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PRIMARY SJOGREN PRESENTING WITH PERICARDIAL EFFUSION

Medicine

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ABSTRACT

a case report of a 19 year old hypothyroid female presenting with right sided heart failure and pericardial effusion being diagnosed with primary sjogren syndrome

KEYWORDS

Introduction:
Though pericardial effusion and serositis is common in both rheumatoid arthritis and SLE the occurrence of the condition is found only in case reports, although tuberculosis is one of the most common causes of pericardial effusion endocrine and rheumatological causes should always be rouled out in such a case

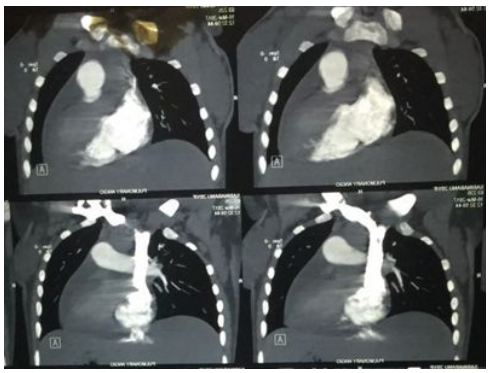


Figure 1 transverse section showing pericaedial effusion

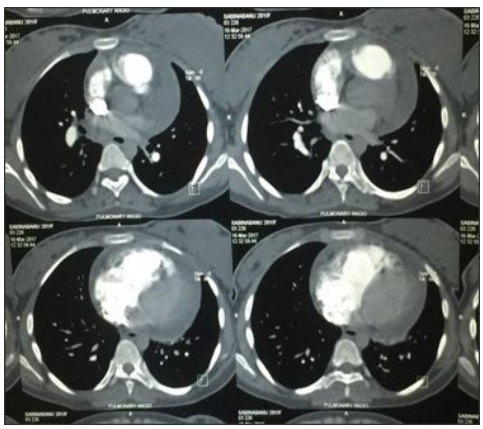


Figure 2 CT thorax showing rim of pericardial effusion

Case report :
A 19 year old muslim female 8 month post-partum presented to the hospital with acute onset dyspnoea, pedal oedema and right hypochondriac pain, after admission a diagnostic 2d echo was performed which was suggestive of mild to moderate pericardial effusion with moderated to severe PAH (rvsp = 86 mmhg) , patients thyroid, protein profile was normal, inflammatory markers ESR was raised to 53 and CRP were positive 25 a confirmatory HRCT was done which confirmed pericardial effusion and PAH. History of parotitis was present so an ANA by immunofluorescence was done was +++ and extractable nuclear antigen was done where anti ro, anti la was strongly positive. Liver and renal functions were normal, HIV, HBsAg and HCV was negative. No evidence of malignancy was found.

Investigations:	On Admission Day	After 5 Days
Hemoglobin	11.5	11.6
WBC Count	7020	5930
Platelets	1,59,000	**1,35,000**
Urea	24	17
Creatinine	0.77	0.70
Sodium	132	132
Potassium	4.3	4.2
SGPT	13	41
Total Billirubin	0.51	0.51
Alkaline Phosphatase	106	101
Serum Proteins	**9.90**	
Serum Albumin	3.35	
Serum TSH	2.23	
Serum B12	798	

Discussion:
Pericardial involvement in CTD
Primary Sjögren's syndrome (pSS) is a chronic autoimmune disease characterised by lymphocytic infiltration of the salivary and lacrimal glands. [6]

Similar lymphocytic infiltrates may invade visceral organs, and this results in several extraglandular manifestations. [6]

Among these, a clinically overt heart disease is very rare. [7-9]

Rantapää-Dahlqvist and colleagues [10] reported signs of present or previous pericarditis in nine of 27 (33%) pSS patients. Of the echocardiographic measurements, the right ventricular anterior wall and the left ventricular posterior wall were significantly smaller in patients with pericarditis than in those without pericardial serositis. Moreover, in the pericarditis patients, the regional fractional shortening of the left ventricle was significantly higher and the hypokinesia of the left ventricle significantly more frequent, when compared with those without pericarditis. Sclerosis of the aortic cups and a slight aortic regurgitation were seen in 11 and three patients, respectively. No patient had mitral valve prolapse or indirect signs of pulmonary hypertension.

Mita et al [11] evaluated 112 patients with SS, primary in 33 and secondary in 79, by two dimensional echocardiography. They reported abnormal findings in 69 (61.6%) of the total cohort and in 55.5% of the

pSS patients. In this second group, pericardial eVusion was seen in 21.2%, thickening/calcification of the aortic valve in 10.3%, decrease in the diastolic descent rate of mitral valve in 6.9%, thickening/ calcification of mitral valve in 3.4%, mitral regurgitation in 3.3%, and mitral prolapse in 3.2%. No pSS patient had pulmonary hypertension.

The most common types of pericardial involvement with systemic inflammatory diseases are acute (or recurrent) pericarditis and asymptomatic pericardial effusions. Pericardial involvement in systemic inflammatory disease is thought to be immune-mediated, although concomitant infection may play a role in some cases. Systemic inflammatory diseases may be either "etiologic" or "permissive" of increased susceptibility to an unrelated primary cause (eg, viral) [3].

While some patients may manifest with the typical signs and symptoms of pericarditis (fever, pleuritic chest pain) or a hemo dynamically significant effusion (dyspnea, fatigue, hypot ension), many are incidentally found to have pericardial involvement during other testing (electrocardiography, echocar diography, computed tomography of the chest, or magnet icsonance imaging of the heart). Symptoms of pericardial involvement in systemic inflammatory disease may be insidious or may present suddenly with rapid progression. Cardiac tamponade and constrictive pericarditis are possible, but rare, complications.

Cardiac manifestation in sjogrens

Valvular regurgitation, pericardial effusion, pulmonary hypertension and increased left ventricular mass index occur with dispropo rtionately high frequency in patients with primary Sjogrens syndrome and no clinically apparent heart disease[1]

In a Chinese study A total of 124 cases were included in this analysis (mean age 47.4 years old; 5 males; average disease duration 85.5 months). Cardiac involvement in Sjogren syndrome is usually asymptomatic. Pericardial effusion (PE) were evidenced in 20.2%, left ventricular diastolic dysfunction (LVDD) in 13.7%, pulmonary artery hypertension (PAH) in 12.9%, left atrium enlargement/in 7.3%, mitral insufficiency in 4.8%, aortic dilation in 5.6%, tricuspid insufficiency in 3.2%, left ventricular enlargement in 2.4% and left ventricular systolic dysfunction in 0.8% patients by echocardiography examinations[2]

Cardiac involvement includes pericarditis, pericardial effusion, and atrioventricular conduction block Pleural and pericardial effusions in association with SS are rare. [6].

Process	Examples
Infectious	Viral Bacterial Mycobacterial Fungal Protozoal HIV
Neoplastic	Primary (mesothelioma, sarcomas, fibroma, lipoma) Secondary (breast, lung, melanoma, sarcoma, lymphoma, leukemia, ovarian)
Immune/ inflammatory	Autoimmune and connective tissue disorders (e.g., SLE, RA, ankylosing spondylitis, scleroderma, sarcoidosis, dematomyositis, polyarteritis nodosa, Wegener granulomatosis, Sjögren syndrome) IBD Löffler syndrome Stevens-Johnson syndrome Myocardial infarction Dressler syndrome Postpericardiotomy syndrome Posttraumatic Pleural and pulmonary diseases
Metabolic	Uremia Dialysis-associated Myxedema Gout
Iatrogenic	Radiation injury Related to cardiac catheterization or placement of implantable cardioverter-defibrillators and pacemakers
Traumatic	Blunt trauma Penetrating trauma Chylopericardium
Drug-induced	Hydralazine, procainamide, minoxidil, isoniazid, anticoagulants

HIV, Human immunodeficiency virus; IBD, inflammatory bowel disease; RA, rheumatoid arthritis; SLE, systemic lupus erythematosus.

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