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# Case Report of a Mediastinal Vascular **Malformation Mimicking Esophageal Varices** on Endoscopy, with Emphasis on Radiological **Findings**

Authors' Contribution: Study Design A Data Collection B Statistical Analysis C Data Interpretation D Manuscript Preparation E Literature Search F Funds Collection G

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None declared

**Patient:** 

Male, 48-year-old

**Final Diagnosis:** 

Mediastinal vascular malformation

**Symptoms:** 

Cough • fever

**Medication:** 

**Clinical Procedure:** Specialty:

Radiology

Objective: Background: Mistake in diagnosis

**Case Report:** 

symptoms. Imaging can be challenging to interpret, but plays an important role in diagnosis and prognostication. We present the case of a 48-year-old man with history of intravenous drug abuse and incompletely treated pulmonary tuberculosis. A computed tomography (CT) scan done for respiratory symptoms showed an extensive soft-tissue mass in the mediastinum and upper abdomen, initially thought to represent tuberculous adenitis with possible esophageal involvement, which appeared variceal in nature on endoscopy. Further investigation with open mediastinal biopsy and magnetic resonance imaging (MRI) eventually led to the diagnosis of a low-flow venous mediastinal vascular malformation. The patient responded well to conservative manage-

Mediastinal vascular malformations are rare, and most patients are asymptomatic or present with unrelated

ment, with the malformation remaining stable on follow-up CT up to a decade later.

**Conclusions:** 

Radiologists should be aware of the rare but important differential diagnosis of a vascular malformation, particularly when an extensive infiltrative calcified mediastinal soft-tissue mass is encountered. Multi-modality imaging, particularly MRI, which can demonstrate typical features, is crucial for diagnosis and prognostication, thereby avoiding unnecessary invasive procedures and treatment.

MeSH Keywords:

Esophageal and Gastric Varices • Mediastinum • Radiology • Tuberculosis • Vascular Malformations

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## **Background**

Mediastinal vascular malformations are rare, with only isolated case reports in the existing literature. Most patients are asymptomatic or present with unrelated symptoms. We present a case of an extensive mediastinal vascular malformation with challenging clinical features, with particular focus on the role of radiology and the radiological findings necessary for image-based diagnosis. We also outline the classification and general treatment principles for vascular malformations.

### **Case Report**

A 48-year-old man presented to our hospital's Emergency Department with fever, cough, and epigastric pain of 3 days' duration. He was known to have longstanding alcoholism, a history of intravenous drug abuse, and incompletely treated pulmonary tuberculosis (TB) more than 20 years ago. Chest radiograph (Figure 1) showed left upper-zone fibrocalcific changes consistent with prior granulomatous infection, and patchy increased opacification over the left middle to lower zones. His inflammatory markers were elevated.

Computed tomography (CT) of the chest (performed to look for evidence of TB reactivation) (Figure 2) revealed extensive ill-defined confluent nodular soft tissue with multiple calcific foci in the lower mediastinum, engulfing the heart and lower esophagus, extending to the subcutaneous left chest wall and upper abdomen. No consolidation, tree-in-bud nodularity, or pleural effusion were seen. Calcified granulomas in the left upper lobe and both adrenal glands were consistent with prior TB. Our initial impression was that of chronic granulomatous adenitis with no evidence of active TB infection. There was also nodular thickening and calcification of the esophageal wall, which raised concern for granulomatous involvement.

He underwent esophagogastroduodenoscopy (OGD) the following day, which showed multiple bluish submucosal lesions along the length of the esophagus, raising suspicion of the presence of dilated varices (Figure 3). His symptoms resolved and he had no further febrile episodes during his hospital stay, even with cessation of antibiotic therapy. Three sets of acid-fast bacilli (AFB) sputum cultures were negative, and he was discharged with a diagnosis of upper-respiratory tract infection (URTI).

He was re-admitted 1 month later with fever and cough of several days' duration. In view of persistent respiratory symptoms despite negative sputum cultures, he underwent anterolateral thoracotomy and open biopsy (under cardiothoracic surgery) of the pericardial mediastinal soft tissue to search for a source of active infection (and simultaneously obtaining tissue cultures to guide therapy). Histology showed fibroadipose tissue



Figure 1. Chest radiograph demonstrates left upper-zone scarring and calcified granulomas, in keeping with the history of tuberculosis. Note the subtle increased opacification over the left middle to lower zones.

with anastomosing variably-sized vascular channels (mainly veins and capillaries) and no granulomatous inflammation or evidence of malignancy, in keeping with a vascular malformation (Figure 4). The post-operative hospital stay was uneventful and he was discharged in good condition.

At outpatient follow-up with Gastroenterology prompted by the findings on the earlier OGD, he underwent elective magnetic resonance imaging (MRI) of the liver with additional sequences for coverage of the thoracic abnormality.

MRI revealed no evidence of liver cirrhosis. The known vascular malformation in the lower mediastinum to upper abdomen showed serpiginous, increased T2-weighted signal with post-contrast enhancement (Figure 5). We found scattered foci of low T1- and T2-weighted signals throughout the lesion, corresponding to calcification on the CT, which were consistent with phleboliths. No fluid-fluid levels or cystic spaces were identified. No large feeding artery was detected on magnetic resonance angiography (MRA).

Overall features were consistent with an extensive low-flow venous vascular malformation in the mediastinum and upper abdomen, indenting on the esophageal wall, and the patient was discharged from Gastroenterology. In view of the extensive infiltrative nature of the vascular malformation, necessitating challenging surgery if resected, as well as the lack of significant mass effect on critical structures, our patient was managed conservatively. The malformation remained stable on follow-up CT more than a decade later (Figure 6).

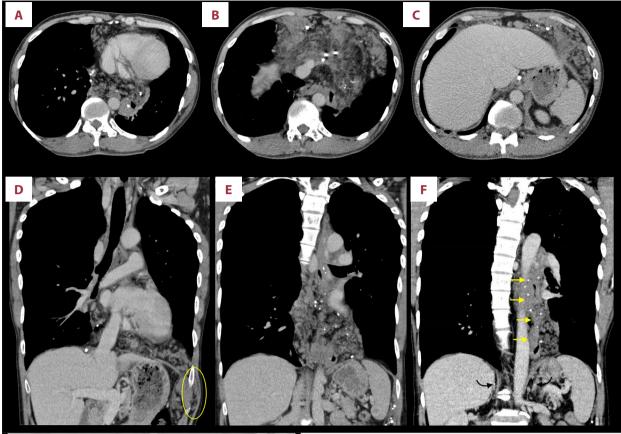




Figure 2. Sequential axial (A-C) and coronal (D-F) CT chest images show a large infiltrative nodular soft-tissue mass containing multiple calcific foci in the lower mediastinum surrounding the heart and esophagus, extending to the subcutaneous left chest wall (D, yellow oval) and upper abdomen. There is nodular thickening and calcification inseparable from the esophageal wall (F, yellow arrows). Calcified granulomas in the left lung upper lobe (G, coronal CT lung window) and bilateral adrenal glands (F, curved black arrows) are in keeping with prior TB.

#### **Discussion**

Vascular anomalies are a rare heterogeneous group of lesions which can affect any part of the body. The International Society for the Study of Vascular Anomalies (ISSVA) classification (revised 2014) [1] is the most widely recognized classification system, dividing vascular anomalies into vascular tumors (e.g., hemangiomas) and malformations, which can comprise any combination of arterial, venous, capillary, and lymphatic components.

Vascular malformations are congenital in nature, usually growing proportionally with the individual, without regression [2]. They are often infiltrative, involving multiple tissue planes and compartments, unlike vascular tumors.

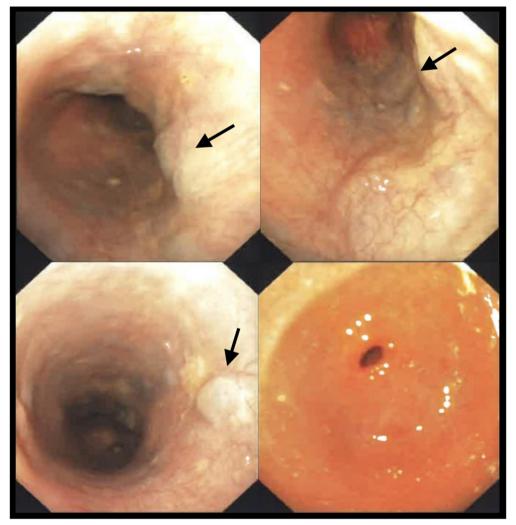


Figure 3. Direct visualization on OGD demonstrating tortuous dilated submucosal lesions with bluish tinge in the lower esophagus mimicking varices (arrows); bottom right-normal upper esophagus for comparison.

Ultrasound (US) and MRI are the primary noninvasive imaging modalities for vascular malformations [3]. Both lack ionizing radiation, are able to clearly identify the vascular nature of a lesion, and provide valuable information on flow dynamics. MRI is preferred for its reproducibility and ability to delineate the extent of non-superficial malformations, such as in our case.

On US, vascular malformations are predominantly heterogeneous hypoechoic lesions with internal color flow. High-flow malformations show arterial waveforms or arteriovenous shunting on Doppler interrogation [4]. On MRI, vascular malformations appear as septated lobulated masses with low T1-weighted and high T2-weighted signals. High-flow malformations contain enlarged feeding arteries and draining veins, often forming a tangled "nidus" of abnormal vessels, early arterial enhancement, and flow voids [2,3]. Low-flow venous malformations show gradual venous and delayed enhancement, absence of flow voids, low-signal-intensity phleboliths, and occasional perilesional edema.

Presence of non-enhancing cystic areas containing fluid-fluid levels suggests a concomitant lymphatic component [2,3].

CT has good spatial resolution and can be used to delineate the extent of the malformation as well as involvement of adjacent structures [3]. Calcification, thrombosis, and phleboliths, which are hallmarks of low-flow malformations, are also clearly visible on CT but requires exposing the patient to ionizing radiation.

Differentiation into high-flow (containing an arterial component) and low-flow malformations is critical for therapeutic planning and prognostication, with the latter accounting for the majority (up to 90%) of lesions outside of the central nervous system [2,5]. Low-flow venous malformations are seen in 1–4% of the population [3,6] and are usually located in the head and neck (40% of cases), extremities (40%), and trunk (20%) [2,7–9]. Mediastinal vascular malformations are very rare,

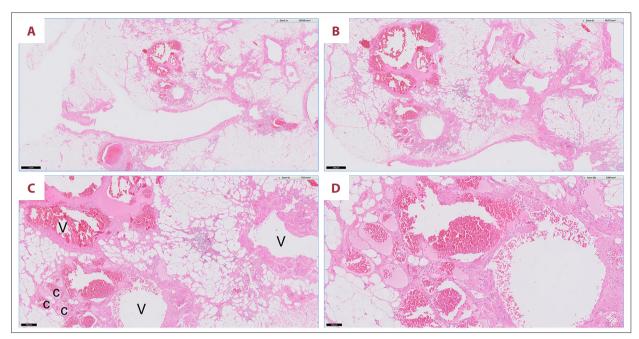
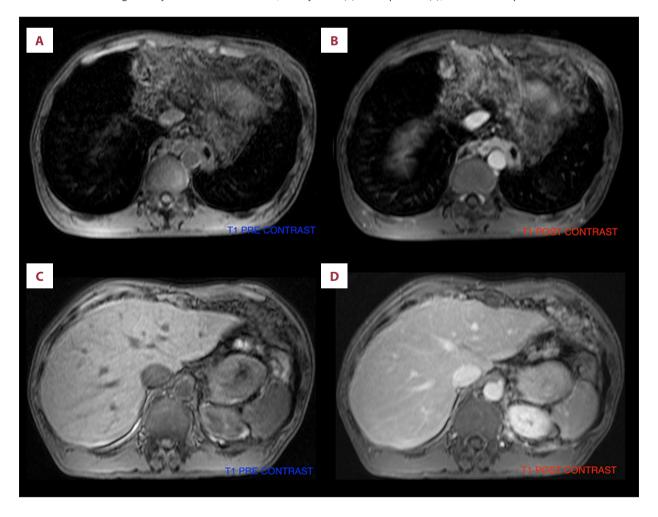


Figure 4. Histology photomicrographs (hematoxylin and eosin staining; (A) ×1, (B) ×2, (C) ×5, and (D) ×10 magnification) show multiple anastomosing variably-sized vascular channels, mainly veins (V) and capillaries (C), within fibroadipose tissue.



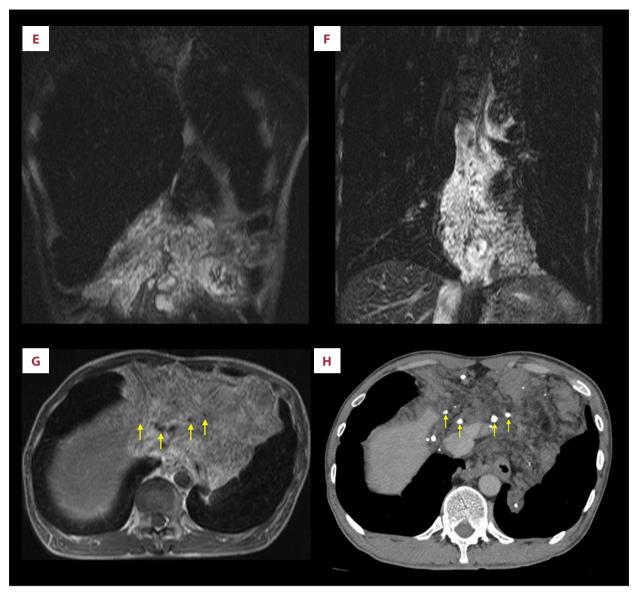


Figure 5. Paired axial T1-weighted pre- (A, C) and post- (B, D) contrast MRI images show avid serpiginous enhancement in keeping with the underlying vascular nature of the mass. Coronal T2-weighted fat saturation (E, F) MRI reveals marked T2-weighted hyperintensity. Scattered intralesional hypointense foci corresponding to calcification on the CT (G, H, yellow arrows) are in keeping with phleboliths.

with only isolated case reports in the existing literature; of 6 previously described cases, 2 were high-flow malformations (arteriovenous) and 4 were low-flow malformations (venous or lymphovenous) [10–14].

In our patient, the mediastinal vascular malformation was first detected on a CT done for respiratory symptoms. Given the clinical history of recurrent acute respiratory symptoms on a background of incompletely treated pulmonary TB and CT findings of extensive calcified nodular soft tissue in the mediastinum, tuberculous lymphadenitis [15] was initially thought to be the primary consideration, ultimately resulting in performing

a biopsy for confirmation. Interestingly, the abnormal dilated vessels comprising the mediastinal vascular malformation were also initially mistaken for esophageal varices on OGD.

MRI performed at a later date showed imaging features in keeping with a mediastinal low-flow venous vascular malformation. In retrospect, had the differential diagnosis of a vascular malformation been considered, an earlier corroborative MRI might have spared the patient from invasive open biopsy and OGD.

Management of vascular malformations requires a complex multidisciplinary approach, given the widely variable presentation,

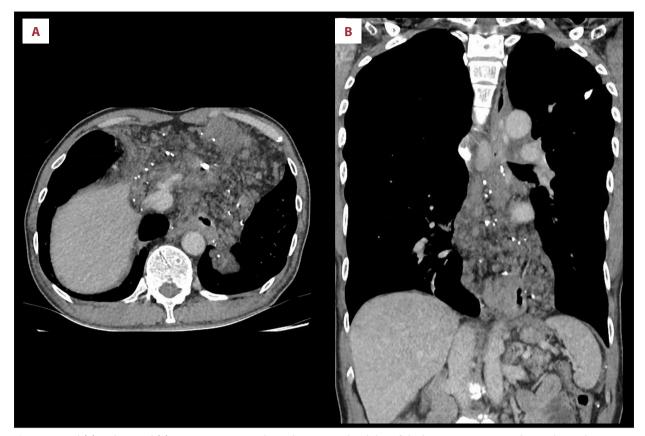


Figure 6. Axial (A) and coronal (B) CT images 11 years later show interval stability of the known extensive mediastinal vascular malformation.

prognosis, and configuration of these lesions [4]. Treatment is generally indicated if the patient has significant complications or functional impairment (e.g., hemorrhage, mass effect on critical structures, threatened limb, intractable pain). Localized intralesional coagulopathy can result in thrombosis and increased risk of deep-vein thrombosis, pulmonary embolism, or, rarely, stroke [14]. Lesion morphology, available expertise, and patient preference are key in deciding between conservative, surgical, or interventional radiologic (such as sclerotherapy or embolization) management.

Our patient remained well on conservative management and did not manifest any compressive or thrombogenic complications; the malformation was stable on follow-up imaging more than 1 decade later. In previous case reports, the majority of low-flow vascular malformations (3 out of 4) were treated conservatively [10–14], with 1 showing stability on follow-up 3 years later [10].

#### **Conclusions**

Mediastinal vascular malformations are rare lesions for which diagnosis can be challenging in the setting of a complicated clinical presentation. Radiologists should be aware of this important differential diagnosis for extensive calcified soft-tissue lesions in the mediastinum involving multiple soft-tissue planes and compartments. Multi-modality imaging, particularly MRI, which can demonstrate typical features, is crucial for image-based diagnosis and prognostication, thereby avoiding unnecessary invasive procedures and treatment.

#### **Conflict of interest**

None.

#### **References:**

- 1. ISSVA Classification of Vascular Anomalies ©2018 International Society for the Study of Vascular Anomalies. issva.org/classification.
- Flors L, Carlos LS, Ismaeel MM et al: MR imaging of soft-tissue vascular malformations: Diagnosis, classification and therapy follow-up. Radiographics, 2011; 31: 1321–40
- 3. Usman B, Samd S, Sunil J et al: Magnetic resonance (MR) imaging of vascular malformations. Pol J Radiol, 2017; 82: 731–41
- Gerald ML, Manraj KSH: Venous malformations: Classification, development, diagnosis and interventional radiologic management. Radiol Clin N Am, 2008; 46: 545–97
- Moukaddam H, Pollak J, Haims AH: MRI characteristics and classification of peripheral vascular malformations and tumors. Skeletal Radiol, 2009; 38(6): 535–47
- Ryu CW, Kim JK, Kim SJ et al: Head and neck vascular lesions: Characterization
  of the flow pattern by the use of three-phase CT. Korean J Radiol, 2009;
  10(4): 323–32
- Dubois J, Garel L: Imaging and therapeutic approach of hemangiomas and vascular malformations in the pediatric age group. Pediatr Radiol, 1999; 29(12): 879–93

- Fayad LM, Hazirolan T, Bluemke D et al: Vascular malformations in the extremities: Emphasis on MR imaging features that guide treatment options. Skeletal Radiol, 2006; 35(3): 127–37
- Laor T, Burrows PE: Congenital anomalies and vascular birthmarks of the lower extremities. Magn Reson Imaging Clin N Am, 1998; 6(3): 497-519
- Andres R, Daniel R, Michael B et al: Mediastinal venous vascular malformations: Report of two cases, with discussion of imaging findings and classification systems. Clinical Imaging, 2014; 38: 218–20
- Oztekin O, Kivanc M, Mehmet G et al: A congenital arteriovenous malformation originating from the aorta locating in the posterior mediastinum. Ann Thorac Cardiovasc Surg, 2012; 18: 387–90
- Yoshihisa K, Tomoki U, Tomohiro K et al: Lymphatic and venous malformation or "lymphangiohemangioma" of the anterior mediastinum: Case report and literature review. Gen Thorac Cardiovasc Surg, 2011; 59: 575–78
- Charlene T, Tom R, Anthony C et al: Arteriovenous malformation in the anterior mediastinum. Ann Thorac Surg, 2010; 90: e9–10
- Rajiah P, Kanne JP: Mediastinal vascular malformation presenting with stroke. Br J Radiol, 2010; 83: e138–42
- Woo KM, Jung-Gi I, In KY et al: Mediastinal tuberculous lymphadenitis: MR imaging appearance with clinicopathologic correlation. Am J Roentgenol, 1996; 166: 21–5