Case Report

J Korean Soc Radiol 2024;85(1):204-209
https://doi.org/10.3348/jksr.2023.0039
eISSN 2951-0805

JOURNAL of THE KOREAN SOCIETY of RADIOLOGY

CT and MRI Findings of Low-Flow Mediastinal Vascular Malformation: A Case Report

종격동에 발생한 저혈류성 혈관 기형의 영상 소견: 증례 보고

Hanlim Song, MD, Mi Sook Lee, MD, Soo-yeon Jeong, MD*

Department of Radiology, Presbyterian Medical Center, Jeonju, Korea

ORCID iDs
Hanlim Song https://orcid.org/0000-0001-6701-4296
Mi Sook Lee https://orcid.org/0000-0002-1488-2249
Soo-yeon Jeong https://orcid.org/0000-0003-2630-2245

Received April 12, 2023
Revised June 20, 2023
Accepted August 10, 2023

***Corresponding author**
Soo-yeon Jeong, MD
Department of Radiology, Presbyterian Medical Center, 365 Seowon-ro, Wansan-gu, Jeonju 54987, Korea.

Tel 82-32-230-2398
Fax 82-32-230-8387
E-mail jsooyeon87@naver.com

Mediastinal vascular malformations are rare and their diagnosis can be challenging. Imaging is vital for diagnosing mediastinal vascular malformations and can help avoid unnecessary invasive procedures. Herein, we report the detailed CT and MRI findings of a rare low-flow mediastinal vascular malformation in an asymptomatic 63-year-old male.

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

INTRODUCTION

Vascular malformations constitute a major category of vascular anomalies, along with vascular tumors. Vascular malformations can be divided into high- and low-flow types based on their hemodynamic characteristics. Malformations containing arterial components are regarded as the high-flow type. Simultaneously, any single or a combination of the venous, capillary, and lymphatic systems is considered a low-flow type (1). Accurate diagnosis and differentiation between high- and low-flow malformations are critical for appropriate treatment planning (2).

CASE REPORT

An asymptomatic 63-year-old male was referred to our hospital for abnormal chest radiographic findings during a routine healthcare examination. Posteroanterior chest radiography revealed a blunted right cardiophrenic angle with an obscured right cardiac border (not shown).

Contrast-enhanced chest CT revealed widespread soft-tissue density lesions along the mediastinum, diaphragm, and upper abdomen, engulfing adjacent organs (Fig. 1A). The lesion showed interspersed fat and calcified densities (presumed to be phleboliths). No demonstrable invasion of adjacent organs was observed. The lesion exhibited gradual delayed enhancement on multiphase abdominal CT, without feeding arteries (Fig. 1B). On axial (Fig. 1C) and sagittal reformatted (Fig. 1D) imaging, we suspected partial connectivity among the lesions located in each mediastinal compartment, diaphragm, and retroperitoneal space.

On MRI, the lesion demonstrated high signal intensity on fat-saturated T2-weighted images (Fig. 1E, first row) and intermediate signal intensity on T1-weighted images (Fig. 1E, second row). The lesion showed delayed enhancement, similar to the CT scan (Fig. 1E, third row), with no arterial feeders. T2-weighted imaging revealed a large draining vein into the inferior vena cava (IVC) (Fig. 1F). Diffusion restrictions were not observed (data not shown).

Fig. 1. A 63-year-old male with incidentally discovered mediastinal vascular malformation.
A. Chest CT images show widespread soft tissue lesions along the mediastinum, diaphragm, and abdomen. Non-contrast images (first column) show scattered calcifications (phleboliths) and interspersed fat. Post-contrast (acquired in the arterial phase) images (second column) do not show any arterial feeders.
B. Non-contrast, arterial, portal, and delayed phase (in order) abdominal CT images show gradual delayed enhancement.

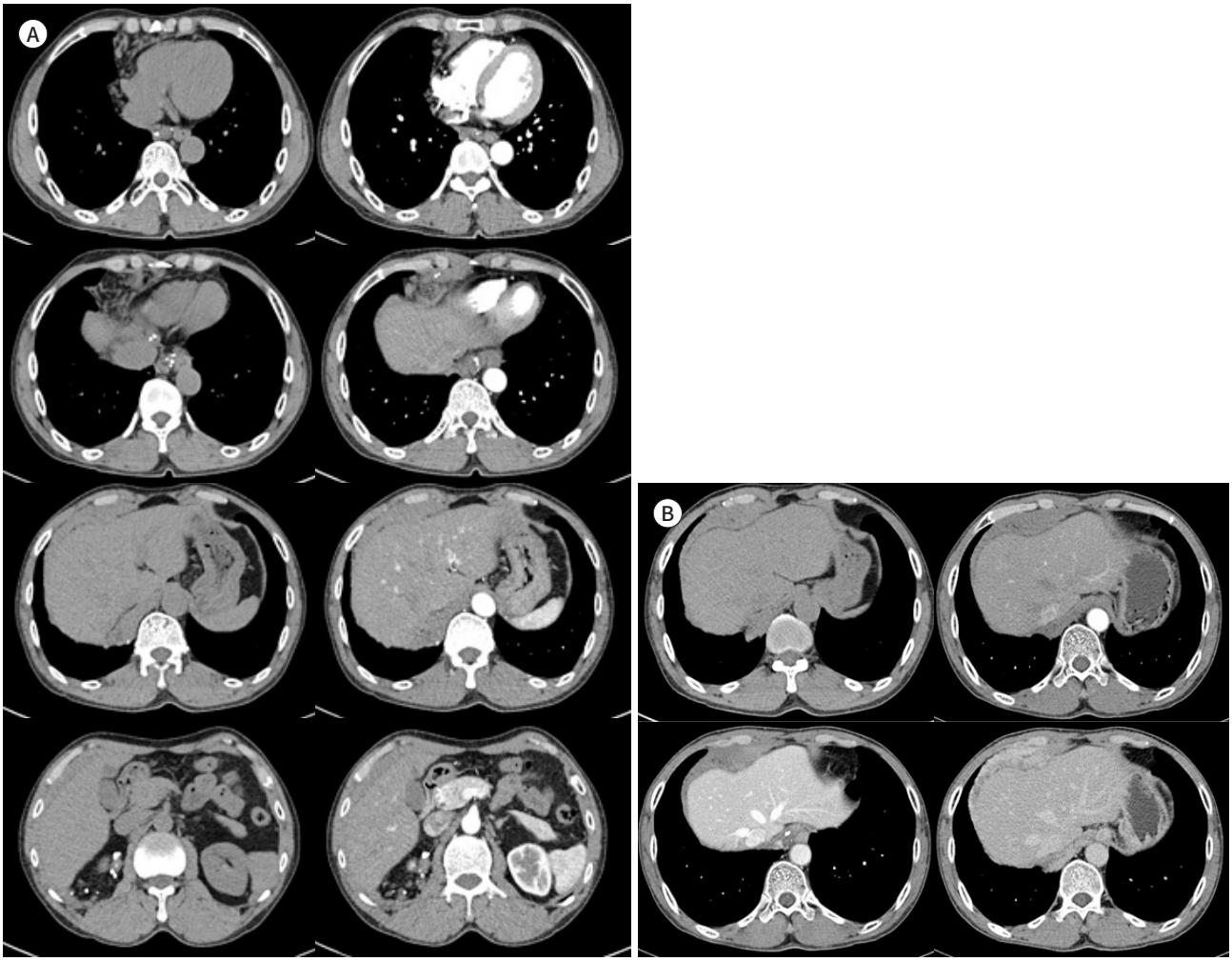
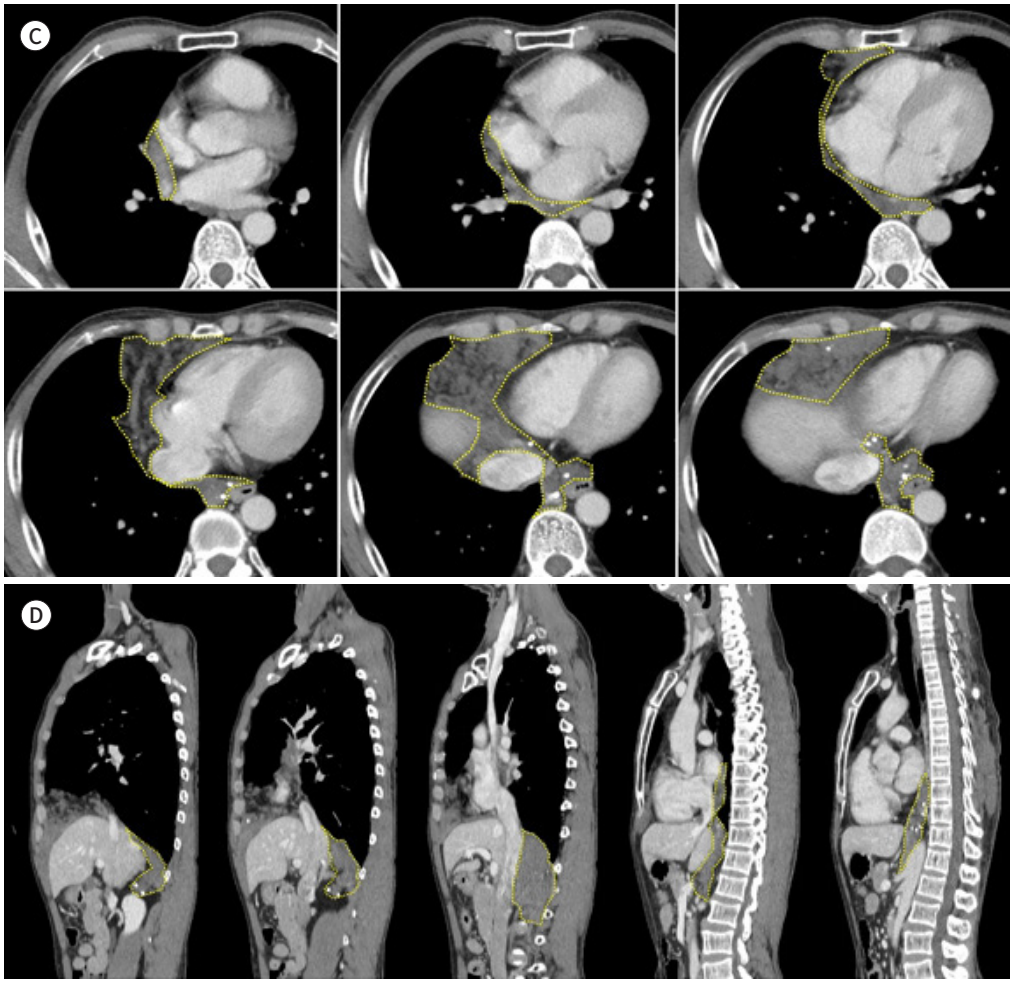


Fig. 1. A 63-year-old male with incidentally discovered mediastinal vascular malformation.
C. The axial chest CT images demonstrate partial continuity of the lesions (delineated by the yellow dotted line) among the mediastinal compartments.
D. The sagittal reformatted images show potential continuity of the lesions (delineated by the yellow dotted line) between the mediastinum, diaphragm and the right retroperitoneal space.



The CT and MRI features were consistent with a vascular malformation, with a large vein draining into the IVC without any arterial feeders, suggesting a low-flow nature of the lesion. Tissue biopsy was not performed because the lesion was a deeply located low-flow vascular malformation. The lesion remained stable for several years on follow-up CT (not shown).

This study was approved by the Institutional Review Board of our institution, which waived the requirement for informed consent (IRB No. E2022-18).

DISCUSSION

Vascular anomalies encompass a broad spectrum of rare vascular lesions, which are classified as vascular tumors and malformations (Table 1) according to the International Society for the Study of Vascular Anomalies (ISSVA) classification, last updated in 2018 (3).

In existing literature, the terms 'vascular tumors', 'vascular malformations', and 'hemangio-

Fig. 1. A 63-year-old male with incidentally discovered mediastinal vascular malformation.
E. On MRI, the lesion shows high signal intensity on fat-saturated T2 weighted images (first row) and intermediate signal intensity on T1 weighted images (second row). Upon contrast enhancement, this shows gradual delayed enhancement in arterial, portal, and delayed phase images (third row, in order).
F. The MR T2 weighted image shows a large draining vein into the inferior vena cava (arrows).

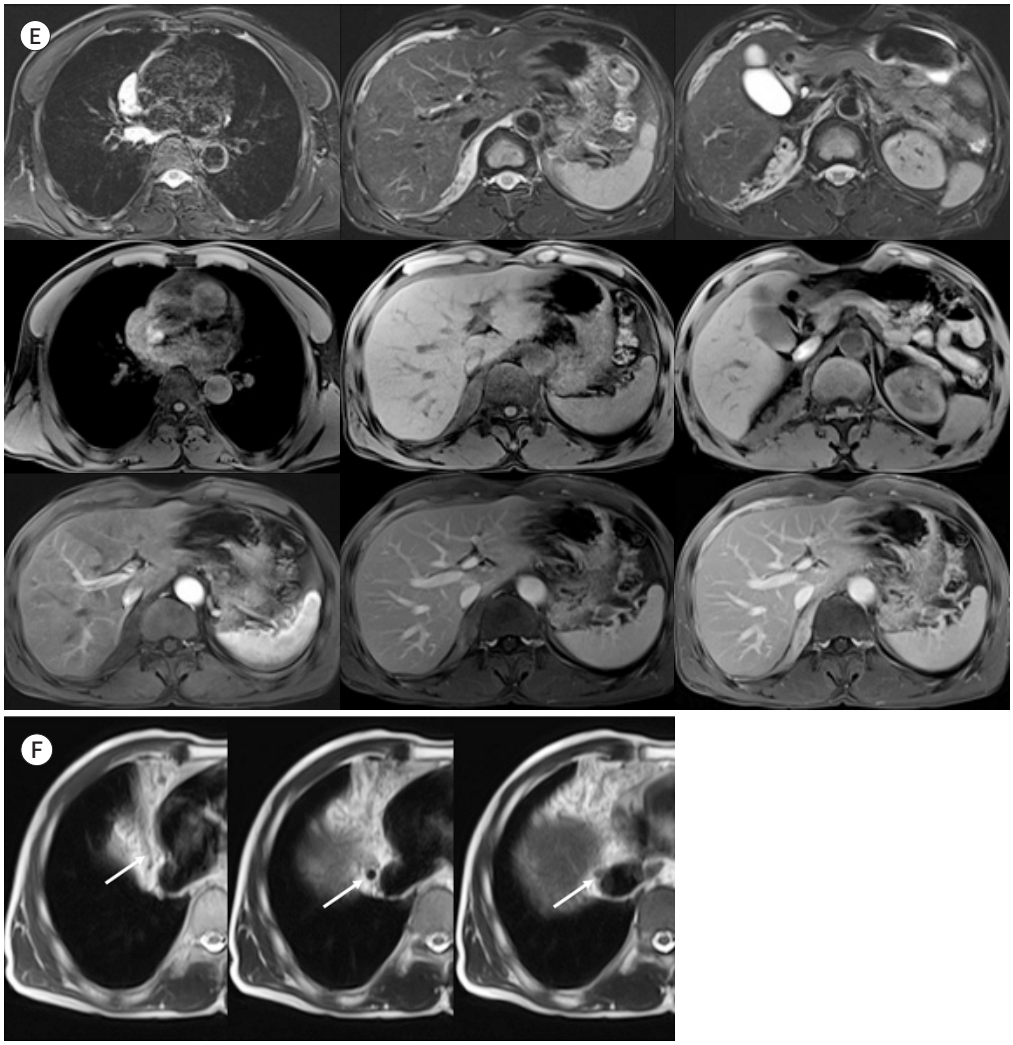


Table 1. International Society for the Study of Vascular Anomalies Classification for Vascular Anomalies

Vascular Tumors	Vascular Malformations
Benign	Simple (CM, LM, VM, AVM*, AVF*)
Locally aggressive or borderline	Combined† (CVM, CLM, LVM, CLVM, CAVM*, CLAVM*, others)
Malignant	Of major named vessels‡
	Associated with other anomalies‡

*High-flow lesions.
†Defined as two or more vascular malformations found in one lesion.
‡Please refer to original sources (3).
AVF = arteriovenous fistula, AVM = arteriovenous malformations, CAVM = capillary-arteriovenous malformation, CLAVM = capillary-lymphatic-arteriovenous malformation, CLM = capillary-lymphatic malformation, CLVM = capillary-lymphatic-venous malformation, CM = capillary malformations, CVM = capillary-venous malformation, LM = lymphatic malformations, LVM = lymphatic-venous malformation, VM = venous malformations

mas' have often been used interchangeably without clear distinction. The primary inquiry for diagnostic physicians will revolve around the distinction between vascular tumors and malformations. Vascular malformations are thought to represent inborn errors of vessel morphogenesis, unlike vascular tumors (true neoplasms) that show clonal cellular proliferation with mitosis (4). Whereas, most vascular tumors have a mass-like appearance (eg. hemangioma, a category of benign vascular tumors), vascular malformations often grow with the patient's body and show infiltrative features that cross multiple tissue planes (1, 2, 4).

They are composed of a single system or a combination of the arterial, capillary, venous, or lymphatic systems. Among these, venous malformations represent the most frequently occurring type of congenital vascular malformation, with an incidence rate ranging from 1 to 2 cases per 10000 individuals and an overall prevalence of approximately 1% (5).

Vascular malformations can be broadly divided into high-flow (containing arterial feeders such as arteriovenous malformations or fistulas) and low-flow types (1, 2). This subclassification is vital for treatment planning and prognostication. Several imaging modalities may play essential roles in this regard.

CT is an excellent imaging modality for assessing the exact anatomical extent and relationship with adjacent structures, providing high spatial resolution and multiplanar reformatted images. Calcifications, phleboliths, and thrombosis, which are the hallmarks of low-flow vascular malformations, can be observed on CT (6).

MRI is an excellent technique for tissue characterization. Typical MRI features of low-flow vascular malformations include T2 hyperintensity and T1 intermediate signal intensity, with phleboliths (when these exist) appearing as dark signal intensity foci on all pulse sequences (6). On contrast enhancement, slow and gradual enhancements are observed.

High-flow vascular malformations can lead to complications such as bleeding, pain, ischemia, or high-output cardiac failure if left untreated and therefore require treatment (7). In contrast, low-flow vascular malformations present treatment challenges. In the absence of symptoms, patients with low-flow vascular malformations can be monitored through regular follow-up and managed conservatively (8). In cases with symptoms, treatment options such as surgical resection or percutaneous sclerotherapy may be considered (6).

Our case demonstrated the characteristic features of a low-flow vascular malformation. To the best of our knowledge, vascular malformations involving the mediastinum are rare, and only a few cases have been reported (6, 9, 10). Radiologists should familiarize themselves with the CT and MRI findings of mediastinal vascular malformations to avoid unnecessary invasive procedures.

Author Contributions

Conceptualization, all authors; supervision, J.S.; writing—original draft, S.H.; and writing—review & editing, J.S.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Funding

None

REFERENCES

1. Steiner JE, Drolet BA. Classification of vascular anomalies: an update. *Semin Intervent Radiol* 2017;34:225-232
2. Sadick M, Müller-Wille R, Wildgruber M, Wohlgemuth WA. Vascular anomalies (part I): classification and diagnostics of vascular anomalies. *Rofo* 2018;190:825-835
3. ISSVA. ISSVA classification of vascular anomalies ©2018 International Society for the Study of Vascular Anomalies. Available at. https://www.issva.org/classification. Accessed June 11, 2023
4. Merrow AC, Gupta A, Patel MN, Adams DM. 2014 revised classification of vascular lesions from the international society for the study of vascular anomalies: radiologic-pathologic update. *Radiographics* 2016;36:1494-1516
5. Behravesh S, Yakes W, Gupta N, Naidu S, Chong BW, Khademhosseini A, et al. Venous malformations: clinical diagnosis and treatment. *Cardiovasc Diagn Ther* 2016;6:557-569
6. Robert A, Raymond D, Bolen M, Renapurkar R. Mediastinal venous vascular malformations: report of two cases, with discussion of imaging findings and classification systems. *Clin Imaging* 2014;38:218-220
7. Rosen RJ, Nassiri N, Drury JE. Interventional management of high-flow vascular malformations. *Tech Vasc Interv Radiol* 2013;16:22-38
8. Rajiah P, Kanne JP. Mediastinal vascular malformation presenting with stroke. *Br J Radiol* 2010;83:e138-e142
9. Munechika M, Tobino K, Okahisa M, Gotou Y, Murakami K, Sueyasu T, et al. A case of huge lymphatic and venous malformations of the mediastinum. *Respir Med Case Rep* 2019;26:53-55
10. Kadota Y, Utsumi T, Kawamura T, Inoue M, Sawabata N, Minami M, et al. Lymphatic and venous malformation or "lymphangiohemangioma" of the anterior mediastinum: case report and literature review. *Gen Thorac Cardiovasc Surg* 2011;59:575-578

종격동에 발생한 저혈류성 혈관 기형의 영상 소견: 증례 보고

송한림 · 이미숙 · 정수연*

종격동 혈관 기형은 매우 드물며 진단이 어려울 수 있다. 영상은 이러한 종격동 혈관 기형의 진단에 있어 중요한 역할을 하며 불필요한 침습적 시술을 피하는 데 도움을 줄 수 있다. 이에 저자들은 63세 무증상 남성에서 발생한 저혈류성 종격동 혈관기형의 자세한 전산화단층촬영 및 자기공명영상 소견을 보고하고자 한다.

전주예수병원 영상의학과