial chest CT (Somatom Definition Edge, Siemens Healthcare, Erlangen, Germany) showed a heterogeneously low attenuated (18–24 Hounsfield unit [HU]) anterior mediastinal mass measuring approximately 20 cm × 12 cm × 15 cm. Contrast-enhanced axial chest CT showed heterogeneous enhancement (28–36 HU) with focally well-enhanced areas (Fig. 1B). The mass encompassed the heart and the great vessels and extended to the pericardium with a small right sided pericardial effusion (Fig. 1B). In addition, multiple enlarged mediastinal, bilateral hilar, and left internal mammary lymph nodes were noted. Based on these findings, malignant anterior mediastinal tumor such as thymic carcinoma or malignant mesenchymal tumor was suspected.

To further evaluate this mediastinal mass, a chest MRI was performed using a 3.0 Tesla scanner (Ingenia, Philips Healthcare, Best, Netherlands). Black-blood T2-weighted image with and without fat saturation, black-blood T1-weighted image, enhanced T1-weighted image with fat saturation, and DWI were performed. DWI was conducted using b values of 0, 100, and 1000 s/mm^2. The tumor showed heterogeneous low to intermediate signal intensity on black-blood T1-weighted image and heterogeneous low to high signal intensity on black-blood T2-weighted image (Fig. 1C), and heterogeneous enhancement with areas of focal nodular and patchy intense enhancement (Fig. 1D). There were no definite fat components within the mass. Most of the tumor showed heterogeneous high signal intensity on DWI and intermediate to high signal intensity on the apparent diffusion coefficient (ADC) map (Fig. 1E) with corresponding poor enhancement (Fig. 1D). However, focal nodular and patchy areas of the tumor showed very high signal intensity on DWI and very low signal intensity on the ADC map (Fig. 1E) with corresponding intense enhancement (Fig. 1D). Combining these findings, the mass was thought to be a soft tissue tumor containing many myxoid and hypercellular solid components.

CT-guided percutaneous transthoracic biopsy revealed a pathologic result of a spindle cell tumor with low cellularity. Mass excision with sternotomy was performed. Only the lower half of the tumor was resected because the mass invaded the innominate vein and main pulmonary artery. Grossly, a myxoid component and hemorrhage were observed on the resected tumor. Hematoxylin and eosin staining showed spindle cells with atypia in the myxoid and fibrous background (Fig. 1F). Immunohistochemistry showed positive results for CD99 and negative results for Bcl-2, CD34, and S-100 protein. The Ki-67 index was 30%. Tumor necrosis was not observed. Based on these histopathological findings, the tumor was confirmed to be myxofibrosarcoma, Fédération Nationale des Centers de Lutte Contre le Cancer (FNCLCC) grade 2 (intermediate).

The patient underwent adjuvant chemotherapy (67.8 mg doxorubicin + 1357 mg ifosfamide) following surgical resection. After three months, a follow-up chest CT revealed that the size of the residual mass had increased. The patient visited the last outpatient clinic

JOURNAL of
THE KOREAN SOCIETY of
RADIOLOGY

Fig. 1. A 66-year-old male with anterior mediastinal myxofibrosarcoma.
A. Chest radiography shows a markedly enlarged cardiac silhouette with mediastinal widening.
B. Contrast-enhanced axial chest CT (left) shows a huge heterogeneously enhancing anterior mediastinal mass with focal, enhanced area, and contrast-enhanced coronal chest CT (right) shows that the mass encompassed the heart and great vessels, with minimal right-sided pericardial effusion (arrows).
C. The mass shows low to intermediate signal intensity on black-blood T1WI (left) and low to high signal intensity on black-blood T2WI with fat saturation (right), heterogeneously, and minimal pericardial effusion (arrow) with abnormal signal void due to cardiac motion is also noted.
T1WI = T1-weighted image, T2WI = T2-weighted image

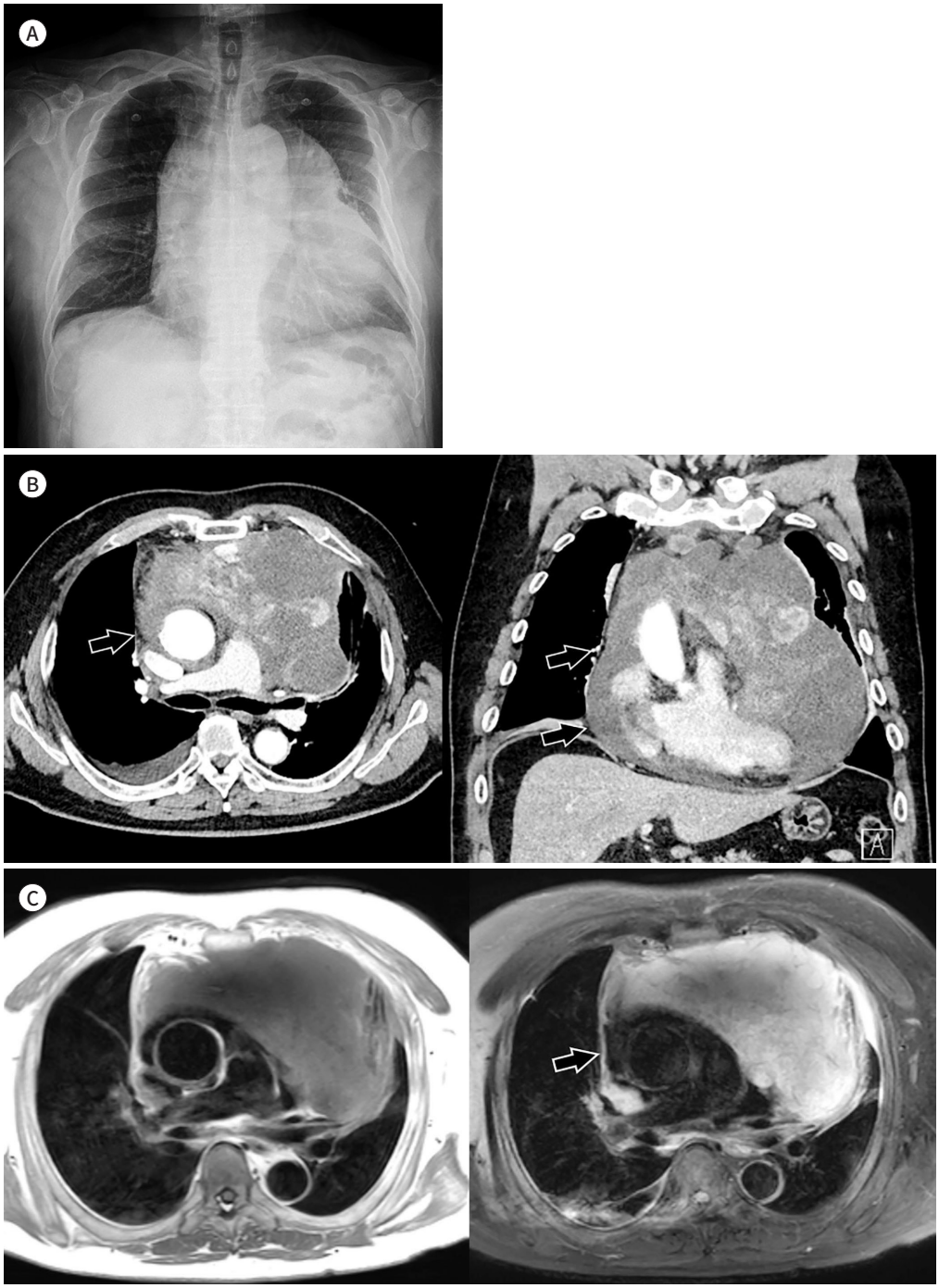
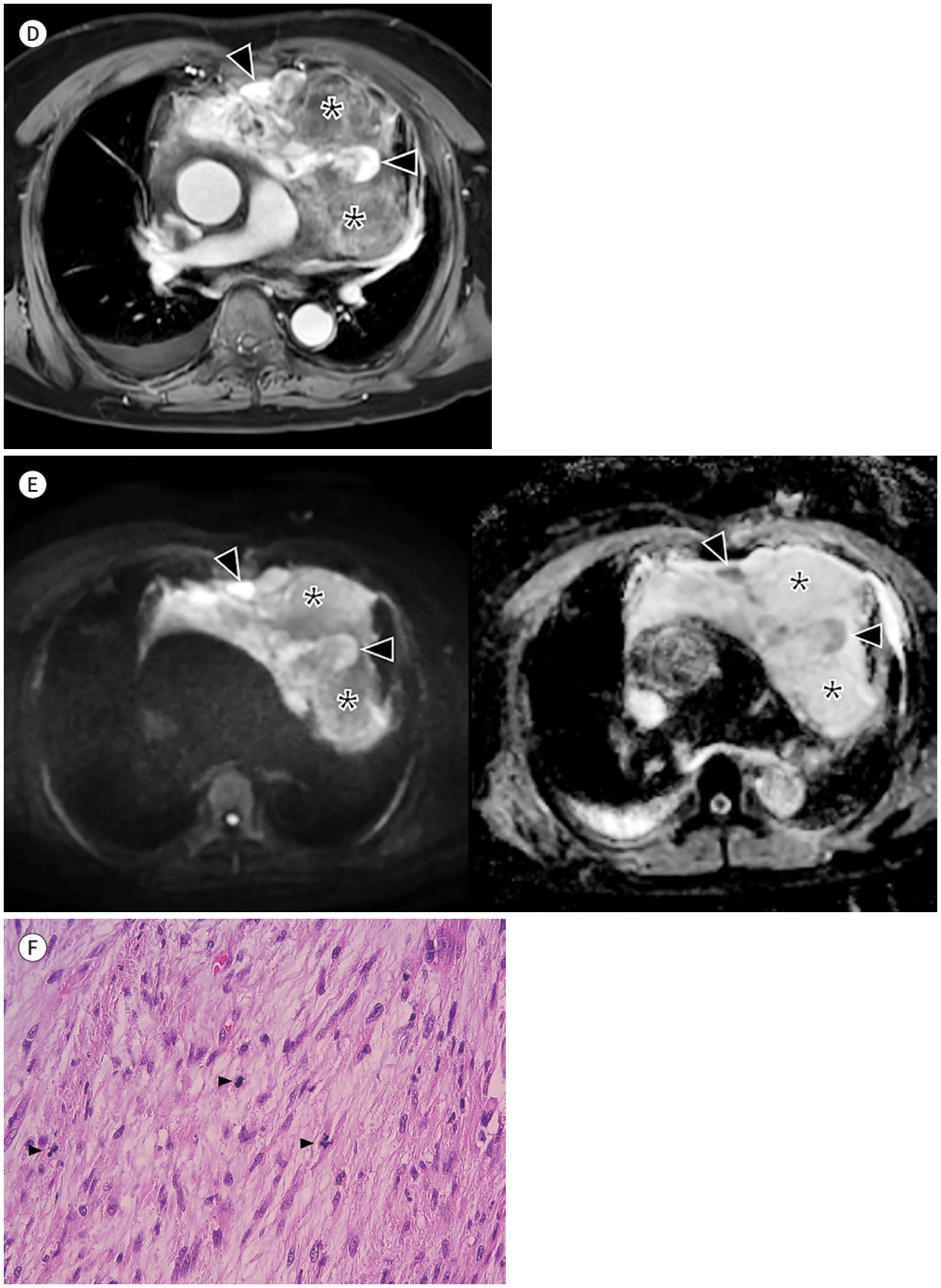


Fig. 1. A 66-year-old male with anterior mediastinal myxofibrosarcoma.
D. Contrast-enhanced T1WI with fat saturation shows heterogeneous enhancement (asterisks) with focal nodular and patchy intense enhancing area (arrowheads).
E. Most of the tumors with heterogeneously poor enhancement on enhanced T1WI (asterisks) shows heterogeneous high signal intensity on DWI and intermediate to high signal intensity on the ADC map, and focal nodular and patchy areas of the tumor with intense enhancement on enhanced T1WI (arrowheads) shows very high signal intensity on DWI and very low signal intensity on the ADC map.
F. Histopathological examination shows spindle cells with atypia in myxoid and fibrous background (arrowheads) (hematoxylin & eosin stain, × 400).
ADC = apparent diffusion coefficient, DWI = diffusion-weighted imaging, T1WI = T1-weighted image, T2WI = T2-weighted image



JOURNAL of THE KOREAN SOCIETY of RADIOLOGY

about 6 months after surgery and did not visit the hospital after that.

This case report was approved by our hospital's Institutional Review Board, and the requirement for informed consent was waived (IRB No. WKUH 2021-12-044).

DISCUSSION

Myxofibrosarcoma is one of the most common soft tissue neoplasms in elderly patients (1, 2). Myxofibrosarcoma was formerly classified as the myxoid variant of malignant fibrous histiocytoma, but it is has been newly classified as a distinct pathologic entity from malignant fibrous histiocytoma according to the 2002 World Health Organization (WHO) criteria, which emphasizes its fibroblastic nature and abundant myxoid stroma (1, 3). Myxofibrosarcoma typically arises on the extremities, particularly the lower extremities. Approximately two-thirds of cases develop in dermal or subcutaneous tissues, while the remainder occurs in the underlying fascia and skeletal muscle (1-3). Myxofibrosarcoma originating from the mediastinum is extremely rare, and only six cases have been reported in the literature (4-6). Clinicoradiologic features of the seven reported cases, including the current case report, are exhibited in Supplementary Table 1 (in the online-only Data Supplement). The age ranged from to 38–81 years (median age, 70 years). The tumors were located in the anterior mediastinum in five cases, and only one case each was located in the middle and posterior mediastinum.

Histologically, myxofibrosarcoma demonstrates distinct morphological features of multinodular growth with incomplete fibrous septa and a myxoid stroma consisting of hyaluronic acid (1). It varies from a low-grade hypocellular, mainly myxoid lesion with purely spindle cell appearance to a high-grade hypercellular pleomorphic lesion (1). Histologic features of the high water content of the myxoid component are reflected in imaging findings of various modalities, including hypoechogenicity on ultrasound, low attenuation on CT, very high signal intensity on T2-weighted image, and low to intermediate signal intensity on T1-weighted image, which is similar to that of fluid (3, 7). However, unlike cysts, nodular or diffuse enhancement is often seen in both the solid and more vascularized myxoid component of myxofibrosarcoma (3, 7, 8). These findings are concordant with the imaging findings of the patient in the current case and three previous cases (4-6) that describe the MRI findings of mediastinal myxofibrosarcoma.

In addition, we obtained DWI with MRI for tissue characterization. To our knowledge, this is the first report of DWI findings in mediastinal myxofibrosarcoma. Nowadays, DWI has become a useful tool for discriminating tumor characteristics (9, 10). DWI reflects variation in the Brownian motion of water caused by changes in the tissue microstructure, and the ADC is a quantitative measure of Brownian motion. Low ADC values indicate a highly cellular microenvironment where diffusion is restricted, whereas high ADC values reflect hypocellular regions that permit free diffusion of water (9, 10). However, ADC values might be affected not only by cellularity but also by the nature of the extracellular matrix (9, 10). Myxoid-containing soft tissue tumors have significantly higher ADC values than non-myxoid soft tissue tumors (9, 10). In addition, in myxoid tumors, ADC values between benign and malignant tumors overlap significantly, limiting the differential diagnosis (10). In the current case, most of the tumor showed high ADC values with heterogeneous poor enhancement in the corre-

sponding area, which suggested a hypocellular myxoid component. On the other hand, focal nodular and patchy areas of the tumor showed very low ADC values with intense enhancement, which reflected a hypercellular solid component.

Myxofibrosarcoma can be graded from grade 1 to grade 3 under the FNCLCC system (3). Hypocellular cases tend to have low mitotic activity and no necrosis and are thus considered low-grade. On the other hand, hypercellular cases tend to have high mitotic activity and necrotic and hemorrhagic foci and are thus considered high-grade (3). The current case revealed an intermediate grade (grade 2) myxofibrosarcoma.

In summary, we describe a rare case of myxofibrosarcoma arising from the anterior mediastinum. DWI can be a useful sequence for tumor characterization when chest MRI is performed to evaluate mediastinal tumors. In addition, when an aggressive anterior mediastinal mass reveals a myxoid component on chest MRI, radiologists could suggest the possibility of myxofibrosarcoma.

Supplementary Materials

The online-only Data Supplement is available with this article at http://doi.org/10.3348/jksr.2022.0005.

Author Contributions

Conceptualization, K.S.R.; investigation, K.M.J.; supervision, K.S.R.; validation, K.M.J.; visualization, K.M.J.; writing—original draft, K.M.J.; and writing—review & editing, K.S.R., R.J.Y.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Funding

This study was supported by Wonkwang University in 2021.

REFERENCES

1. Mentzel T, Van den Berg E, Molenaar WM. *Myxofibrosarcoma*. In Fletcher CDM, Unni KK, Mertens F, eds. *World Health Organization classification of tumours. Pathology and genetics of tumours of soft tissue and bone*. Lyon: IARC Press 2002:102-103
2. Mentzel T, Calonje E, Wadden C, Camplejohn RS, Beham A, Smith MA, et al. Myxofibrosarcoma. Clinicopathologic analysis of 75 cases with emphasis on the low-grade variant. *Am J Surg Pathol* 1996;20:391-405
3. Roland CL, Wang WL, Lazar AJ, Torres KE. Myxofibrosarcoma. *Surg Oncol Clin N Am* 2016;25:775-788
4. Sugiura Y, Fujimoto H, Hashizume T, Ogura G, Nemoto E. An entire clinical course of mediastinal myxofibrosarcoma treated by surgery and radiation. *Gen Thorac Cardiovasc Surg* 2018;66:748-752
5. Yoon HE, Shimizu S, Tokura S, Okuma T, Kawaguchi T, Ito I, et al. Primary high-grade myxofibrosarcoma in the anterior mediastinum: a case report and review of the literature. *Cancer Treat Res Commun* 2016;9:44-47
6. Fukui T, Wakatsuki Y, Matsukura T. Immunotherapy for advanced lung cancer combined with surgery for mediastinal myxofibrosarcoma: a case report. *Surg Case Rep* 2019;5:37
7. Petscavage-Thomas JM, Walker EA, Logie CI, Clarke LE, Duryea DM, Murphey MD. Soft-tissue myxomatous lesions: review of salient imaging features with pathologic comparison. *Radiographics* 2014;34:964-980
8. Kransdorf MJ, Murphey MD. *Imaging of soft tissue tumors.* 2nd ed. Philadelphia: Lippincott Williams & Wilkins 2006:320
9. Costa FM, Ferreira EC, Vianna EM. Diffusion-weighted magnetic resonance imaging for the evaluation of musculoskeletal tumors. *Magn Reson Imaging Clin N Am* 2011;19:159-180
10. Maeda M, Matsumine A, Kato H, Kusuzaki K, Maier SE, Uchida A, et al. Soft-tissue tumors evaluated by line-scan diffusion-weighted imaging: influence of myxoid matrix on the apparent diffusion coefficient. *J Magn Reson Imaging* 2007;25:1199-1204

JOURNAL of
THE KOREAN SOCIETY of
RADIOLOGY

전종격동에 발생한 매우 드문 점액섬유육종의 자기공명영상 소견

김미진 · 강세리* · 노지영

점액섬유육종은 고령자에서 가장 흔한 연조직 육종 중 하나이며 사지, 몸통 및 후복막에서 흔히 발생한다. 그러나 종격동 점액섬유육종은 매우 드물며 문헌에서도 몇 가지 사례만이 보고되었다. 이에 저자들은 66세 남성에서 수술로 확진된 전종격동 점액섬유육종의 영상 소견을 MRI 소견 중심으로 보고하고자 한다.

원광대학교 의과대학 원광대학교병원 영상의학과