- **Answer:**
  d) Generalized lymphangiomatosis(GLA)

- **Findings:**

- Computed tomography (CT) of the chest and abdomen and pelvis revealed confluent cystic mediastinal masses enveloping mediastinal structures without compression effect. lungs parenchyma showed interlobular septal thickening, prominent peribronchovascular interstitium and multifocal ground glass opacities.
- Dilated lymphatic vessels were identified in the retroperitoneum.
- A chest tube was placed for pleural effusions and the laboratory study of the pleural fluid was consistent with chylothorax with elevated lactate dehydrogenase and triglyceride and chylomicrons. The patient underwent a CT-guided biopsy of mediastinal lymph nodes which was in favor of reactive lymphadenopathy without evidence of malignancy.
- Multiple lucent lesions with trabecular appearance were present throughout the spine on CT which were confirmed on magnetic resonance imaging. A Technetium-99 whole body bone scan demonstrated multiple foci of abnormal increased radiotracer uptake throughout the spine, ribs and pelvic girdles. An echocardiography was performed which was unremarkable.
- The characteristic imaging findings and laboratory tests are suggestive of GLA with a possible superimposed pulmonary infection.

- **Discussion:**

- Generalized lymphangiomatosis (GLA) is a rare lymphatic abnormality, mostly affects children and young individuals with an equal gender prevalence. It can be a diagnostic challenge because of broad spectrum of manifestations and variety of organs involved. The systemic manifestations are wide and may include chylothorax, pleural and pericardial effusion, lung infiltration, mediastinal soft tissue and cystic masses, lytic bone lesions, abdominal organs and mesenteric involvement, disseminated intravascular coagulation and skin lesions, and cervical lymphadenopathy.

- The mediastinal soft tissue masses may be suspicious for lymphoma; although cystic appearance of the lesions without displacement of adjacent structures and negative lymph node biopsy would favor GLA. The differential diagnoses for osseous lesions include Gorham-Stout disease and hemophagocytic lymphohistiocytosis (HLH). The osseous lesions in GLA are well defined and centered in the medullary space while Gorham-Stout disease may result in progressive osseous destruction known as "disappearing bone disease" and LHL lesions are usually ill-defined and associated with bone marrow edema, periosteal reaction and enhancing soft tissues.

- The Erdheim Chester disease is characterized by proliferation of lipid-containing foamy histiocytes in the skeleton and other organs; the absence of sclerotic changes in the diaphysis and metaphysis of long bones may exclude this diagnosis.

- **References:**

- Putta T., Irodi A., Thangakunam B., Oliver A., Gunasingam R. Young patient with generalized lymphangiomatosis: differentiating the differential. Indian J Radiol Imaging. 2016;26:411–415.

- Faber D.L., Galili R., Nitzan O., Sharoni E. Systemic generalized lymphangiomatosis: a diagnostic challenge. Isr Med Assoc J. 2015;17:785–786.
- Arda K.N., Akay S., Kizilkanat K.T. Generalized cystic lymphangiomatosis incidentally recognized in an asymptomatic adult: peroperative CT, MRI, and histopathological findings of a very rare case. Niger J Clin Pract. 2019;22:1778–1780.
- Yu W et al. Diffuse pulmonary lymphangiomatosis: a rare case report in an adult. Medicine (Baltimore) 2019;98:e17349.

“Case courtesy of Mersad Mehrnahad , MD, Assistant professor of Radiology, Qom University of Medical Sciences.”