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CASE REPORT
Other

A case report of generalized lymphangiomatosis with chylopericardium: the crucial role of magnetic resonance lymphangiography

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Background Chylopericardium due to generalized lymphangiomatosis is a rare clinical entity. Its aetiology and treatment remain unclear.

Case summary We report one case of a 51-year-old man who was diagnosed with generalized lymphangiomatosis with idiopathic chylopericardium by bilateral inguinal intranodal contrast-enhanced magnetic resonance lymphangiography. Magnetic resonance lymphangiography demonstrated abnormal communications between the pericardial sac and the thoracic duct. The patient with idiopathic chylopericardium was therefore successfully treated by exclusive surgical ligation of the abnormal communications and partial pericardiectomy by thoracotomy. The patient's postoperative recovery was uneventful, and no recurrence of pericardial effusion occurred during the 13-month follow-up.

Discussion Magnetic resonance lymphangiography showed a good capability for evaluating the extent of generalized lymphangiomatosis and therefore is helpful for delineating the anatomy of the thoracic duct and identifying chyle leakage and abnormal communications between the thoracic duct and the pericardial sac. This makes a simple ligature of the abnormal communications, instead of thoracic duct mass ligation possible in the treatment of chylopericardium.

Keywords Generalized lymphangiomatosis • Chylopericardium • MR lymphangiography • Case report

Learning points

- Chylopericardium caused by generalized lymphangiomatosis is rare and can be first found in middle-aged people with no symptoms in young age.
- Magnetic resonance (MR) lymphangiography is a powerful tool to characterize chylopericardium and to search for an underlying disease.
- A simple ligature of the abnormal communications could replace thoracic duct mass ligation with the guide of MR lymphangiography.

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Introduction

Chylopericardium is an uncommon clinical condition characterized by the accumulation of chylous fluid in the pericardial cavity.[1] It is generally associated with lymphatic agenesis and malformations, lymphangiectasia, Behçet disease, infection, malignancy, cardiac surgery, and trauma.[2] When generalized lymphangiomatosis leads to pericardial effusion, it is referred to as idiopathic pericardium—a rarely reported phenomenon. The treatment and prognosis of this disease depends upon the organs involved and the extent to which they are affected.[3]

Timeline

Time	Events
Before admission	The patient suffered abdominal distension and mild diarrhoea 5–6 times a day for 6 months, with slight dyspnoea while doing physical work for 1 month
September 2018	Admission. Transthoracic echocardiography revealed a large amount of pericardial effusion, which was confirmed to be chylopericardium via pericardiocentesis
Within 1 month after admission	Subxiphoid exterior tube drainage was maintained, and a low-fat and medium-chain triglyceride diet was instated. Significant chylopericardium continued
1 month after admission	The patient was diagnosed with generalized lymphangiomatosis involving the mediastinum or mediastinal lymphangiectasia by magnetic resonance lymphangiography, which also presented the abnormal communication between the pericardial sac and the thoracic duct
1 month and 1 week after admission	Surgery. The patient underwent exclusive surgical ligation of this abnormal communication and partial pericardiectomy by thoracotomy
10 days after surgery	The patient was discharged
8 weeks after surgery	The patient returned to normal diet, full-time work, and full activities
A 13-month follow-up after surgery	Echocardiography showed no recurrence of chylopericardium

Case presentation

A 51-year-old man was accidentally found pericardial effusion and referred to the Cardiology Department. Before admission, the patient had suffered abdominal distension and mild diarrhoea 5–6 times a day for 6 months, with slight dyspnoea while doing physical work for 1 month. He just had a history of left tibia and fibula fracture when he was 21 years old and no other special past medical history.

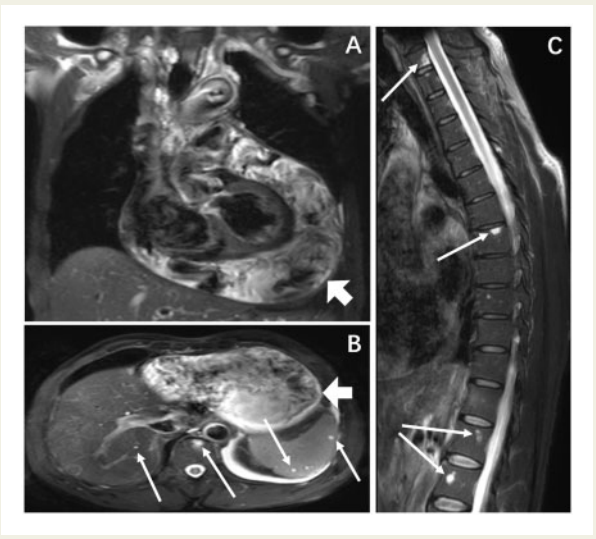

Figure 1 Thoracic and abdominal T2-weighted magnetic resonance imaging with fat suppression showed the extent of the disease. (*A* and *B*) Large amount of effusion with inhomogeneous hyperintensity within pericardium (coarse arrows). (*B* and *C*) Multiple high-signal intensity lesions in the liver and the spleen, and multiple hyperintense well-defined osteolytic lesions in the vertebral bodies (thin arrows).

The patient did not present with fever on examination. His blood pressure was 125/78 mmHg and the pulse was 76 b.p.m. The bilateral lung auscultation showed no wheezing or crackles. Cardiovascular examination demonstrated a regular heart rhythm without murmurs, and the heart sound was distant. An electrocardiogram showed low QRS voltages. Transthoracic echocardiography revealed a large amount of pericardial effusion with 12–25 mm of fluid in the apex and basal area but no atrial or ventricular collapse, with left ventricular ejection fraction (LVEF) 68%.

Pericardiocentesis was performed three times within 2 weeks to alleviate the symptom of dyspnoea, and ~1500–1800 mL of reddish milky fluid was removed each time. The effusion was confirmed to be chylopericardium and had the following findings: specific gravity ≥1.028; glucose, 7.5 mmol/L; total protein, 78 g/L; albumin, 32 g/L; lactate dehydrogenase, 135 U/L; triglycerides, 15.2 mmol/L; and total cholesterol, 5.1 mmol/L; cholesterol/triglyceride ratio >1; red blood cell count, 3540/mm^3; white blood cell count, 830/mm^3; monocytes, 87%; and neutrophils, 13%. Sudan III staining of the fluid revealed fat globules. Bacterial and tuberculosis cultures were negative. Cytology demonstrated an abundance of lymphocytes and individual heterotypic cells without tumour cells.

Subxiphoid exterior tube drainage was maintained, and a low-fat and medium-chain triglyceride diet was instated. After initial success with this treatment, significant chylopericardium continued. The patient was subsequently referred for surgery.

Magnetic resonance (MR) lymphangiography was performed prior to surgery. Before that, abdominal and thoracic T2-weighted MR imaging with a fat suppression sequence was conducted to evaluate

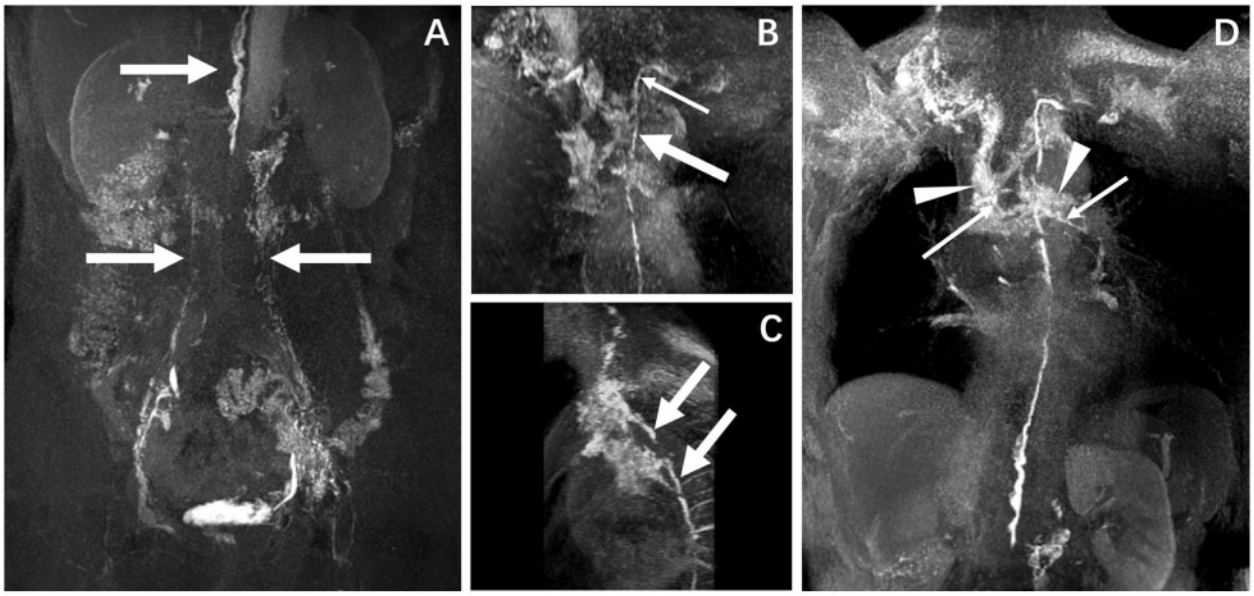

Figure 2 Three-dimensional maximum intensity projection of a T1-weighted enhanced lymphangiogram showed the lymphatic trunks, thoracic duct, and its branches. (*A*) Abdominal and pelvic position, contrast material appeared in the lumbar lymphatic trunks (coarse arrows) and cisterna chyli (coarse arrow) within 5 min after inguinal lymph nodes injection. (*B*) Partial thoracic position, contrast material appeared in the thoracic duct (coarse arrow) and left venous angle (thin arrow) within 10 min. An extensive network of collateral channels to the right side of the neck. (*C*) Lateral thoracic position, contrast material appeared in the thoracic duct (coarse arrows) and the upper mediastinal area. (*D*) Thoracic and partial abdominal position, certain abnormal communicating branches between the thoracic duct and the bilateral bronchomediastinal trunks abutting the pericardial sac (thin arrows) were observed ~10 min after gadopentetate injection. Pooling of contrast material was observed around bilateral bronchomediastinal trunks within the pericardial spaces and towards right neck (arrowheads).

the extent of the disease. On T2-weighted MR images, a large amount of effusion with inhomogeneous hyperintensity was identified within the pericardium (*Figure 1A and B*). Multiple high-signal intensity lesions involved the liver and spleen (*Figure 1B*). Additionally, multiple hyperintense well-defined osteolytic lesions were noted in vertebral bodies (*Figure 1C*). Intranodal lymphangiography was performed as described by Krishnamurthy *et al.*[4] using 22-gauge needles that were placed into the inguinal lymph nodes bilaterally under sonographic guidance. Paramagnetic contrast agent (2 mL of gadopentetate dimeglumine, 0.5 mol/L, Bayer Schering Pharma, Berlin, Germany) was injected into each inguinal lymph node. After injection, MR lymphangiography was performed using a three-dimensional fat-saturated spoiled gradient echo sequence. Contrast material appeared in the retroperitoneal lymphatics and cisterna chyli within 5 min after intranodal injection (*Figure 2A*) and in the thoracic duct (*Figure 2B and C*) and left venous angle (*Figure 2B*) within 10 min. An extensive network of collateral channels to the right side of the neck (*Figure 2B*). These collaterals communicated with the right supraclavicular vein. Certain abnormal communicating branches between the thoracic duct and the bilateral bronchomediastinal trunks abutting the pericardial sac (*Figure 2D*) were observed ~10 min after gadopentetate injection. Chylous regurgitation was observed from the thoracic duct towards the bilateral bronchomediastinal trunks, and pooling of the contrast material presented within the pericardial spaces and towards right neck (*Figure 2D*). Based on MR findings, the patient was diagnosed as generalized lymphangiomatosis with chylopericardium.

Surgery was conducted after MR lymphangiography. During surgery, the pericardium appeared thickened with numerous lymphangiectasias. After the descending aorta was mobilized, the thoracic duct and surrounding lymphatics were observed. The thoracic duct was branched and showed multiple dilated vessels connected with the left bronchomediastinal trucks and pericardium. Thus, the patient underwent ligation of several communicating branches, including the branches between the thoracic duct and left bronchomediastinal trucks. The thoracic duct-left internal jugular vein connections were preserved. Concomitant pericardial fenestration was performed. Pericardial biopsies showed a chronic inflammatory process with haemorrhagic infiltrate and a large amount of lymphangiectasias.

After 10 days, the patient was discharged, and after 8 weeks, he returned to normal diet, full-time work, and full activities. Echocardiography showed no recurrence of chylopericardium at a 13-month follow-up.

Discussion

Generalized lymphangiomatosis is traditionally known as pulmonary lymphangiectasis, generalized lymphangiectasis, and intrathoracic lymphangiomatosis[5,6] when the chest is involved. Pulmonary lymphangiectasis describes the pathological dilation of lymphatics and appears in children and young adults; however, it is quite rare in patients over the age of 40.[7] The case we describe here is therefore especially unusual due to the age at which it first presents. We propose that this patient should be categorized as having generalized lymphangiectasis with primarily mediastinal involvement and less severe pulmonary disease according to the classification proposed by Noonan *et al.*[8] Moreover, to our knowledge, mediastinal involvement of pulmonary lymphangiectasis is rare.

In this case, bilateral inguinal intranodal contrast-enhanced MR lymphangiography was performed, and intra-abdominal lymphatic vessels, cisterna chyli, and a thoracic duct were definitively demonstrated. The progression of the contrast medium showed the area of communications between the branches of the thoracic duct and the pericardial sac. Consistent with previous studies involving the introduction of MR lymphangiography, imaging of the central lymphatic system has changed our understanding of intrathoracic lymphatic diseases.[9,10] This technique used in this case is more sensitive than traditional lymphangiography and lymphoscintigraphy due to its higher contrast resolution and the lower viscosity of gadolinium compared to iodinated oil-based contrast agents, and it therefore provides more distal contrast propagation and allows the demonstration of the source of a chylous leak and small abnormal lymphatic communications, which can be traced from the abdomen.

The optimal treatment for patients with chylopericardium is unclear. Pericardiocentesis followed by pericardiostomy, a medium-chain triglyceride diet, or total parenteral nutrition can be attempted. However, conservative treatment has failed in 58.1% of previously reported cases.[2,11] In most cases, there are difficulties in identifying the site of chyle leak owing to anatomic variation of the thoracic duct, and thus the most commonly used surgical method is mass ligation of the thoracic duct above the level of the diaphragm along with the establishment of a pericardial window.[12] However, this approach cannot be used to restore the normal physiology of the thoracic duct.[13] In this case, preoperative MR lymphangiography clearly delineated the intrathoracic course of the thoracic duct and several tributaries between the thoracic duct and the pericardial sac. After the culprit communications were located, simple ligatures were employed, and mass thoracic duct ligation or excision was avoided.

Conclusion

People with generalized lymphangiomatosis can present with no symptoms in young age and be first diagnosed in middle age when it causes chylopericardium. Intranodal contrast MR lymphangiography provides an accurate and comprehensive imaging tool for diagnosing this rare disease and delineating abnormal anatomical lymphatic communications, which can guide a more targeted surgical approach.

Lead author biography

Dr Qing Lu got the Medical and Doctoral degree from Shanghai Jiao Tong University in 2012. As a senior Radiologic Technologist, he has 24 years of experience of operating MRI, X-ray, and CT technique. He leads and conducts researches on the lymphatic system diseases and cardiovascular diseases in radiology department. In last decade, he developed several novel CT and MR lymphangiography procedures to investigate lymphatic system disease, such as lymphedema, lymphatic vascular malformation, and lymphatic metastasis. He published more than 30 high-impacted papers on lymphatic imaging so far and his research has been publicly shared at national and international academic conferences many times.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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