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PICTORIAL REVIEW

Non-contrast 3D MR lymphography of retroperitoneal lymphatic aneurysmal dilatation: a continuous spectrum of change from normal variants to cystic lymphangioma

Sarah Derhy · Sanaâ El Mouhadi · Ana Ruiz · Louisa Azizi · Yves Menu · Lionel Arrivé

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Abstract

Objective Our objective was to demonstrate the characteristic features of retroperitoneal lymphatic aneurysmal dilatation with three-dimensional (3D) magnetic resonance (MR) lymphography.

Conclusion Three-dimensional MR lymphography demonstrates that retroperitoneal lymphatic aneurysmal dilatation exhibits a continuous spectrum of change from normal variants to lymphatic aneurysmal dilatation and so-called cystic lymphangioma.

Main Message

- *Non-contrast MR lymphography with very heavily T2-weighted fast spin echo sequences is a useful non-invasive technique without the need of contrast medium injection to obtain a unique evaluation of the lymphatic system*
- *To prove the lymphatic origin of a cystic formation, it is essential to demonstrate the communication with retroperitoneal lymphatic vessels*
- *3D MR lymphography demonstrates that retroperitoneal lymphatic aneurysmal dilatation exhibits a continuous spectrum of change from normal variants to lymphatic aneurysmal dilatation and so-called cystic lymphangioma*

Keywords Magnetic resonance · Lymphography · Lymphatic aneurysmal dilatation · Retroperitoneal

S. Derhy · S. El Mouhadi · A. Ruiz · L. Azizi · Y. Menu · L. Arrivé (✉)
Department of Radiology Hospital Saint-Antoine, Faculté de Médecine Pierre et Marie Curie, 184 rue du Faubourg Saint-Antoine, 75012 Paris, France
e-mail: lionel.arrive@sat.aphp.fr

Introduction

The retroperitoneal lymphatic system is markedly complex and characterised by numerous anatomical variants [1].

For a long time X-ray lymphography performed by injecting a radio-opaque material into a lymphatic vessel, surgically exposed and cannulated, has been the single method to explore the lymphatic system [2]. Presently, X-ray lymphography, which is an invasive procedure, is uncommonly performed in particular selected indications, such as preoperative evaluation before surgery for chylothorax

Non-contrast magnetic resonance (MR) lymphography with very heavily T2-weighted fast spin echo sequences appears to be a useful non-invasive technique without the need of contrast medium injection to obtain a unique evaluation of the lymphatic system [3–5].

In this pictorial review, we analyse the characteristic features of retroperitoneal lymphatic aneurysmal dilatation with three-dimensional (3D) MR lymphography.

Non-contrast MR lymphography protocol

MR lymphography was performed on a 1.5-T unit (Magnetom Symphony; Siemens Medical Solutions/Signa HDXt; GE Medical System) with phased-array body coil. Our protocol has already been described by our group [5].

Non-contrast MR lymphography techniques exploit the high signal intensity of static fluids in the lymphatic vessels with heavily T2-weighted sequences, which also result in decreased signal from background tissues. A free-breathing 3D high-spatial-resolution fast spin-echo sequence with a very long echo time (TE) and a voxel size of 1 × 1 × 1 mm is used. The main advantage of 3D isotropic MR lymphography is thinner section source images, which allow optimal post-processing of image

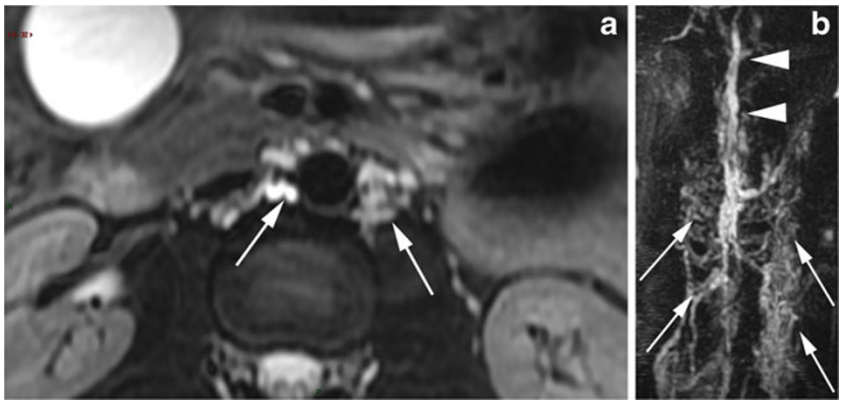

Fig. 1 A 39-year-old man with primary sclerosing cholangitis. **a** Axial T2-weighted MR image shows multiple high signal-intensity cystic formations (*arrows*) corresponding to lymphatic channels around aorta. **b** Coronal MIP image shows bilateral plexus of lymphatic channels (*arrows*) which continues with thoracic duct (*arrowheads*)

data to obtain maximum intensity projection (MIP) images and multiplanar reformatted (MPR) images. Use of 3D acquisition allows an improvement of both spatial resolution and contrast-to-noise ratio. Therefore, MR lymphography combines the advantages of projectional imaging with those of cross-sectional imaging. MIP 3D images provide an overview of lymphatic anatomy and abnormalities, while MPR images help visualise small or localised abnormalities such as connexion with normal lymphatic system.

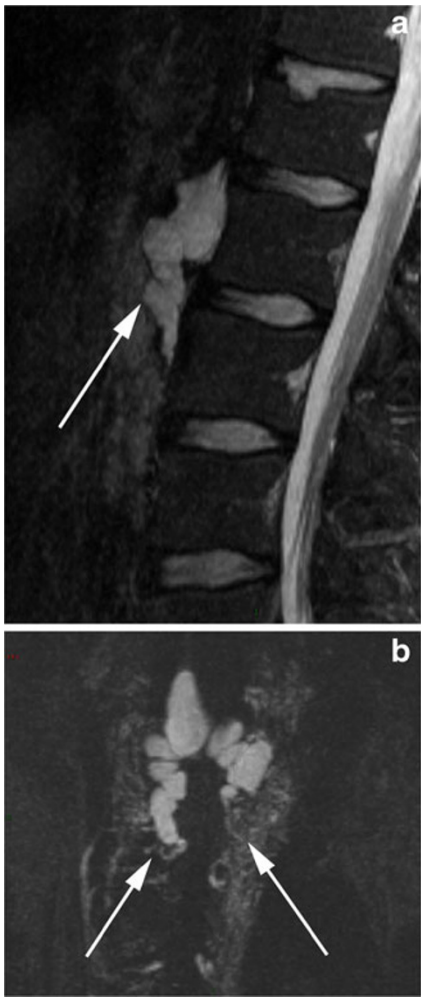

Fig. 2 A 25-year-old man who underwent MR imaging for back pain. **a** Sagittal T2-weighted MR image demonstrates a prevertebral cystic lesion (*arrow*) **b** Coronal MIP image shows giant cisterna chyli at confluence of left and right markedly dilated retroperitoneal trunks (*arrows*)

Lymphatic aneurysmal dilatation

The retroperitoneal lymphatic system is characterised by numerous anatomical variants [1]. The lumbar lymphatics, which are the continuation of the iliac lymphatics, ascend on either side of the aorta as two main trunks. They are not single vessels but a plexus of vessels that connect by lymphatics around the aorta. The appearance of retroperitoneal lymph

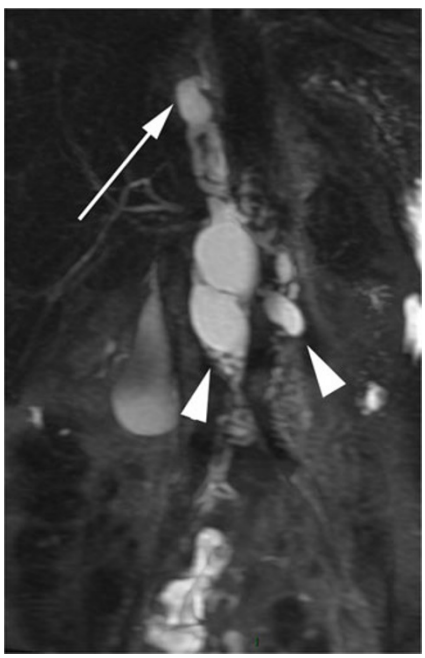

Fig. 3 A 37-year-old woman in which a cystic mass was discovered incidentally in a prevertebral location. Coronal MIP image shows a dilatation of cisterna chyli (*arrow*) and marked dilatation of both right and left retroperitoneal trunks (*arrowheads*)

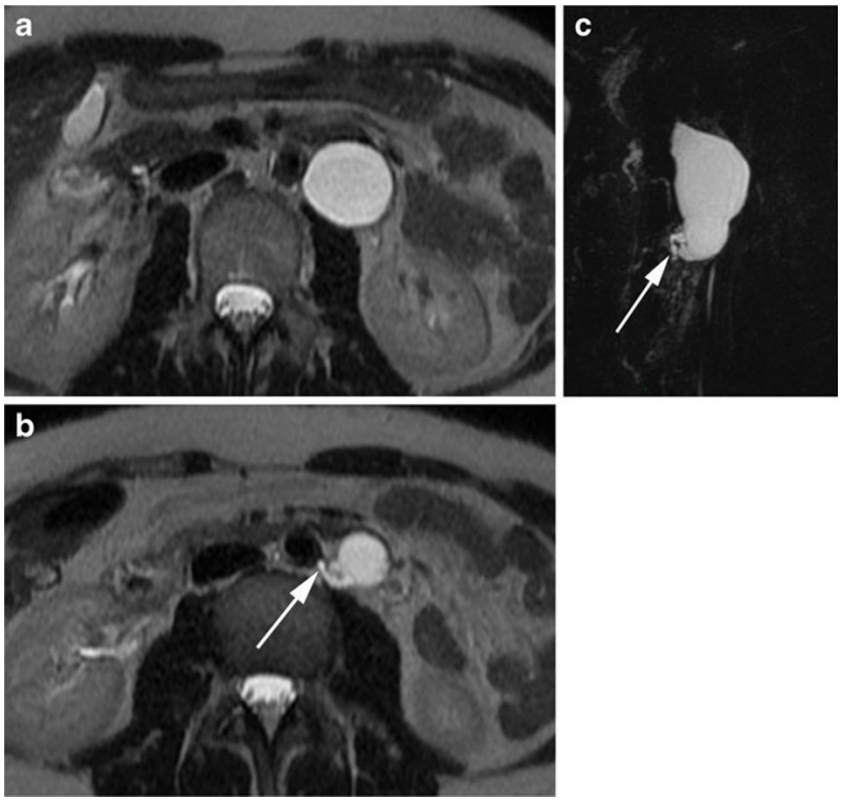

Fig. 4 A 47-year-old woman presenting with non-specific abdominal discomfort. Axial T2-weighted images (**a**, **b**) show a retroperitoneal, round mass with thin wall and homogeneous high signal intensity corresponding to fluid content. A mass effect on the small bowel is demonstrated. Extensions are present at the bottom of the lesion (*arrow*) (**b**). MR lymphography with coronal MIP image (**c**) confirms the diagnosis by demonstrating extension at the bottom of the lesion that communicates with dilated lymphatic channel (*arrow*)

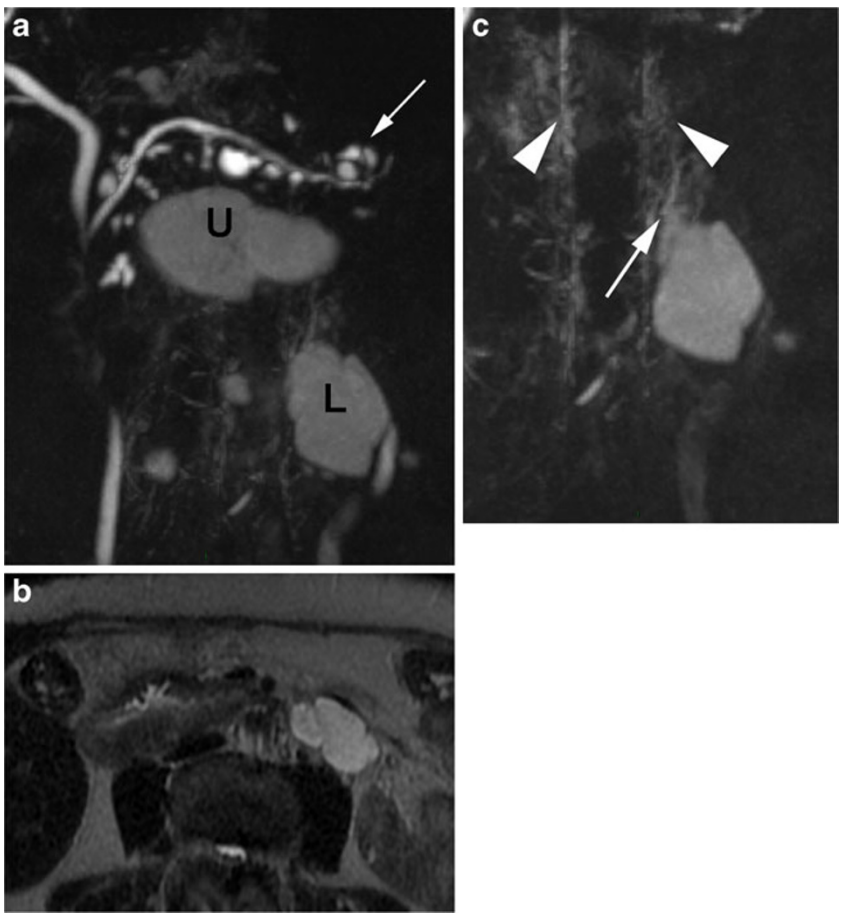

Fig. 5 A 60-year-old woman with intraductal papillary mucinous neoplasm (IPMN). Branch duct type IPMN (*arrow*) and two retroperitoneal cystic lesions (upper [*U*] and lower [*L*]) are demonstrated on coronal MIP image (**a**). Branch duct type IPMN and retroperitoneal lymphatic aneurysmal dilatation were proved at surgery. At follow-up, the second location of lymphatic aneurysmal dilatation that was not resected is well demonstrated (**b**). Extension of the lesion (*arrow*) is well demonstrated on coronal MIP image (**c**). Retroperitoneal trunks (*arrowheads*)

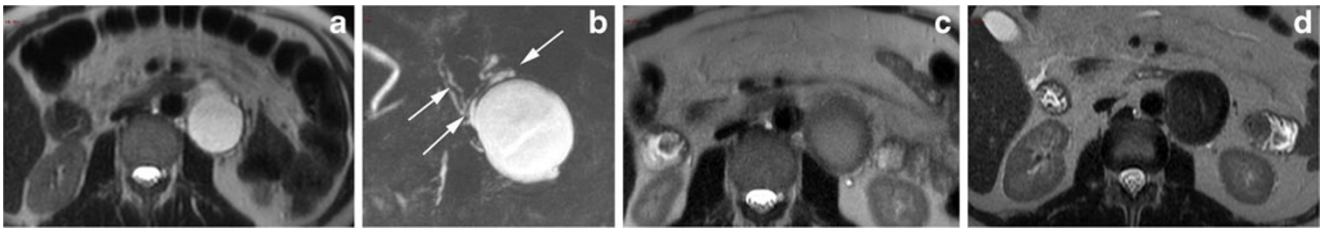

Fig. 6 A 41-year-old man with biopsy-proved intestinal lymphangiectasis and history of chylous ascites. On first MR examination the lesion has high signal on T2-weighted images (**a**). MR lymphography (**b**) shows the communication with dilated lymphatic channel (*arrows*). On first follow-up MR examination the lesion has lower signal on T2-weighted images (**c**) with very low signal intensity on MR lymphography on second follow-up (**d**). Low signal intensity on T2-weighted MR image is likely related to haemorrhage changes

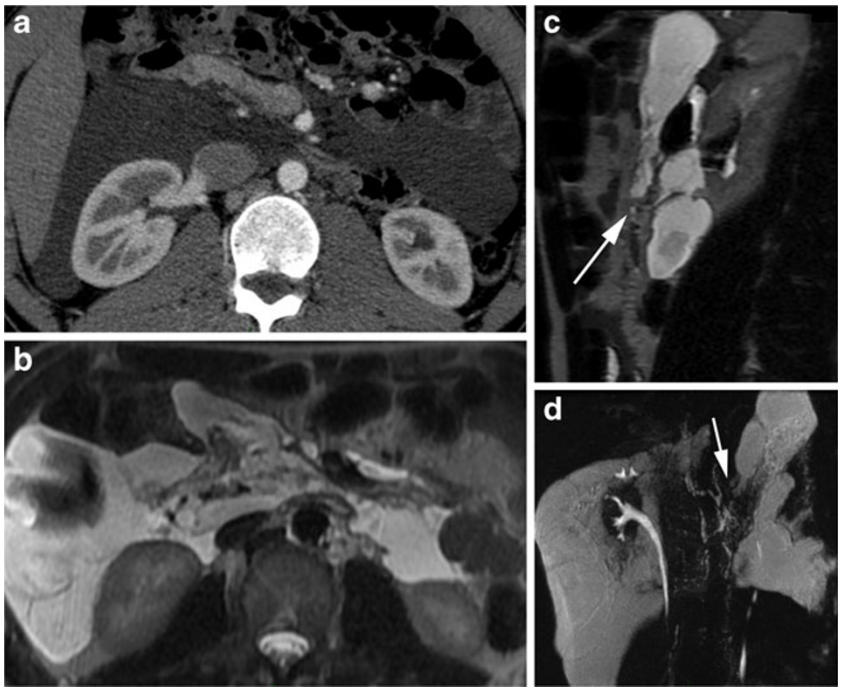

Fig. 7 An 18-year-old man presenting with abdominal discomfort. CT scan (**a**) demonstrates a large infiltrative retroperitoneal cystic lesion. On T2-weighted MR image (**b**) it appears multiloculated, with thin wall and septa. The content is serous with flow phenomena, appearing as characteristically shaped signal heterogeneity on T2-weighted images. Sagittal T2-weighted MR image (**c**) and coronal MIP image (**d**) show extensions (*arrow*) adjacent to surrounding lymphatic channels

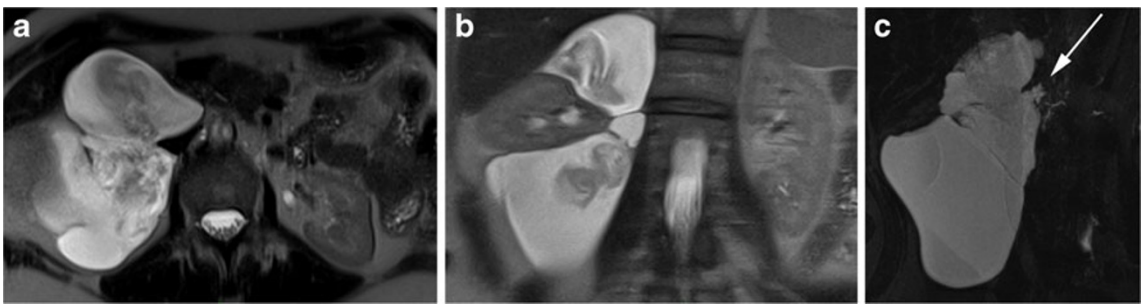

Fig. 8 A 60-year-old woman with non-specific abdominal pain. Axial (**a**) and coronal (**b**) T2-weighted images show a large multiloculated bulky lesion with serous content and characteristic shaped flow phenomena. Coronal MIP reconstruction of 3D MR lymphography (**c**) shows extensions adjacent to lymphatic vessels (*arrow*). Cystic lymphangioma was found at surgery

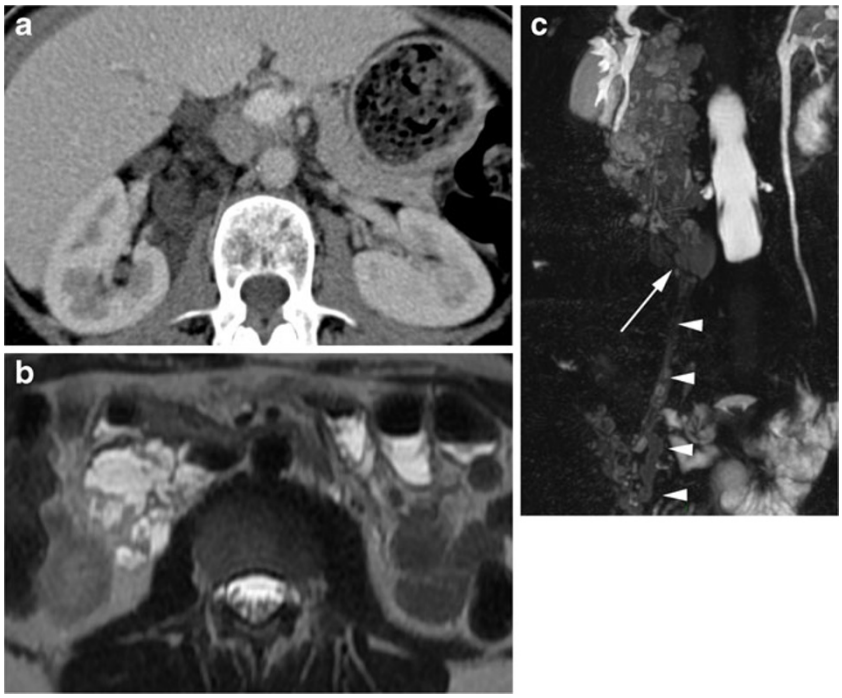

Fig. 9 A 71-year-old woman presenting with diffuse non-specific abdominal pain. CT scan (**a**) demonstrates an infiltrative multiloculated retroperitoneal cystic lesion. Axial T2-weighted image (**b**) shows infiltrative, multiloculated lesion in the right retroperitoneum with high signal intensity content. MR lymphography with MIP reconstruction (**c**) shows communication (*arrow*) between lesion and markedly dilated right iliac lymphatic vessels (*arrowheads*)

trunks markedly varies from thin to prominent thick channels, parallel or converging channels, or a plexus (Fig. 1). Alternating bands of constriction and dilatation are characteristic of the appearance of lymphatic vessels. Areas of constriction correspond to lymphatic valves [6].

The cisterna chyli receives the right and left lumbar lymphatic trunks. The cisterna chyli lies in front of the first and second lumbar vertebrae behind the crus of the diaphragm. It is a single sac, usually 1–2 cm wide and up to 5 cm long. Despite this classic description, the cisterna chyli has a highly variable appearance. This wide variation of the cisterna chyli has led some authors to prefer the descriptive term "abdominal confluence of the lymphatic trunks" [7].

The size of the cisterna chyli is markedly variable, with cases of very large cisterna chyli frequently called giant cisterna chyli [8, 9] (Fig. 2).

In other cases, aneurysmal dilatation of the retroperitoneal lymphatic system may be more diffuse with dilatation of

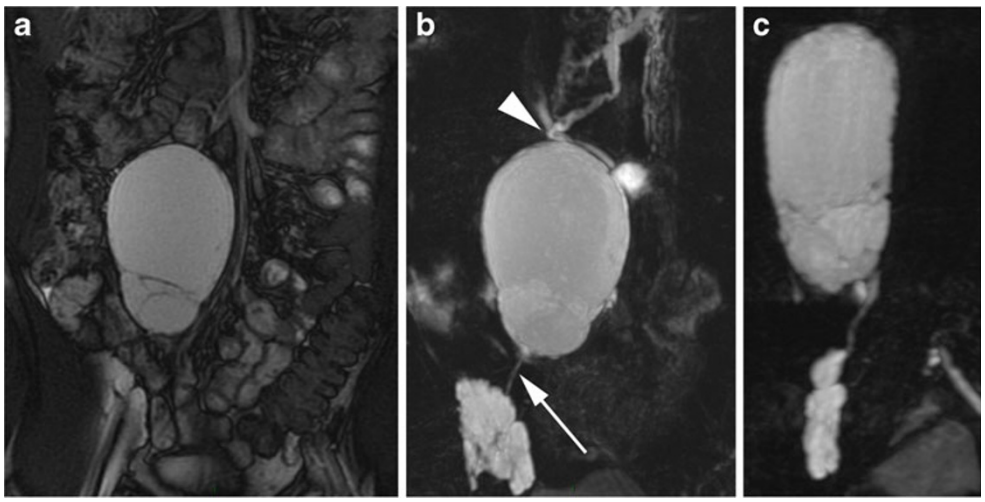

Fig. 10 A 25-year-old man presenting with non-specific abdominal discomfort. Two communicating cystic lesions are demonstrated. The upper lesion is a round bulky cystic lesion with mass effect on the small bowel on T2-weighted MR image (**a**). MR lymphography with coronal MIP image (**b**) demonstrates communication of the upper portion with retroperitoneal lymphatic system (*arrowhead*). The lower position communicates (*arrow*) with the lower lesion which is infiltrative and multiloculated. Communication between the two locations is also well demonstrated on MIP sagittal reconstruction (**c**). Lymphatic nature of the upper lesion and communication with the lower lesion was proved at surgery

cisterna chyli and both right and left retroperitoneal lymphatic trunks (Fig. 2). In other cases, aneurysmal dilatation of right or left retroperitoneal lymph trunk may be observed (Fig. 3). This lymphatic aneurysmal dilatation appears as fluid-filled cystic spaces that may be responsible for a mass effect on adjacent organs (Fig. 3). To prove the lymphatic origin of the unilocular or multilocular tortuous cystic formation, it is essential to demonstrate the communication with retroperitoneal lymphatic vessels (Figs. 1, 2, 3, 4, 5 and 6). Demonstration of communication of cystic lesion with the main lymphatic vessels allows a definitive non-invasive diagnosis of lymphatic malformation.

These lymphatic aneurysmal dilatations are unilateral in most cases, but bilateral location may also be observed (Fig. 4).

The content of cystic dilatation is usually homogeneous, but a fluid-fluid level may be observed related to sedimentation caused by haemorrhagic debris or by fatty content of the formation (Fig. 6). Therefore, a signal intensity of lymphatic aneurysmal dilatation may be variable in the same patient at different MR examinations (Fig. 6).

Retroperitoneal cystic lymphangioma

Lymphangiomas are found predominantly in children, usually in the head and neck region, and retroperitoneal location is uncommon [10].

The aetiology of lymphangioma is poorly understood but is believed to be a developmental abnormality characterised by failure of lymphatic tissue to establish normal communication with regional lymphatic vessels, resulting in dilatation of the abnormal channels [11].

Histopathologically, lymphangioma is characterised by thin-walled unilocular or multilocular cysts that are lined by a single endothelial layer and contain clear or milky fluid. The wall contains lymphatic tissue, small lymphatic spaces and smooth muscles [12].

Lymphangiomas are classified as capillary, cavernous or cystic depending on the size of the lymphatic spaces.

Retroperitoneal lymphangioma is mainly the cystic form, composed of large macroscopic lymphatic spaces. It usually appears as a cystic multiseptated mass with thin walls.

Clinical presentation is variable and may be misleading. Although it may be diagnosed incidentally on ultrasonography or computed tomography (CT) scan, chronic non-specific symptoms are relatively common. With MR lymphography, retroperitoneal cystic lymphangioma appears usually as a multilocular mass. These fluid-filled cystic lesions may present as infiltrative lesions with concave margins or bulky round lesions with a mass effect on adjacent organs (Figs. 7, 8). An elongated shape and the involvement of multiple compartments of the retroperitoneum are characteristic features of lymphangioma (Fig. 9). Most lesions are fluid-filled masses with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig. 10). Sometimes, there are some flow phenomena with signal heterogeneity on T2-weighted images in very large lesions (Figs. 7, 8).

In summary, we believe that 3D MR lymphography demonstrating a communication of the lymphatic aneurysmal dilatation with main lymphatic vessels suggests a continuous spectrum of change from normal variants to lymphatic aneurysmal dilatation. However, in the so-called cystic lymphangioma, lack of communication with lymphatic channels may be observed.

Conflicts of interest The authors declare no conflicts of interest. No funding was received for this work.

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