

Episode 33

Problem Representation

A 58-year-old man with a history of lower extremity lymphedema and COPD presented with subacute dyspnea, found to have bilateral chylothoraces and nail dystrophy.

Schemas

The CPSers' schema for pleural effusions utilizes Light's criteria (fluid protein/serum protein ratio > 0.5; fluid/serum LDH ratio > 0.6; pleural fluid LDH > 200 IU or >2/3 of the upper limit of normal) to distinguish between transudative and exudative effusions.

We also included a schema for lymphedema that distinguishes between primary (due to lymphatic hypoplasia) and the far more common secondary (due to lymphatic obstruction/insufficiency) causes of lymphedema.

Diagnosis

Based on the findings of nail dystrophy, lymphedema, and chylothorax, the patient was found to meet diagnostic criteria for yellow-nail syndrome, a rare cause of primary lymphedema and pulmonary findings!

Teaching points

- A chylothorax¹ occurs when chyle (milky fluid containing lymph and fats transported by chylomicrons) accumulates in the pleural space. This most commonly occurs due to disruption of the thoracic duct secondary to trauma during surgery or as the result of malignant compression. The diagnosis is supported by finding elevated triglyceride levels (i.e. > 110 mg/dL) in the pleural fluid.
- Lymphedema² refers to the accumulation of protein-rich interstitial fluid that occurs due to impaired lymphatic drainage in the setting of normal capillary function. The most common cause of secondary lymphedema worldwide is lymphatic filariasis³ (infection with *Wuchereria bancrofti* or *Brugia malayi*), while malignancy/cancer treatment and cellulitis predominate in the developed world. Complications of lymphedema include recurrent skin infections (due to regional immune dysfunction), cutaneous ulceration, and elephantiasis nostras verrucosa (hyperkeratotic skin changes).
- Yellow-nail syndrome⁴ (YNS) is a rare syndrome (~1/1,000,000 people) characterized by dystrophic/discolored nails, pulmonary abnormalities, and primary lymphedema. While the etiology of this syndrome is unknown, its pathogenesis is thought to largely be related to impaired lymphatic flow. Respiratory manifestations include cough, pleural effusions (including chylothoraces), bronchiectasis, and recurrent sino-pulmonary infections.

Clinical Reasoning Pearl

As learners, we will inevitably encounter clinical scenarios with which we are unfamiliar. In those instances, our reasoning should then focus on how to best *describe* the problem, so that someone familiar with the diagnosis (or our search engine) may immediately recognize it.

For example:

Reza ultimately turned to google to help him solve the case – but not before distilling down what he believed to be the essential descriptive elements of the patient's presentation.

References

1. McGrath EE, Blades Z, Anderson PB. Chylothorax: aetiology, diagnosis and therapeutic options. *Respir Med.* 2010 Jan;104(1):1-8.
2. Taylor MJ, Hoerauf A, Bockarie M. Lymphatic filariasis and onchocerciasis. *Lancet.* 2010 Oct 2;376(9747):1175-85.
3. Grada AA, Phillips TJ. Lymphedema: Pathophysiology and clinical manifestations. *J Am Acad Dermatol.* 2017 Dec;77(6):1009-1020.
4. Vignes S, Baran R. Yellow nail syndrome: a review. *Orphanet J Rare Dis.* 2017 Feb 27;12(1):42.