Chylotamponade
An Unusual Manifestation of Gorham-Stout Syndrome
Namal Wijesinghe, MBBS, MD, MRCP, FRACP; Zaw Lin, MBBS, FRCS, FRACS; Michael J. Swarbrick, FRCR, FRCS; Dilesh M. Jogia, MBChB, FRACP, PhD

Gorham-Stout syndrome is a rare disorder that leads to extensive lymphangiomatosis in bones with resorption of adjacent bone matrix. As a result, the disease is also known as massive osteolysis or “vanishing bone disease.” Although the disease primarily occurs in bone, it may also involve adjacent soft tissues. We report a case of Gorham-Stout syndrome presented with chylotamponade as a result of extensive intrathoracic lymphatic hyperproliferation.

History
A 50-year-old woman, who is known to have Gorham-Stout syndrome, presented with progressively worsening dyspnea for 1 week. She was hemodynamically stable and her cardiorespiratory examination was unremarkable except for elevated jugular venous pressure. Chest radiography revealed an enlarged cardiac silhouette and a small left pleural effusion. Transthoracic echocardiography confirmed a large pericardial effusion with evidence of tamponade. Urgent pericardiocentesis was performed, and 1.2 L of milky fluid, identified as chyle on biochemistry, was drained. She became asymptomatic soon after pericardiocentesis; however, pericardial fluid reaccumulated over the next few days. She had been diagnosed as having Gorham-Stout syndrome from the age of 12 years. Massive osteolysis of the mandible developed, which required reconstruction. Osteolysis subsequently recurred in the reconstructed mandible. Also, bone involvement in the skull base had resulted in cerebrospinal fluid rhinorrhea and recurrent meningitis. The disease was in remission until recently.

The magnetic resonance lymphangiogram revealed a narrowed thoracic duct but extensive lymphatic tissue infiltration between the major vessels and visceral structures in the upper mediastinum as well as invasion and apparent breach of the pericardial sac at the most superior aspect of pericardium (Figure 1). It was presumed that the chylous fluid was leaking into the pericardial sac at this point. Radiography of the humeri, scapulae, and thoracic spine did not reveal evidence of bone resorption.

She underwent surgery for thoracic duct ligation, drainage of pericardial fluid, and prophylactic surgical pleurodesis of the right pleura. Left pleurodesis was not done because it was thought that this procedure would make a repeat surgical intervention, such as reexploration of thoracic duct if required, difficult. She was also given intravenous octreotide infusion for 1 week followed by 100 μg octreotide subcutaneously twice daily for 1 more month. She was maintained on a low fat diet with Monogen, an enteral low long-chain triglyceride formula, during this period. MRI performed before surgery revealed abnormal lymphatic tissue invading the mediastinum and a large collection of pericardial fluid (Figure 2). She remained asymptomatic, and a repeat MRI done 6 months later revealed no further soft tissue invasion by lymphatics (Figure 3).

Discussion
Gorham-Stout syndrome was first described in 1955 as a “vanishing bone disease” that associates with intraosseous vascular malformation. However, a case of a 12-year-old boy, who had features of Gorham-Stout syndrome, had been reported long before this description. The cause of this disease is not known. Fewer than 200 cases have been reported.

Involvement of thoracic cavity is common among patients with Gorham-Stout syndrome and carries a poor prognosis. The most common manifestation of thoracic involvement, chylothorax, can occur in 1 in every 5 patients. Chylothorax usually associates with the shoulder girdle and thoracic vertebral osteolysis and results from invasion of the thoracic duct or communication of the dysplastic lymphatics with the pleural cavity. Chylopericardium as a manifestation of Gorham-Stout syndrome is very rare, and only 1 case of chylotamponade and 1 other case of asymptomatic chylopericardium have been reported to date.

MRI was helpful in the diagnosis and follow-up of our patient. A single breath-hold 3D fast-recuperation fast spin-echo (FRFSE) sequence, similar to that used for magnetic resonant cholangiopancreatography, was used for the preoperative lymphangiogram. This heavily T2-weighted sequence demonstrates stationary or slowly moving fluid and showed the thoracic duct in the lower thorax and the abnormal lymphangiomatous tissue in the superior mediastinum. It did not show pericardial effusion due to cardiac-related movement in this fluid. The pericardial fluid was well demonstrated using a coronal steady-state free precession (2D FIESTA, GE Healthcare) sequence, which is relatively motion insensitive. This sequence also demonstrated high signal from the abnor-
mal lymphangiomatous tissue in the superior mediastinum though it did not suppress signal from mediastinal fat as strongly as the 3D FRFSE sequence.

Management of chylothorax and chylopericardium associated with Gorham-Stout syndrome includes drainage of fluid, thoracic duct ligation, minimizing chyle production, and the prevention of lymphatic hyperproliferation. Strategies to minimize chyle production include dietary restriction of fat, replacing long-chain triglycerides in the diet with medium-chain triglycerides, and total parenteral nutrition. Octreotide, a long-acting somatostatin analog, also helps to minimize chyle excretion by acting directly on vascular somatostatin receptors. Lymphatic hyperproliferation can be minimized by external beam radiotherapy, a total dose of 40 to 45 Gy delivered in daily 2-Gy doses. Oral clodronate and interferon-α are also thought to be effective to reduce lymphatic hyperproliferation. Interferon-α was thought to be effective for its antiangiogenic properties.

Our patient responded well to thoracic duct ligation and right-sided pleurodesis. Other conservative management methods including dietary restriction of fat, supplementation of medium-chain triglycerides (Monogen), and octreotide may have contributed to her recovery. She was also seen by a radiation oncologist who recommended deferring radiation therapy on that occasion because she was responding well to management described above. However, this remains an option if there is any evidence of further soft tissue invasion by lymphatics on follow-up MRI.

Disclosures

None.

References


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