


## ORIGINAL RESEARCH

# MR-lymphangiography identifies lymphatic pathologies in patients with idiopathic recurrent cervical swelling

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## Abstract

**Background:** Idiopathic recurrent cervical swelling may be caused by lymphatic abnormalities.

**Methods:** Ten patients (9 females, mean age  $51.2 \pm 7$ ) with idiopathic recurrent cervical swelling underwent MR-lymphangiography (MRL). MR-lymphangiograms were evaluated regarding lymphatic anatomy and flow. Individualized treatment was recommended according to MRL-findings.

**Results:** 8/10 patients presented with left-sided, 2/10 with right-sided swelling. Pathological lymph-flow was identified in all cases: thoracic duct dilatation in patients with left-sided and right lymphatic duct dilatation in right-sided swelling, accessory thoracic lymphatics in 7/10 and reflux in 8/10 cases. In two cases, a lymphatic thrombus was identified.

After treatment, symptoms resolved completely in 6/10 cases and partially in 1/10 cases. The remaining three patients have intermittent swellings but have no treatment wish.

**Conclusion:** Idiopathic recurrent cervical swelling can be caused by lymphatic anomalies. MRL displays impaired lymphatic drainage, lymphatic vessel dilatation, and chylo-lymphatic reflux as hallmarks of this condition and may aid in targeted treatment planning.

## KEYWORDS

cervical swelling, chylothorax, lymphangiography, lymphedema, magnetic resonance imaging

## 1 | INTRODUCTION

Permanent swelling of the neck is a common finding caused by a variety of diseases. In contrast, recurrent or intermittent cervical swelling—sometimes referred to as recurrent cervical swelling syndrome (RCS)—is a

rare condition.<sup>1–8</sup> Patients are usually concerned because of sudden onset and the inexplicable occurrence of symptoms.<sup>4</sup>

The etiology of RCS is poorly understood and most of the cases were thought to be idiopathic. Recently, however, recurrent swelling episodes have been linked to lymphatic disorders such as lymph

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vessel obstruction.<sup>3,4</sup> So in cases with a typical history, the diagnostic work-up should focus on the lymphatic system to detect anomalies that may predispose to ruptures of lymph vessels with leakage or pathological reflux of lymph. However, diagnosis of lymphatic flow pathologies is challenging.<sup>9</sup>

The imaging work-up of the lymphatic system has been significantly improved with the recent introduction of MR-lymphangiography (MRL).<sup>9</sup> Hence, MRL may be a useful tool to visualize an underlying lymphatic pathology in patients with RCS.

In the following, we want to report our diagnostic and therapeutic experience with RCS to further elucidate this rare condition.

## 2 | METHODS

### 2.1 | Patient inclusion

#### 2.1.1 | Patients were included into the study when they

- Had a history of at least two episodes of cervical swelling, and other causes for cervical swelling like tumors, lymph node enlargement, etc. had been excluded and
- Had undergone MRL for diagnostic work-up of the swelling at our institution.

Retrospective data analysis was approved by the local institutional review board of the Medical Faculty of the Rheinische Friedrich-Wilhelms-University Bonn with a waiver for written informed patient consent for data analysis.

### 2.2 | Clinical history and examination

Side and duration of cervical swelling as well as the time interval between swelling attacks were recorded; particular attention was given to predisposing/causative factors like increased abdominal/thoracic pressure (e.g., Valsalva maneuver). Prior diseases and surgeries as well as radiation therapies were recorded.

### 2.3 | MR Imaging

MRL was performed as part of our standard clinical work-up of patients with suspected lymphatic vascular disease. All patients underwent contrast-enhanced MRL on a 1.5-Tesla MR-system (Ingenia, Philips, Best, The Netherlands) with lymphatic contrast-medium application to visualize the central lymphatic system. Patients were informed about off-label contrast-agent use and gave their written informed consent for MRI.

The examination protocol included axial fat-suppressed T2-weighted and coronal dynamic post-contrast T1-weighted multi-gradient-echo (mDIXON) sequences (repetition time: 5.2 ms, echo

time: 1.8 ms and 4 ms, field of view: 430 mm, matrix: 480 × 480 mm). Gadobutrol (Gadovist, Bayer Vital, Leverkusen, Germany) was used as a contrast agent in a 1:2 dilution with physiological saline.

Two types of MR-lymphangiographic examinations were performed:

- Central MRL (either transpedal [interdigital] interstitial or nodal contrast agent application)
- Peripheral MRL of the right arm (manual [interdigital] interstitial contrast agent application)

Central nodal MRL was performed as described before.<sup>9</sup> In short, the patient was placed in supine position and noncontrast-enhanced sequences were acquired. Then, the patient was transported out of the MRI-room on a detachable table and an inguinal lymph node in each groin was accessed under sonographic guidance with a 25G needle. Thereafter, the patient was brought back into the MR-scanner. During slow injection of the contrast medium solution via the accessed lymph nodes (flow rate of 1 ml/min), T1-weighted mDIXON sequences were acquired with a temporal resolution of ~40 s.

The methodology of transpedal interstitial MRL of the central lymphatics has also been described before.<sup>10,11</sup> One milliliter of diluted contrast agent was injected intracutaneously into each interdigital space of both feet (overall 8 ml of diluted contrast agent). The patient was then asked to ambulate for 5 min to facilitate lymphatic transport of the contrast agent. Imaging was obtained analogous to nodal MRL.

Interstitial MRL of the right arm was performed, if the swelling occurred on the right side, and central MRL did not explain the cervical swelling. MRL of the arm was conducted similarly to transpedal interstitial MRL with injection of 1 ml of diluted contrast agent into all interdigital spaces of the right hand followed by dynamic imaging as described above.

### 2.4 | Image analysis

All images were analyzed in consensus by two radiologists (M.V. and C.C.P with 1 and 10 years of experience with lymphatic imaging) on a standard workstation (Impax, Agfa Healthcare, Mortsel, Belgium). The radiologists were blinded to patients' clinical data, and evaluated MR-lymphangiograms regarding lymphatic anatomy and flow dynamics, as well as abnormalities.

The following imaging parameters were analyzed:

- presence and extent of cervical swelling
- presence of pleural effusions
- side of thoracic duct outlet (left or right)
- thoracic duct morphology (normal anatomy, nonpathological normal variants (e.g., thoracic duct multiplications), (terminally) circumscribed dilatation, general dilatation)
- enhancement of accessory thoracic lymphatic pathways (e.g., additional paravertebral/mediastinal lymph vessels other than

TABLE 1 Patient demographics and clinical presentation at baseline

Patient	Age (years)	Gender	Side of swelling	Duration of symptoms prior to MR-L (months)	Number of swelling episodes	Minimal time between swelling attacks	History of chylothorax during swelling episode	Side of chylothorax	Ascites	Trigger/aggravating factor of swelling	Potentially relevant prior illnesses
1	44	Female	Left	25	10	4 weeks	Yes	Left	No	Physical stress	
2	43	Female	Left	18	>20	2 weeks	Yes	Left and right	Yes	Physical stress	
3	50	Female	Right	10	4	7 weeks	No	-	No	Physical stress	Retrotonsillar abscess on the right
4	47	Female	Left	22	>20	5 days	No	-	No	Physical stress	Persisting thoracic outlet syndrome on the left
5	44	Female	Right	4	5	2 weeks	No	-	No	Physical stress	
6	53	Female	Left	43	12	2 weeks	No	-	No	Physical stress	
7	53	Female	Left	3	10	1 week	Yes	Left and right	No	Physical stress	Breast Cancer on the left
8	52	Female	Left	12	>20	1 week	Yes	Left and right	No	Physical stress	APC resistance, recent left subclavian vein thrombosis
9	57	Female	Left	29	>20	3 weeks	Yes	Left	No	Physical stress	
10	69	Male	Left	3	3	3 weeks	Yes	Right	No	Physical stress	Gamma disease

TABLE 2 Imaging findings in MRL

Patient	MRL technique	Examination during symptoms	Pleural effusion at time of MRL	Side of swelling	Pharynx/Larynx involvement	Side of thoracic duct junction	Thoracic duct morphology	Right lymphatic duct	Accessory thoracic lymphatic pathways	Lymphatic outflow obstruction	Lymphatic reflux
1	Central	Yes	No	Left	Yes	Left	Generally dilated	Generally dilated	Yes	Yes	Neck
2	Central	Yes	No	Left	Yes	Left	Generally dilated	Generally dilated	Yes	Yes	Neck and mediastinum
3	Central + arm	Yes	No	Right	Yes	Left	Normal	Generally dilated	No	Yes	No
4	Central	Yes	No	Left	Yes	Bilateral	Generally dilated	Generally dilated	Yes	Yes	Neck and mediastinum
5	Central + arm	No	No	Right	No	Left	Normal	Normal	No	No	No
Arm	Yes	Yes	No	Right	No	Left	Generally dilated	Generally dilated	No	Yes	Neck
6	Central	Yes	No	Left	Yes	Left	Terminally dilated	Terminally dilated	No	Yes	No
7	Central	Yes	Yes	Left	Yes	Left	Generally dilated	Generally dilated	Yes	Yes	Neck and mediastinum
Central	No	No	No	None	Yes	Left	Normal	Normal	No	No	No
8	Central	Yes	No	Left	Yes	Left	Terminally dilated	Terminally dilated	Yes	Yes	Neck
9	Central	Yes	No	Left	Yes	Left	Terminally dilated	Terminally dilated	Yes	Yes	Mediastinum
10	Central	Yes	Yes	Left	No	Left	Terminally dilated	Terminally dilated	Yes	Yes	Mediastinum

Note: MRL techniques: central (i.e., transpedal or nodal MRL); arm (i.e., interstitial MRL of right arm).

the thoracic duct draining lymph from the abdomen into the thorax/cervical region)

- lymphatic reflux (none, into lymph vessels of the neck, into lymph vessels of the mediastinum, both)
- cervical lymphatic extravasation.

The thoracic duct was considered generally dilated if the diameter of the whole duct was >5 mm; it was terminally dilated if the terminal portion measured more than 10 mm in diameter.<sup>12,13</sup> The diagnosis of thoracic duct outflow obstruction was made if the thoracic duct was dilated and outflow of the contrast agent into the venous system was absent.<sup>14</sup> Lymphatic reflux was defined as reversal of lymph flow away from the thoracic duct and the venous termination.

Overall, pathological lymph flow within the central lymphatic system was defined as outflow obstruction and/or lymphatic reflux.

## 2.5 | Treatment and clinical course

Therapeutic recommendations were made based on the imaging findings of MRL. As a basic measure medium-chain triglyceride (MCT) diet was recommended in patients with pathological lymph flow in an effort to reduce symptoms by reducing lymph flow in central lymphatics.<sup>15</sup> Causal or (minimally)-invasive therapy including interventional recanalization of obstruction was recommended when a target for intervention was identified on imaging. In some cases because of patients' choice/preference, the actual treatment differed from the therapy recommendation (see below).

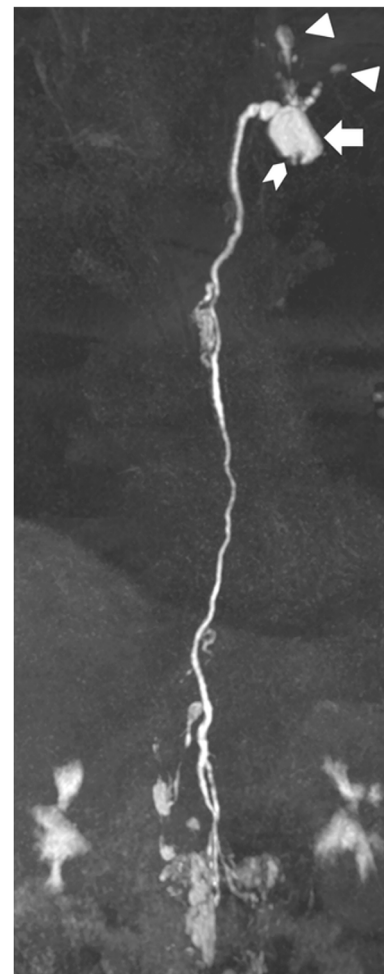
Patients' further clinical course was recorded. Further imaging (sonography, repeated MRL) was performed depending on the clinical course of the patient.

## 3 | RESULTS

### 3.1 | Patient characteristics

Between 08/2016 and 01/2020 10 patients (9 females, 1 male; mean age  $51.2 \pm 7$  [range 43–69] years) were referred to us for diagnostic work-up because of recurrent cervical swelling. Recurrent swellings had been observed for a median time of 15 (range 3–43) months with a median number of swelling attacks of 11 (range 3–over 20). Swelling was left-sided in eight, and right-sided in two cases and was accompanied by dyspnea in all patients.

In all cases, swelling was triggered or aggravated by physical stress, typically sport or yoga. A history of accompanying pleural effusions was present in six patients. Effusions were chylous in all cases, as shown by laboratory examination. None of the patients had accompanying salivary gland disease or autoimmune/inflammatory conditions. Patient demographics, relevant prior illnesses as well as symptoms at baseline are given in Table 1.



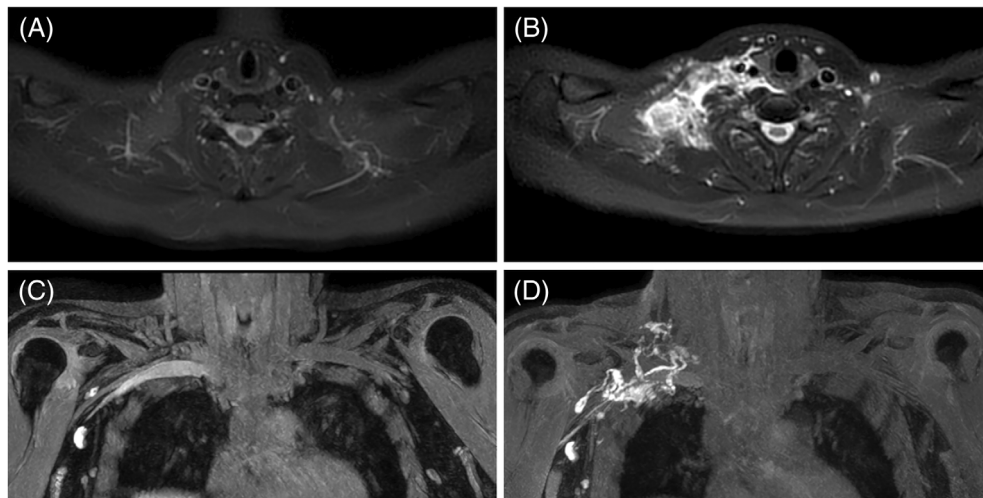
**FIGURE 1** Maximum-intensity projection of a contrast-enhanced, T1-weighted MR-lymphangiogram of a patient with recurrent cervical swelling on the left side. The terminal thoracic duct is dilated (white arrow) and there is lymphatic reflux in the neck on the left side (white arrow heads). There is a small contrast filling defect in the terminal thoracic duct where it flows into the subclavian vein (square arrow head)

### 3.2 | MRL examinations

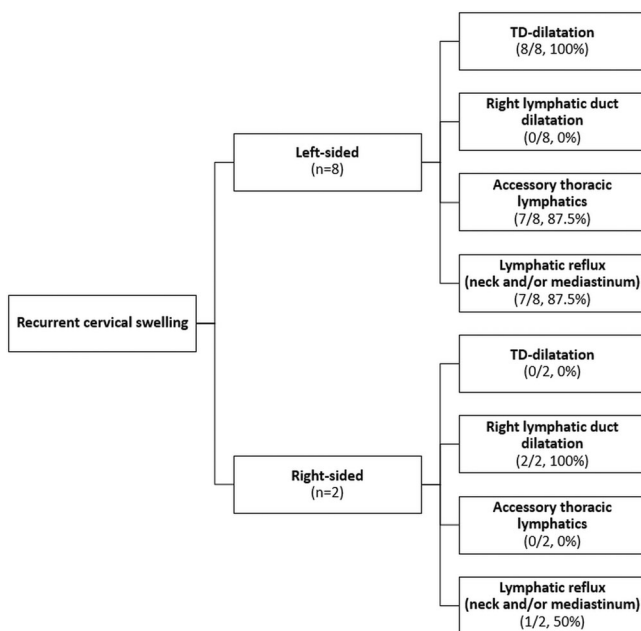
Ten primary MRLs were performed in 10 patients (Table 2). All patients received central MRL (four transpedal and six nodal). Two patients with right-sided swelling additionally received MRL of the right arm during the initial examination.

Nine of ten patients were examined while swelling was still present. One patient with right-sided swelling (#5) was first examined in a symptom-free interval, and a repeat examination was performed later when the patient had symptoms. In one patient (#7), a nodal follow-up MRL was carried out after interventional thoracic duct recanalization.

All MRL examinations were uneventful, that is, no adverse effects were observed during the examination or patient follow-up.



**FIGURE 2** 44-year old female with right-sided recurrent swelling. (A,B) Fat-suppressed, axial T2-weighted images in a symptom-free interval with normal findings (A) and during an acute swelling attack clearly showing edema in the right periclavicular region (B). (C,D) Corresponding contrast-enhanced, T1-weighted, coronal images demonstrating normal enhancement of axillary lymph nodes and venous run-off into the right subclavian vein in the symptom-free interval (C) and dilated right-sided lymphatics and reflux without adequate venous drainage during swelling (D)



**FIGURE 3** Overview of major imaging findings in patients with left- and right-sided recurrent swelling during swelling episodes

### 3.3 | MRL findings

All MRL studies performed during a swelling episode showed interstitial fluid accumulation in the subcutaneous tissue from the mid-cervical area down to the clavicle on the affected side. Swelling was restricted to one side, and did not cross the midline. Edema extended to the larynx or pharynx in 8/10 patients. Of the six patients with a history of proven chylothorax during at least one swelling episode only two had a pleural effusion (one large, one moderate) at the time of the MRL.

The thoracic duct terminated into the subclavian vein on the left side in 9/10 cases and into both subclavian veins in one case. In 8/10 patients (all with left-sided swelling), the thoracic duct was dilated (4/10 general dilatations and 4/10 terminal dilatations; Figure 1). The remaining 2/10 patients had right sided swelling and showed a normal thoracic duct with termination in the left venous angle. In these two patients the right lymphatic duct was dilated as shown by interdigital MRL of the right arm (Figure 2).

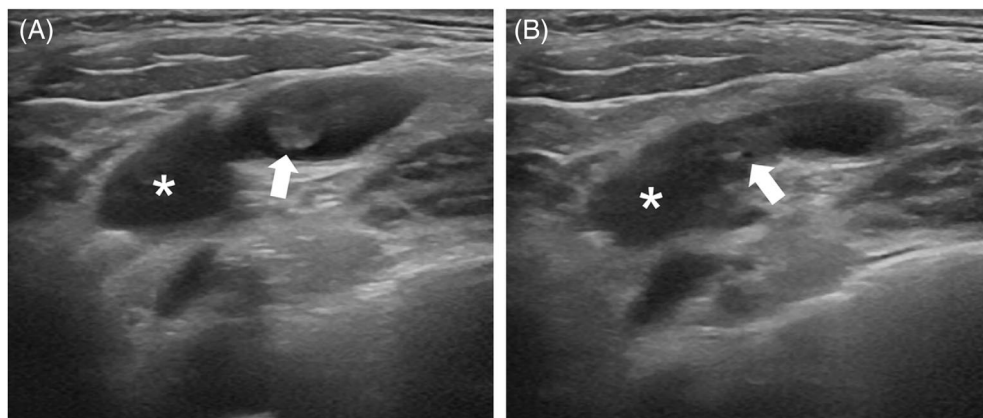
Lymphatic reflux was present in 7/10 cases, and occurred into lymph vessels of the neck (2/10 cases), into lymph vessels of the mediastinum (2/10 cases) or into both (3/10 cases). Enhancement of accessory thoracic lymphatic vessels was seen in 7/10 cases. Frank lymphatic extravasation was not observed.

Patient #5—the only patient without evident lymphatic flow abnormalities in the initial MRL—was first examined in a symptom free interval. A second MRL was done when the patient had a cervical swelling attack, and then demonstrated dilated right-sided periclavicular lymphatics with reflux into lymphatics of the neck (Figure 2).

Overall, pathological lymph flow within the central lymphatic system—defined as outflow obstruction and/or lymphatic reflux—was observed in all cases (eight left and two right). Individual imaging findings are summarized in Table 2 and Figure 3. In 2/10 cases, a thrombus within the terminal thoracic duct could be identified as the cause of lymphatic outflow obstruction (Figure 4).

### 3.4 | Correlation with prior illnesses

In 4/10 patients, a relationship between swelling and prior illness seemed plausible. One patient with right-sided swelling had a history of retrotonsillar abscess with respective surgery on the right side. One patient with left-sided swelling had bilateral thoracic outlet syndrome with resection of



**FIGURE 4** Ultrasound images of the same patient as in Figure 1 with left cervical swelling. There is a slightly hyperechogenic mass/thrombus in the distal thoracic duct (white arrow, star = left subclavian vein). The thrombus intermittently impeded the transvalvular lymph flow between the terminal thoracic duct and the left subclavian vein (B, white arrow, star = left subclavian vein). The patient had a history of recent left subclavian vein thrombosis.

**TABLE 3** Suggested and performed therapy and clinical presentation at follow-up (MCT)

Patient	Recommended therapy after MRL	Actually performed therapy	Symptoms at follow-up	Follow-up duration (months)
1	MCT-diet	None (MCT-diet was perceived as too stressful)	Continued cervical swelling episodes	43.2
2	MCT-diet	None (no treatment wish)	Continued cervical swelling episodes	45.0
3	MCT-diet	None (no treatment wish)	No further symptoms	29.4
4	MCT-diet	None (MCT-diet was perceived as too stressful)	Continued cervical swelling episodes	35.0
5	MCT-diet	MCT-diet	No further symptoms	34.3
6	MCT-diet	MCT-diet	No further symptoms	20.5
7	Thrombus fragmentation	Thrombus fragmentation	No further symptoms	21.1
8	Continuation of heparin treatment, thrombus fragmentation	Continuation of heparin treatment	No further symptoms	8.4
9	MCT-diet	MCT-diet	Less frequent and lighter cervical swelling with MCT-diet	9.0
10	MCT-diet	MCT-diet	No further cervical swelling	10.5

the first rib on the right and persisting thoracic outlet syndrome on the left. Of the two patients with a lymphatic thrombus, one had a history of recent left subclavian vein thrombosis immediately before symptom onset. Venous thrombosis had completely resolved under heparin therapy, but a fresh thrombus in the terminal thoracic duct persisted. The other patient with lymphatic thrombosis had a history of left-sided radiation therapy because of breast cancer several years before symptom onset without recent venous thrombotic events.

### 3.5 | Treatment

Treatment recommendations and the actually performed treatments are listed in Table 3. In the patient with a fresh thrombus in the terminal thoracic duct continuation of heparin and thrombus fragmentation was recommended. Symptoms resolved on continued heparin

3 weeks after MRL. The patient with lymphatic outflow obstruction due to a consolidated lymphatic thrombus required interventional recanalization of the terminal thoracic duct by thrombus fragmentation. This was performed by direct ultrasound-guided puncture of the cervical thoracic duct using an 18G needle with subsequent mechanical fragmentation of the thrombus, which was technically successful. In the remaining 8/10 cases, MCT diet was recommended, but only four patients followed this recommendation. Two patients perceived MCT-diet as too stressful in daily life; another two had no further desire for treatment in view of the clarified diagnosis.

### 3.6 | Follow-up

Clinical follow-up was available for all patients. Follow-up time was  $25.6 \pm 13.8$  months (range 8–45 months). On follow-up, symptoms



resolved completely in 6/10 cases (1 under continued heparin in a patient with fresh thrombosis, 1 after thrombus fragmentation, 3 with MCT-diet, 1 spontaneously without therapy); in another patient symptoms resolved partially with MCT-diet (swelling less frequent and lighter). The remaining 3/10 patients without therapy still have intermittent swellings.

To evaluate the therapeutic effect of thrombus fragmentation, patient #7 underwent MRL after lymphatic thrombus fragmentation, showing normalization of thoracic duct caliber, and disappearance of formerly visible accessory lymphatic pathways.

## 4 | DISCUSSION

RCS is a rare clinical entity with only 12 cases reported in the literature to date.<sup>1-8</sup> The disease preferentially affects females (9/10 in our cohort; 10/12 in literature), which suggests a possible contributing hormonal component.<sup>16</sup> Also, it seems to be a disorder of adult age, with our youngest patient being 43 years, and the youngest patient in the literature being 33 years of age.

The typical clinical presentation is a sudden painless swelling extending from above the clavicles up into the neck caused by soft-tissue edema. This occurs or is aggravated in the context of physical stress, minor trauma or strain in the head/neck region (e.g., during sport, yoga), as also seen in our patients.<sup>3,4</sup> Prior local disease, surgery or radiation therapy may be predisposing like other conditions that obstruct the normal lymph flow (e.g., thoracic outlet syndrome). The swelling subsides over time, but recurs with symptom-free intervals between 5 days and 7 weeks in our patients.

Since the thoracic duct drains the largest amount of lymph of the body and typically terminates into the left jugulo-venous angle,<sup>17</sup> cervical swelling linked to lymphatic flow pathologies occurs mostly on the left side.

In most patients, the swelling is accompanied by dyspnea, which besides the sudden and inexplicable onset, is usually the patients' main concern.<sup>4</sup> Whether dyspnea is due to the space occupying effect of the edema, due to larynx involvement, or results from accompanying pleural effusion (often presumed to be chylothorax) can only be speculated. Although pleural effusions have been described to accompany a swelling attack in a number of cases,<sup>1,3</sup> our imaging results demonstrate, that effusions are not necessarily present during every swelling episode.

The diagnosis of RCS is based upon the typical history and is usually made by exclusion of other forms of temporary cervical swelling, including venous causes or angioedema.<sup>18</sup> The pathophysiology of RCS has been poorly understood. Underlying lymphatic vascular pathologies have recently been suggested,<sup>3,4</sup> which is supported by our findings.

The main pillar of our diagnostic work-up was MR lymphangiography, which has increasingly become a focus of diagnostic attention over the last years.<sup>9,11,19</sup> Especially, contrast-enhanced MRL has shown its capability to reliably demonstrate central lymphatic anomalies, and to provide functional information.

In all of our patients with the clinical diagnosis of RCS, MRL identified a lymphatic flow anomaly, usually a combination of lymphatic obstruction and chylolymphatic reflux, which was seen in 8/10 patients. In the remaining two cases, MRL revealed an obstruction without reflux, which may be a lesser manifestation of the same disease spectrum. The exact cause of outflow obstruction could only be determined in two cases where a thrombus could be unequivocally identified. In the other patients no clear cause for obstruction was seen. Prior illnesses or therapies (such as radiation therapy) or even underlying congenital lymphatic anomalies may play a role.<sup>20</sup>

A follow-up MRL after successful thrombus removal in one patient showed that the flow obstacle had been removed, and accessory lymphatics were no longer delineated, suggesting that accessory lymphatics are likely lymphatic collaterals that present in times of altered endolymphatic pressures.

That MR findings may directly correlate with the clinical findings/swelling episodes was also observed in one patient with normal MRL in the symptom-free interval, but clear signs of obstruction and reflux during a swelling episode. We therefore advocate to perform MRL evaluation during the acute stage of the disease.

Although the pathophysiology of RCS has not been elucidated in all details, lymphatic drainage anomalies seem to be of fundamental importance. This provides targeted therapeutic approaches. Both of our patients with obvious obstruction benefitted from thrombus removal with no further swelling episodes during follow-up periods of 8 and 21 months.

When MRL shows only signs of a functional obstruction, triggering movements should be avoided. Furthermore, it seems prudent to reduce the lymph flow by means of an MCT-diet<sup>15</sup> when the swelling is on the side of thoracic duct termination. All of our patients placed on MCT-diet, were symptom-free ( $n = 3$ ) or had less frequent and less pronounced swelling episodes ( $n = 1$ ), while undergoing the MCT-diet. More invasive measures, like lymphovenous anastomosis<sup>1</sup> or interventional embolization of pathological lymph vessels<sup>5,21</sup> have been described anecdotally. As lymph vessel occlusion may even worsen lymphatic flow pathologies these more invasive treatment options should be reserved as a last resort only, as RCS in general is a rather benign and not life threatening condition. This also explains that—after the disease was explained to the patients—some refused any therapeutic measures as these are perceived as burdensome. Although MCT-diet is a noninvasive treatment option, the treating clinician should keep this in mind when recommending treatment of cervical swelling as it will have an impact on treatment adherence of the patients. The treatment strategy should therefore be discussed with the patient depending on the individual perception of the burden of the condition on patients' life. It is important to note in this context that so far the necessary duration of dietary treatment has not been established as the patients in our cohort continued the diet after resolution of symptoms. Furthermore, other treatment options that are possibly less burdensome for the patients, including local compression, drainage therapy or medical treatment (e.g., octreotide) should be explored.

Of note, one of the patients refusing treatment showed no further swelling episodes after MRL over a follow-up time of 2.5 years without any treatment. RCS may therefore also spontaneously resolve, possibly due to spontaneous recanalization of lymphatic outflow.

The present study has several limitations. With 10 patients our cohort is rather small; however, it is the largest cohort of consecutive patients with RCS reported so far. Due to the retrospective character of this analysis, different types of contrast-enhanced MRL (nodal and transpedal contrast injection) were performed. However, both techniques are able to visualize the central lymphatic system and associated pathological conditions.<sup>9</sup> Follow-up imaging was only available in one patient as there was no clinical indication for further imaging. It would, however, certainly be interesting to investigate how different therapeutic approaches change lymphatic flow in this condition. In this respect, local manual lymphatic drainage or local compression therapy may be pursued as a therapeutic approach in the future not performed in our cohort. Due to considerable anatomical variation of the lymphatic system it is difficult to distinguish normal variants from abnormalities. As four patients chose not to adhere to the recommended treatment and have continued symptoms, the connection between the described imaging findings of the lymphatic system and cervical swelling cannot be proven with certainty in these patients. However, more invasive catheter angiography was not indicated and MRL-findings are consistent with patients who responded to therapy, strongly suggesting the causative role of the lymphatic system in RCS. Nonetheless, further research into lymphatic flow dynamics in larger patient cohorts is warranted in the future to prove the connection between lymph flow anomalies and cervical swelling suggested by this case series.

In conclusion, recurrent cervical swelling seems to be caused by lymphatic flow anomalies. MRL can demonstrate impaired lymphatic drainage, lymphatic vessel dilatation, and chylolymphatic reflux as hallmarks of this condition. As treatment options depend on the results of MRL, this should be performed in all patients presenting with RCS, preferably in centers with experience in lymphatic imaging.

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