Case Report

Pericardial effusion with Cardiac Tamponade and Pulmonary Arterial Hypertension in A Patient with Primary Sjogren's Syndrome-A Rare Case Report

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Abstract
Pericardial effusion in Pulmonary arterial hypertension is associated with significant morbidity and mortality, and its pathogenesis is complex and poorly understood. We present a case of pericardial effusion with cardiac tamponade and pulmonary arterial hypertension with primary sjogren's syndrome.

Keywords:tamponade,pumnonary

Introduction

Pericardial effusion is a relatively common finding in everyday clinical practice. It may occur in patients with chest pain of pericarditic characteristics or in patients with underlying diseases that can cause pericardial involvement (renal failure, chest irradiation) and thoracic complaints. Other patients, without previous known diseases, seek medical attention because of dyspnea or nonspecific chest discomfort and the thoracic X-ray shows the presence of an enlarged cardiac silhouette with clear lungs. The echocardiogram is the most available and reliable technique in order to verify the presence and the amount of a pericardial effusion; in addition, the echocardiogram offers valuable data for evaluation of hemodynamic repercussion[1,2]. Cardiac tamponade is a medical or traumatic emergency that happens when enough fluid accumulates in the pericardial sac compressing the heart and leading to a decrease in cardiac output and shock. Cardiac tamponade is caused by the buildup of pericardial fluid (exudate, transudate, or blood)[2].Pulmonary arterial hypertension (PAH) is a progressive disease of the lung vascular system, primarily affecting the small pulmonary arterioles. A combination of endothelial dysfunction and increased contractility of small pulmonary arteries, proliferation and remodeling of endothelial and smooth muscle cells, and in situ thrombosis leads to progressive narrowing of the blood vessels. This results in a progressive resistance to blood flow and an increase in pulmonary artery pressures[3,4].Sjogren syndrome is an autoimmune condition with a plethora of extra glandular manifestations. Primary Sjogren syndrome associated with initial presentation as large pericardial effusion with tamponade is exceedingly rare, there have only been a few reported cases of symptomatic pericardial effusion and one case report of cardiac tamponade. However, studies have shown that TTE detects asymptomatic cardiac involvement in up to 30 percent of patients.

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Immunopathology of pericarditis in Sjogren's syndrome remain unclear, it has been proposed that immune complex deposition in the pericardium triggers an inflammatory response leading to effusion.

We present a case of previously healthy young woman who presented to the hospital with profound respiratory distress and shock. This case demonstrates the need to consider the autoimmune etiologies in patients presenting with pericardial effusion and cardiac tamponade. Prompt drainage of effusion and aggressive treatment was the key to success for this rare presentation. Ours is the second such case reported in the literature.

Case Report

An otherwise healthy 15-year-old woman was admitted to the hospital with a 15 day history of progressive dyspnea on exertion and anasarca following out-patient Transthoracic echocardiography (TTE) demonstrating a large pericardial effusion and early signs of cardiac tamponade based on exaggerated mitral inflow velocity, the right ventricle free wall collapse and compromised function (Fig 1).

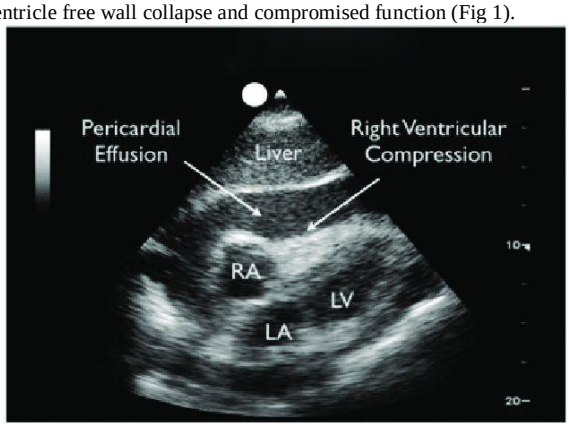

Fig 1: 2D echocardiography

Physical examination revealed jugular venous distension and bilateral pedal edema with no pericardial rub.ECG showed sinus rhythm with signs of left atrial enlargement. Chest X-ray revealed prominent cardiomegaly. Pericardiocentesis removed 800ml of serosanguinous fluid with in-situ pericardial catheter. Cytology showed small clusters

of mesothelial cells, no obvious atypical cells seen, ADA was negative through which tuberculosis was ruled out. TTE the next day revealed re-accumulation of a moderate pericardial effusion. Chest CT scan revealed and enlarged mediastinal lymph nodes and consolidation patchy consolidation of both lower lobes. She was diagnosed with primary Sjogren's syndrome by positive antibodies (Anti-ro and Anti-La antibodies), after the diagnosis she was started on low dose steroid therapy and hydroxychloroquine. Her symptoms resolved and subsequently in the next three days there was no re-accumulation of the pericardial fluid.

Discussion

Sjogren's syndrome is a chronic inflammatory disorder characterized by diminished lacrimal and salivary gland function, although extra glandular disease involvement can involve nearly any organ. Cardiovascular involvement occurs in less than one-third of cases. Pericarditis is the most common form of cardiac involvement in Sjogren's syndromeThe prevalence of pericardial effusion varies depending on the type and severity of PAH studied. The Chinese PAH registry noted that patients with PAH associated with connective tissue diseases had a higher prevalence of pericardial effusion than did individuals with IPAH (21% vs. 13%)[6].Fijalkowska et al described a higher prevalence of pericardial effusion in patients with PH (mostly IPAH) who died than in those who remained alive during follow-up (56% vs. 15%)[7]. A Canadian study reported the incidence and prevalence of pericardial effusion among 154 patients with PAH. During the initial assessment, the authors found a prevalence of 29%, and over a 2-year follow-up period, they found an incidence of 44.1%[8].Anti-SSA/Ro and anti-La/SSB are the hallmark antibodies in primary Sjögren's syndrome (pSS), being present in 60-70%. These antibodies have been associated with an earlier disease onset, glandular dysfunction and extraglandular manifestations as well as with other B cells activation markers. PAH is a rare complication of Primary Sjögren's Syndrome (pSS) and is associated with worse outcomes among patients with Pss[9,10].In 2004, the European executive summary on the diagnosis and management of pericardial diseases suggested that small pericardial effusions should not be drained in patients with PAH[11]. Cardiac tamponade remained a clear indication for drainage. Hence we drained in our case.The presence of pericardial effusion in patients with PAH is an indicator of right heart failure associated with poor outcome. Worsening of symptoms of right heart failure should raise the suspicion for this ominous association. Early detection of pericardial effusion in PAH is accomplished by a high index of suspicion together with echocardiography.

Conclusion

Although rare, physicians must be aware that primary Sjogren's syndrome may cause pericardial effusions. This case report raises the question of whether all patients with Sjogren's would benefit from early TTE screening and early identification of pericardial effusion may warrant closer monitoring to minimize the risk of cardiac tamponade

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