CLINICAL VIGNETTE

Cardiac Manifestation of Lymphedema

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A 26-year-old Caucasian female presented for preoperative risk stratification prior to protein lipectomy of her abdomen and lower extremities. The patient's past medical history was significant for congenital malrotation of the anterior abdominal wall and chylothorax repaired at birth with congenital lymphedema diagnosed at 1 year of age. The patient reported the edema initially was responsive to compression stockings and diuretics but over the past several years began to accumulate and harden mainly in her lower abdomen and bilateral lower extremities despite aggressive treatment. She had been hospitalized 6-8 times per year as a child for management of edema, cellulitis, and infection-related complications. She previously sought care at various national and international medical centers without relief of the edema. After researching further, she found a local plastic surgeon that offered a potential solution via an invasive catheter procedure called suction assisted protein lipectomy (SAPL).

She presented to a nearby family practice physician for preoperative risk assessment. An EKG revealed NSR with low voltage and poor R wave progression. She was referred to cardiology for further evaluation due to her abnormal EKG.

Upon cardiac consultation, the patient denied active cardiac symptoms including chest pain, shortness of breath, or palpitations. Her current symptoms did include severe lower extremity edema limiting her ability to flex her legs and thereby her functional capacity. At times, her lower extremities were painful as well. Despite this, her functional capacity was greater than 4 mets. She denied any cardiac risk factors and her Revised Cardiac Index score calculated to 0 equating to a 0.4% risk of major adverse cardiac events associated with her procedure. Her family history was significant for a grandfather who had an MI at the age 75. She denied smoking, alcohol, and drug use.

On presentation, the patient was afebrile with a blood pressure of 109/76 and a pulse of 72 beats per minute. Here respiratory rate was 12 breaths per minute and her oxygen saturation was 97% on room air. Physical exam revealed a regular rate and rhythm with a normal S1/S2. No murmur, rubs, or gallops were auscultated. JVP was normal at 7 cm H2O. Lungs were clear to auscultation. She had 2+ pitting edema of her lower abdomen and lower extremities with noticeable hardening of the subcutaneous tissue.

At this point, the patient was referred for 2D echocardiogram based on her abnormal EKG. The echocardiogram was essentially normal including normal left ventricular systolic/diastolic function and valvular function. The inferior vena cava was normal size and showed adequate respiratory collapse. The left and right filling pressures were within normal range. A moderate size circumferential pericardial effusion without signs of cardiac tamponade was noted.

The patient was cleared for her medical procedure and successfully underwent her procedure 1 week later without complications.

Discussion

The pathophysiology of lymphedema is based on a complex interplay of capillary hemodynamics and transport of interstitial fluid. Accumulation of increasing protein rich fluid in the interstitium causes inflammation and reactive fibrosis of the affected tissue, which overtime can cause hardening of the extremities.

Primary lymphedemas are hereditary and are encompassed by three main classes based on the age of presentation: Congenital or infantile hereditary lymphedema (Milroy Disease), lymphedema precox occurring at puberty, and hereditary lymphedema tarda usually occurring in the third decade of life. Primary lymphedema has been associated with congenital syndromes including Turner's syndrome and Noonan's syndrome. Secondary forms of lymphedema are most often the consequence of obstruction of the lymphatic system by infection or cancer and trauma.[1]

This patient suffered from congenital lymphedema (infantile hereditary lymphedema). It presents in the first two years of life, and some cases have been linked to mutations in the FLT4 gene. The FLT4 gene is responsible for producing vascular endothelial growth factor receptor 3 (VEGFR-3), which regulates the development of the normal lymphatic system. Mutations in this gene lead to hypoplasia and aplasia of the lymphatic endothelial cells and manifest itself as small or absent lymphatic vessels leading to lymphedema.[1]

This patient was born with a congenital chylothorax that was repaired shortly after birth. Association of lymphedema with chylothorax has been described in both hereditary and sporadic cases. These patients usually have pleural effusions. However, cases involving congenital lymphedema, chylothorax, and pericardial effusions have been rarely reported. One case containing this clinical triad occurred in an older patient who presented with cardiac tamponade on

echocardiogram.[2] Drainage of the pericardial fluid in this case revealed chylous fluid.

Pericardial effusions occur due a transudative or exudative process. Transudative fluids result from obstruction of fluid drainage, which occurs through lymphatic channels. Exudative fluids occur secondary to inflammatory, infectious, malignant, or autoimmune processes within the pericardium.[3] The mechanism behind our patient's pericardial effusion was likely due to impaired lymphatic drainage. The size of her effusion was moderate, and she more than likely did not have symptoms due to the fact that the accumulation of fluid occurred over a long period of time.

The findings in this case illustrate the complex interplay of pathophysiology on organ systems and highlight the importance of a thorough clinical history. Knowing that this patient had an impaired lymphatic system raised suspicion for a pericardial effusion, especially in the setting of a low voltage EKG. Had the effusion been larger or demonstrated signs of cardiac tamponade, she may have been a candidate for pericardiocentesis. At this time, the patient was counseled on signs and symptoms of tamponade and advised to follow-up with an echocardiogram in 1 year.

Images

Figure 1.

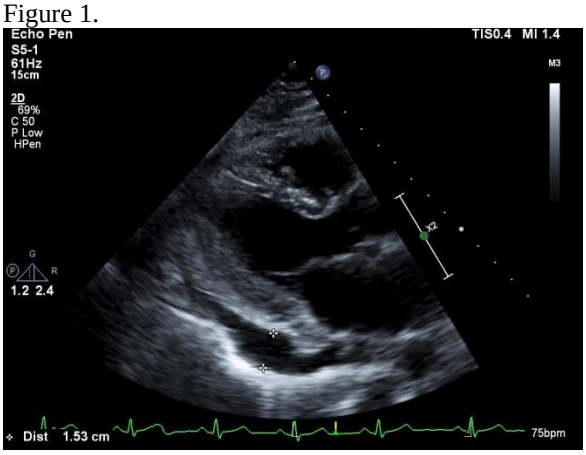
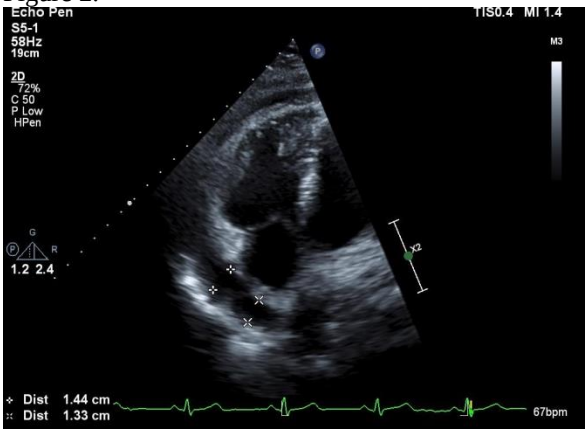


Figure 2.



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Submitted February 22, 2016