

## CASE REPORT

## INTERMEDIATE

## CLINICAL CASE

# To Be or Not To Be

## A Case of Recurrent Swelling Syndrome of Thoracic Duct



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## ABSTRACT

Thoracic duct aneurysm is a rare entity presenting as a stable, asymptomatic, left supraclavicular swelling. We report an unusual case of a thoracic duct aneurysm in a 71-year-old woman presenting as a recurrent swelling syndrome of the left supraclavicular area associated with sporadic episodes of sharp left subcostal pain. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1070-3) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

The patient was a 71-year-old woman with episodic left lower chest pain and associated left supraclavicular swelling for 6 years. She had episodes of sharp pain in the left subcostal area lasting 2 to 6 h. The pain was soon after associated with a swelling in the left supraclavicular area. This occurred over a duration of 6 years and increased in frequency. She reported no pain or tenderness in the neck area. The subcostal pain was almost always associated with this inexplicable neck swelling, which disappears shortly after the resolution of the pain. No inciting factors were recognized. There was no history of chest trauma or surgery prior to this occurring.

On physical examination, her neck was supple. There were no carotid bruits and carotid upstrokes were normal bilaterally. There was a slight fullness noted in the left supraclavicular area without tenderness (**Figure 1**). Her chest was clear to auscultation with normal heart sounds. Her abdomen was soft without tenderness or masses. Her upper extremities were warm and peripheral pulses were normal bilaterally.

## PAST MEDICAL HISTORY

She has a past surgical history of bladder prolapse repair but is otherwise healthy.

## INVESTIGATIONS

The blood counts and biochemical tests did not reveal any pathological findings. Ultrasonography of the soft tissues of the left side of her neck performed during the acute phase of one of her spells demonstrated thin-walled tubular structures with no internal perfusion on color-coded Doppler (**Figure 2**).

## LEARNING OBJECTIVES

- To recognize the atypical and dynamic presentation of a TD cyst.
- To understand the diagnosis and management of recurrent swelling syndrome of the TD.

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Serial computed tomography scans of the chest showed a small, smooth ovoid fluid density structure interposed between the descending thoracic aorta and the thoracic spine at the T5 vertebra initially measuring  $1.1 \times 0.7$  cm increasing to  $2.1 \times 1.2$  cm over 1 year. Similar findings were noted on magnetic resonance imaging.

### MANAGEMENT

Due to suspicion of thoracic duct (TD) origin, lymphangiography was performed. A sonographic survey was performed of the left neck and supraclavicular area prior to the procedure for localization of the previously seen dilated tubular structures. Following the administration of local anesthesia, a puncture was performed of the tubular structure in the left neck using a micropuncture needle under ultrasound guidance. Serial injections of contrast with pressure were performed into this structure, some with digital subtraction angiography. The terminal TD was noted to be aneurysmal and lobulated over a 2.2-cm segment measuring up to 10 mm diameter (Figure 3). There was a high-grade, irregular stenosis of the TD approximately 3 mm proximal to its anastomosis with the left subclavian vein (Video 1). Reflux was seen up the left internal jugular vein. A few lymphatic collaterals proximal to the stenosis were noted. The TD was also dilated over its entire length measuring approximately 3.5 mm in diameter (Figure 4). The cisterna chyli was opacified located in the midline spanning from mid-T<sub>12</sub> through mid-L<sub>1</sub> vertebrae level measuring  $2.9 \times 1.5$  cm. There was transient filling of some outpouching and branching structures arising from the lower TD around the level of the mid-T<sub>10</sub> to mid-T<sub>11</sub> vertebrae. Contrary to imaging studies, there was no filling of any focal dilated lymphatic structures at the T<sub>5</sub> level. The inner dilator of the micropuncture kit was then inserted using Seldinger technique. Several microwires were made to cross the stenosis in the TD with some success (Figure 5, Video 2). The catheter was removed and a sterile dressing was placed. The patient tolerated the procedure well. There were no post-operative complications.

### FOLLOW-UP

Complete resolution of her symptoms was noted post-procedure at 1-year follow-up.

### DIFFERENTIAL DIAGNOSIS

Thyroid lesions are unusual in the supraclavicular area, but metastatic papillary carcinoma of the

thyroid can present here; however, it is usually heterogenous and less likely cystic (1). Thymic cyst, parathyroid cyst, lipoma, cystic hygroma, and branchial cleft cyst can present as a left supraclavicular mass. Less commonly, TD cysts present as a stable, supraclavicular mass.

### DISCUSSION

The TD originates from the cisterna chyli in the abdomen, penetrates the diaphragm at the aortic aperture and travels through the posterior and superior mediastinum serving as a channel for lymph to drain into the venous system. Cystic dilations are rare and may occur anywhere along the course of the duct (1). Furthermore, the terms “thoracic duct cyst” and “thoracic duct aneurysm” have been used interchangeably in the published data. Suzuki et al. (2) described a TD cyst as an aneurysmal dilation of the TD. Livermore et al. (3) reported a TD aneurysm as a cyst in the left supraclavicular area. Review of the published data reports 30 cases of TD cysts, the majority presenting as an asymptomatic stable swelling. Cases of cervical TD cysts have been reported with symptoms of pain, dyspnea, dysphagia, and hoarseness related to compression of neck structures (4). Wan et al. (1) reported a case of large abdominal TD

### ABBREVIATIONS AND ACRONYMS

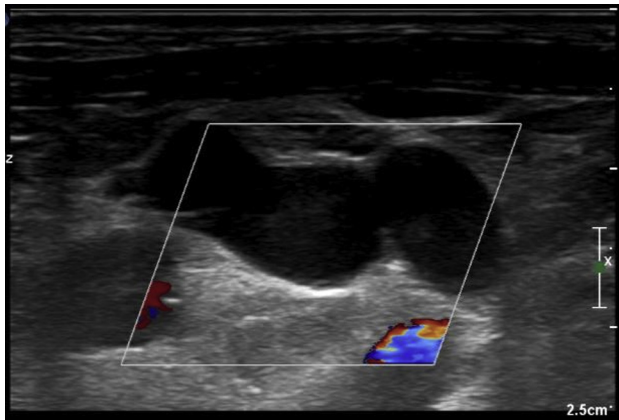
FNA = fine needle aspiration

TD = thoracic duct

FIGURE 1 Thoracic Duct Aneurysm



Left supraclavicular fullness.

**FIGURE 2** Ultrasonography

Multiple cysts with no color Doppler flow.

**FIGURE 3** Terminal Stenosis

Lymphangiogram showing 2.2 × 1 cm dilatation of the thoracic duct with terminal stenosis (arrow).

cyst presenting with abdominal pain managed by surgical resection. Veziant et al. (7) reported a case of recurrent swelling of the terminal TD with spontaneous bilateral chylothorax and chyloperitoneum triggered by a fat-rich diet. The left radicular pain in our case appeared to be related to a lymphocele that enlarges when the patient is experiencing acute lymphatic obstruction with impingement of an adjacent nerve.

Congenital weakness of the duct and acquired degeneration of the TD wall due to inflammation or atherosclerosis have been indicated as etiological factors. There have been reports of cervical TD cysts developing after blunt or whiplash injury to the neck (4). No triggers were found in our patient. The recurrent swelling may reflect the variable flow of chyle and dynamic obstruction of the terminal TD related to stenosis and debris collection.

Both computed tomography and magnetic resonance imaging demonstrate the cystic nature of the lesion along with anatomic location and connections. The addition of lipiodol has been attempted in a few cases to increase specificity by enhancing visualization of the TD (5). Ultrasonography has been suggested for primary diagnosis (6). Fine needle aspiration (FNA) reveals a milky fluid with a high concentration of triglycerides. Due to the ambiguous nature of the cyst and pain syndrome, FNA was deferred to avoid provoking iatrogenic infection, inflammation, or thrombosis of vital neck structures. Lymphangiography helped to confirm the TD origin, delineate the communication of the supraclavicular cyst with the TD, recognize collaterals, and trace cystic areas.

Although the management of stable-sized cysts includes FNA or surgical resection, there is no standard management for recurrent swelling associated with terminal TD occlusion. Two cases of successful cervical lymphovenous anastomosis have been reported (7). In the current case, obstruction of the lymphoid flow could be from a focal stenosis in the terminal portion of TD. This was relieved by dilation and pressure with contrast. It is unclear if this is the absolute treatment of her condition or if her symptoms could recur. No recurrence was noted at 1-year follow-up. We discussed embolization of the TD above the level of cisterna chyli should her symptoms recur. However, reopening the TD stenosis directly could be tried again given the prolonged benefit.

## CONCLUSIONS

TD aneurysm with terminal duct stenosis can present as a recurrent swelling over variable areas along its

**FIGURE 4 Dilated Thoracic Duct**



Origin at the cisterna chyli at T<sub>12</sub>-L<sub>1</sub> level.

**FIGURE 5 Management**



Guidewire passing through the terminal stenosis of thoracic duct into left subclavian vein.

length with associated symptoms due to compression of surrounding structures.

Dilation of the stenotic region was performed under fluoroscopy in this case. Successful surgical

management with TD ligation or lympho-venous anastomosis has also been reported.

Lymphangiography is the diagnostic gold standard.

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**KEY WORDS** chest pain, contrast agent, stenosis, thoracic

**APPENDIX** For supplemental videos, please see the online version of this paper.