Case report

A case of huge lymphatic and venous malformations of the mediastinum

Miyuki Munechikaa,*, Kazunori Tobinob, Masanobu Okahisa, Yuki Gotou, Kojin Murakami, Takuto Sueyasu, Saori Nishizawa, Kouhei Yoshimine

a Department of Respiratory Medicine, Iizuka Hospital, 3-83 Yoshiomachi, Iizuka, Fukuoka 820-0018, Japan
b Department of Respiratory Medicine, Juntendo University, School of Medicine, 2-1-1 Hongo, Bunkyo-Ku, Tokyo 113-8421, Japan

ARTICLE INFO

Keywords:
Lymphatic and venous malformations
Vascular tumors
Vascular malformations

ABSTRACT

We herein report a case of huge lymphatic and venous malformations of the mediastinum (LVM). A 19-year-old man presented for evaluation of a mediastinal mass. On chest computed tomography, the mass demonstrated contrast enhancement and showed dilated veins draining into the superior and inferior vena cava, with multiple scattered calcifications. The lesion was enhanced heterogeneously on T1-weighted magnetic resonance imaging (MRI) and hyperintense on T2-weighted MRI. Contrast-enhanced MRI revealed that the mass was enhanced, with the multilocular part marginally enhanced. From these images, we diagnosed him with LVM. Given that an operation presented a high risk, we decided to follow him up without any treatment.

1. Introduction

Lymphatic and venous malformations (LVMs) are considered tumor-like lesions combining dysplastic lymphatic and venous vessel structures. LVMs of the mediastinum are rare, and to our knowledge, only 14 cases have been reported in the English literature. We herein report a case of huge mediastinal LVM diagnosed by computed tomography (CT) and magnetic resonance imaging (MRI) features.

2. Case presentation

A 19-year-old Japanese man presented to our hospital for further evaluation of abnormal chest X-ray findings on a routine health checkup. This abnormality had been pointed out for the first time. He had no symptoms. He had experienced no developmental or growth problems and never smoked. His vital signs were within normal limits, and the findings on a physical examination was completely normal. Auscultation of the chest revealed clear breath sounds throughout all lung fields. His cardiac sounds were normal, and no murmurs were present. He had no abdominal masses, hepatomegaly, splenomegaly, or peripheral edema. A chest X-ray demonstrated a large right-sided mediastinal mass that obscured the cardiac border (Fig. 1), and an enhanced chest CT showed a smooth marginated huge mass extending from the upper mediastinum to the level of the diaphragm, and the mass also extended from the anterior to the posterior mediastinum at the level of the diaphragm (Fig. 2A–C). Punctate calcific opacities within the mass were identified. On contrast-enhanced CT, the mass demonstrated nodular and tubular enhancing structures, fat attenuation, and multiple scattered calcifications. The flow of contrast medium from the superior vena cava (SVC) into the mass was also identified. Furthermore, in the spleen, multiple thin-walled low-attenuation masses with no enhancement located at the subcapsular region were observed (Fig. 2D–F). On T1-weighted MRI, the lesion appeared as a heterogeneous mass of predominantly intermediate signal intensity (Fig. 3A–C), and on T2-weighted MRI, it appeared as a hyperintense mass with a flow void that was thought to represent an enlarged vein draining into the inferior vena cava (IVC) (Fig. 3D–F). Contrast-enhanced T1-weighted MRI revealed multiple tortuous serpentine enhanced structures and enlarged veins draining into the SVC and IVC. A part of the mass was multilocular and marginally enhanced (Fig. 3G–I). The mass did not have any definite evidence of invasion to adjacent structures or pericardial effusion. Given these imaging findings, the patient was thought to have a mediastinal lymphatic and venous malformation (LVM).

Biopsy of the lesion was needed for a definitive diagnosis, but we opted to avoid biopsy, as the mass was too extensive to remove completely and the risk of bleeding after biopsy was thought to be high. On contrast-enhanced T1-weighted MRI, most of the splenic masses appeared to have low signal intensity, but one appeared to have high signal intensity, which was thought to represent internal bleeding or large amounts of intracystic proteinaceous content. These lesions appeared to be multiloculated hyperintense masses on T2-weighted MRI, and a diagnosis of splenic cysts was given. Thereafter, the patient had no symptoms, and the mass did not show any changes on CT images.

* Corresponding author.
E-mail address: makamatsuh1@aih-net.com (M. Munechika).

https://doi.org/10.1016/j.rmcr.2018.11.014

Received 20 September 2018; Accepted 20 November 2018

2213-0071/ © 2018 The Authors. Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/BY-NC-ND/4.0/).
obtained six months later.

3. Discussion

The International Society for the Study of Vascular Anomalies (ISSVA) mainly classifies vascular anomalies as vascular tumors and vascular malformations [1]. Vascular tumors are vascular anomalies with a proliferative component and are divided into three types: benign, locally aggressive or borderline, and malignant tumors (Table 1). Most vascular tumors occur in infants and undergo a characteristic two-stage process of growth and regression. Therefore, most vascular anomalies in young adults without spontaneous regression are vascular malformations. Vascular malformations are relatively static lesions attributed to inborn errors in vasculogenesis and are divided into four types: simple, combined, of major named vessels, and associated with other anomalies. Simple vascular malformations are further classified on the basis of the main type of vessel: capillary, venous, lymphatic, arterial, arteriovenous, and arteriovenous fistula.

Most vascular tumors and malformations are diagnosed based on the clinical course and findings from a physical examination. Biopsy is needed for a definitive diagnosis but can confer a risk of bleeding due to the abundant vascular flow. Therefore, non-invasive examinations such as an ultrasound examination or MRI are useful for making a diagnosis and determining courses of treatment [2].

Vascular malformation of the mediastinum is rare and accounts for 0.5% of mediastinal tumors [3]. It commonly occurs in the anterior mediastinum. Half of all such patients have no symptoms, while the other half have symptoms such as cough, chest pain, and dyspnea.

CT and MRI features of LVM depend on the proportions of lymphatic and venous components. Lymphatic components are shown as low and markedly high-intensity areas on T1- and T2-weighted MRI, respectively. They show no internal enhancement on dynamic postcontrast imaging [4]. The imaging features of venous components include collections of serpentine structures partitioned by septa, and phleboliths. T1- and T2-weighted MRI show intermediate- and high-intensity masses, respectively [5]. Phleboliths and thromboses are shown as punctate or rounded low-intensity areas, known as “dot signs”. Sequence scans after the injection of contrast material in both CT and MRI are useful for delineating slow-flow areas of venous components. In our patient, CT and MRI showed typical findings, with venous connections to the dilated SVC and IVC. As in our patient, some cases of mediastinal LVM with venous connections to the SVC have been reported [6,7].

While a biopsy is needed to make a definitive diagnosis of vascular malformation, it can confer a risk of bleeding due to the abundant vascular flow. In our patient, biopsy was avoided because the mass was too extensive to remove completely, and the risk of intractable bleeding after partial excision was thought to be high.

Fig. 1. Chest X-ray. A large right-sided mediastinal mass obscured the cardiac border.

Fig. 2. Chest CT images. A-C: Unenhanced chest CT images. The smoothly margined huge mass extended from the upper mediastinum to the level of the diaphragm, and also from the anterior to the posterior mediastinum at the level of the diaphragm. Punctate calcific opacities within the mass were identified (arrows on B). D-F: Contrast-enhanced CT images. The mass demonstrated nodular and tubular enhancing structures, fat attenuation, and multiple scattered calcifications. The flow of contrast medium from the SVC into the mass was also identified (arrow on D). In the spleen, multiple thin-walled low-attenuation masses with no enhancement located at the subcapsular region were observed.
The treatment choices for vascular malformations are surgery and sclerotherapy. Surgery is sometimes useful in isolated, symptomatic venous malformations or following sclerotherapy; however, it is usually difficult to completely remove the malformation. Recurrence following surgery is more common in patients with diffuse malformations or incomplete excision than in those with solitary malformations or successfully treated cases [8]. Sclerotherapy is the primary form of nonsurgical intervention. In some cases, multiple sclerotherapy sessions are needed. Generally, treatment is more successful in patients with pure venous malformations than in those with combined malformations [9]. Thus far, 14 cases of mediastinal LVM have been reported in the English literature, 11 of whom were treated with surgery. Only one case was treated with endoscopic sclerotherapy through the esophagus. In our patient, the LVM was too large to completely remove. In addition, the LVM was directly connected to the SVC and IVC, and sclerotherapy was thought to have a high risk of complications due to pulmonary and systemic infarction. Therefore, we decided to follow the patient without any treatment. The mass remained stable on CT images obtained six months after the diagnosis, a clinical course that suggests our diagnosis was correct. The patient has been followed-up closely.

In conclusion, we presented a case of huge LVM of the mediastinum diagnosed with CT and MRI. This condition is very rare, and biopsy can be difficult; therefore, obtaining characteristic CT and MRI findings is important for making a definitive diagnosis.

Declarations of interest

None.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2018.11.014.

References

[1] O. Enjolras, J.B. Mulliken, Vascular tumors and vascular malformations: new issues, Adv. Dermatol. 13 (1997) 375–423.
[2] A. Yamazaki, H. Miyamoto, Y. Saito, et al., Cavernous hemangioma of the anterior mediastinum: case report and 50-year review of Japanese cases, Jpn. J. Thorac. Cardiovasc. Surg. 54 (2006) 221–224.
[3] Y. Kadota, T. Utsumi, T. Kawamura, et al., Lymphatic and venous malformation or “lymphangiohemangiomata” of the anterior mediastinum: case report and literature review, Gen. Thorac. Cardiovasc. Surg. 59 (2011) 575–578.
[4] L.F. Donnelly, D.M. Adams, G.S. Bisset 3rd, Vascular malformations and hemangioma: a practical approach in a multidisciplinary clinic, AJR Am. J. Roentgenol. 174 (2000) 597–608.
[5] L.J. Abernethy, Classification and imaging of vascular malformations in children, Eur. Radiol. 13 (2003) 2483–2497.
[6] M. Riquet, J. Briere, F. Le Pimpec-Barthes, et al., Lymphangiohemangiomata of the mediastinum, Ann. Thorac. Surg. 64 (1997) 1476–1478.
[7] P.J. Bruyere, G. Trotteur, E. Creemers, et al., Mediastinal lymphangiohemangiomata associated with superior vena cava stenosis, JBR-BTR 89 (2006) 116–117.
[8] Y. Kadota, T. Utsumi, T. Kawamura, et al., Lymphatic and venous malformation or “lymphangiohemangiomata” of the anterior mediastinum: case report and literature review, Gen. Thorac. Cardiovasc. Surg. 59 (2011) 575–578.
[9] C.M. Giguerre, N.M. Başman, Y. Sato, et al., Treatment of lymphangiomata with OK-432 (Picibanil) sclerotherapy: a prospective multi-institutional trial, Arch. Otolaryngol. Head Neck Surg. 128 (2002) 1137–1144.