



Visual Case Discussion

Cardiac tamponade in a patient with Gorham-Stout disease

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A 22 year-old woman with a history of Gorham-Stout Disease¹ and recurrent chylothorax presented to the emergency department with 4 days of shortness of breath. No other symptoms.

Exam: Vital signs remarkable for tachycardia to 130. Well appearing with dyspnea upon walking. Cardiopulmonary exam shows regular rate and rhythm and decreased breath sounds at the bases bilaterally.

Point of care echocardiogram revealed a pericardial effusion² with

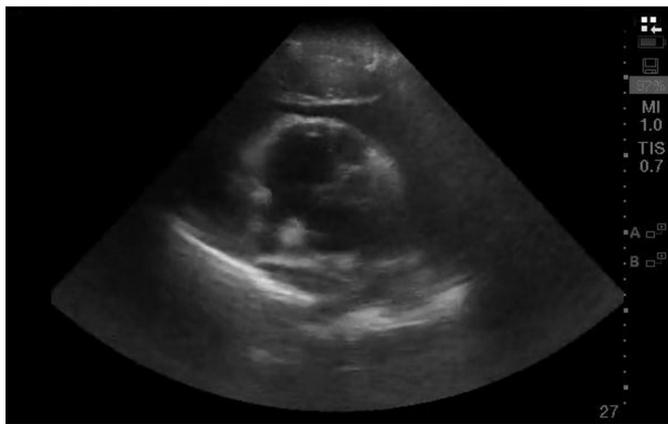


Fig. 1. Bedside transthoracic echo in a parasternal long view showing pericardial effusion with right ventricular collapse.

tamponade physiology, including right ventricular collapse shown in Fig. 1. Cardiomegaly was seen on the chest x-ray shown in Fig. 2.

The patient was admitted to the cardiology service for an urgent

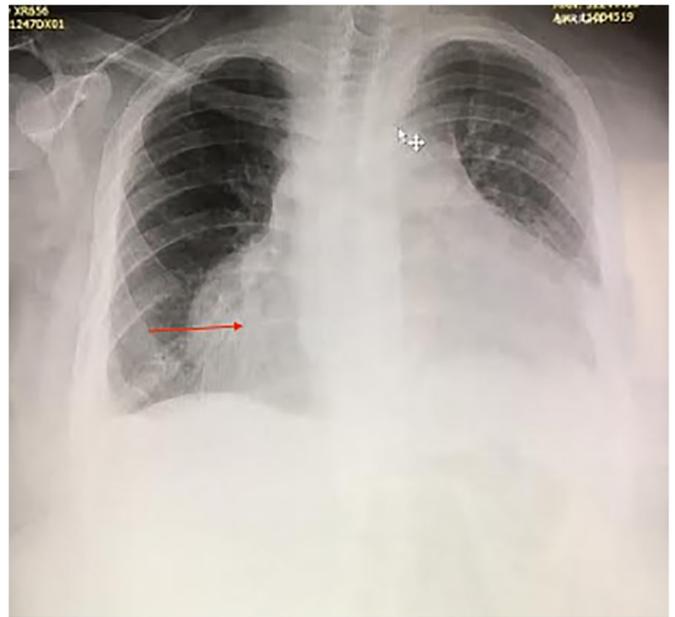


Fig. 2. Chest x-ray before pericardiocentesis showing globe-shaped cardiomegaly and clear lungs.

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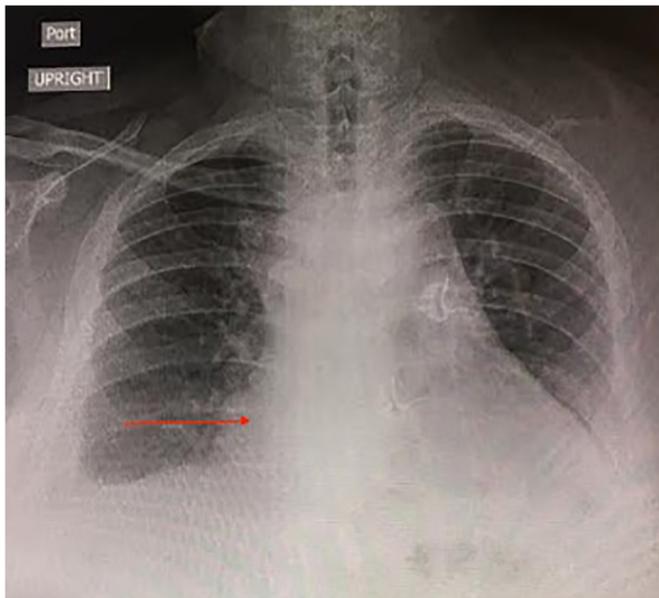


Fig. 3. Chest x-ray after pericardiocentesis showing significant decrease in size of cardiomegaly.

pericardiocentesis draining 2.3 l of fluid. A repeat echo revealed resolution of the effusion. Repeat chest x-ray is shown in **Fig. 3**.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.visj.2018.10.004](https://doi.org/10.1016/j.visj.2018.10.004).

References

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Questions

1. Which of the following is the most common and specific finding to confirm tamponade physiology on an echo?
 - a. Pericardial effusion
 - b. Increased IVC collapse
 - c. Right atrial collapse
 - d. Right ventricle collapse
 - e. Flow velocity paradoxus
2. Which of the following best characterizes Gorham-Stout Disease?
 - a. Benign disorder of enlarged and painful lymph nodes
 - b. Abnormal growth of smooth muscle cells in lungs, lymph, and kidneys.
 - c. Proliferation of abnormal lymphatic channels leading to progressive osteolysis
 - d. Growth delays, malformation of limbs, abnormalities of craniofacial region
 - e. Congenital disorder of lung with abnormal widening of upper airways

Answers

1. Right ventricle collapse. Explanation: Right atrial collapse (c) might be the earliest indicator of tamponade, but it is less specific to cardiac tamponade. Pericardial effusion (a) is a necessary but not sufficient criteria for tamponade. Increased IVC collapse (b) is incorrect; rather, IVC plethora is seen in cardiac tamponade. Flow velocity paradoxes (e) is exaggerated respiratory variations in transmitral and transtricuspid Doppler flow, which can indicate tamponade physiology. However, this finding requires a higher skill level in ultrasound use and is therefore less common.¹
2. Proliferation of abnormal lymphatic channels leading to progressive osteolysis. Explanation: Gorham-Stout Disease, or vanishing bone disease, is characterized by abnormal lymphatic growth and progressive osteolysis (c). Kikuchi disease, or histolytic necrotizing lymphadenopathy (a), lymphangioleiomyomatosis (b), Roberts syndrome (d), and tracheobronchomegaly, or Mounier-Kuhn syndrome (e) are all incorrect.²