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RESEARCH ARTICLE

A CASE OF RHEUMATOID ARTHRITIS ASSOCIATED WITH CONSTRICTIVE PERICARDITIS.

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Abstract

RA is a progressive inflammatory autoimmune disease with articular and systemic effects. Its exact cause is unknown, but genetic and environmental factors are contributory. T cells, B cells and the orchestrated interaction of pro-inflammatory cytokines play key roles in the pathophysiology of RA. The release of cytokines, especially TNF- α , IL-6 and IL-1, causes synovial inflammation. In addition to their articular effects, pro-inflammatory cytokines promote the development of systemic effects, including production of acute-phase proteins (such as CRP), anaemia of chronic disease, cardiovascular disease and osteoporosis and affect the hypothalamic-pituitary-adrenal axis, resulting in fatigue and depression.

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Introduction:-

Constrictive pericarditis manifests in less than 10% on rheumatoid arthritis patients. It should be considered in patients with rheumatoid arthritis who develop unexplained cardiac failure. Early diagnosis requires a high index of suspicion and cardiac imaging is necessary to confirm the diagnosis. Medical treatment is largely ineffective and pericardiectomy should be considered.

Case Description:-

A 34 years old Saudi male referred to the rheumatology outpatient clinic for management of recently diagnosed rheumatoid arthritis. The patient had 10 months history of pain involving MCP, PIP and wrists joints bilaterally. He also complained of occasional pain involving both elbows, shoulders, knees and ankles. His pain was associated with morning stiffness lasting for one hour.

He denied visual symptoms, oral ulcers, skin rash, nail changes, hair loss or fever.

He also complained of progressive exertional shortness of breath for 3 months with lower limbs swelling. He denied chest pain, cough, palpitations, dizziness, diaphoresis, syncope or presyncope.

He never smoked or consumed alcohol or illicit drugs. He denied any family history of rheumatoid arthritis or any other rheumatic or non-rheumatic diseases.

On examination, he was comfortable at rest with obvious icteric sclera. Temperature: 37.1 C°, Blood pressure of 111/87, pulse of 66 irregularly irregular, RR 20, Oxygen saturation of 100% on room air. Skin exam showed no rash or nodules. Head and neck exam was significant for jugular venous distension of 5 cm above the sternal angle. There were no palpable lymph nodes. Cardiopulmonary exam was significant for irregular variable heart sounds

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with no murmurs, bilaterally basal fine inspiratory crackles and mild pitting lower limb edema. Abdomen was soft, lax, not distended and without shifting dullness. The liver was mildly tender on palpation with normal span of around 9 cm on percussion. Spleen was not palpable. Musculoskeletal exam showed synovitis of bilateral MCP, PIP and wrist joints with no obvious deformities. Normal elbows, shoulders, knees and hips.

Laboratory work:-

RF was 25 IU/ml, Anti-CCP antibodies moderately positive, CRP 71.2 mg/L

Total bilirubin 81 $\mu\text{mol/L}$, conjugated bilirubin 31 $\mu\text{mol/L}$, GGT 175 unit/L, albumin 3.6 g/L and BNP 747 pg/ml

He had normal CBC, AST, ALT, basic metabolic panel, renal function tests, electrolytes and coagulation profile

Quantiferon test for TB was negative.

His ECG showed atrial fibrillation with a rate of 73 bpm with no ischemic changes. (Figure 4).

Hands x-ray showed periarticular osteopenia without erosive changes. (Figure 1). Chest X-ray showed cardiomegaly. (Figure 2). Abdominal US showed a cirrhotic liver with minimal ascites.

Rheumatoid arthritis was controlled with prednisone 10 mg daily while his cardiac problem was being sorted out. Cardiology service was consulted and echocardiography showed mildly dilated left ventricle, EF of 20%, severe global hypokinesia, markedly dilated left atrium with elevated filling pressures. Right-sided chambers were mildly dilated with severely depressed right ventricular systolic function. Moreover, the free wall of the right atrium was compressed from the outside by thickened pericardium. No pericardial effusion or valvular abnormalities. Cardiac MRI showed a thickened pericardium with an area of maximal thickening overlying the lateral wall of the right atrium consisting with constrictive pericarditis. (Figure 4).



Figure 1:- Hand X-Ray shows periarticular osteopenia.

The patient was started on oral furosemide as well as an ACE inhibitor and a beta blocker as anti-failure medications. He was anti-coagulated with rivaroxaban. Upon follow-up, his shortness of breath and lower limb edema improved remarkably. His BNP decreased to 270 pg/ml and his total bilirubin dropped from 81 to 70 $\mu\text{mol/L}$. Plaquenil therapy was started and Prednisone dose was reduced to 7.5 mg daily.

He was referred for cardiac surgery for the management of his rheumatoid constrictive pericarditis. Pericardiectomy was performed. Pathological exam of the pericardium showed fibrinous constrictive pericarditis with no evidence of infection or malignancy.

Follow-up echocardiogram after surgery showed that left ventricular ejection fraction improved to 50%.



Figure 2:- Chest X-Ray of the patient shows Cardiomegaly.

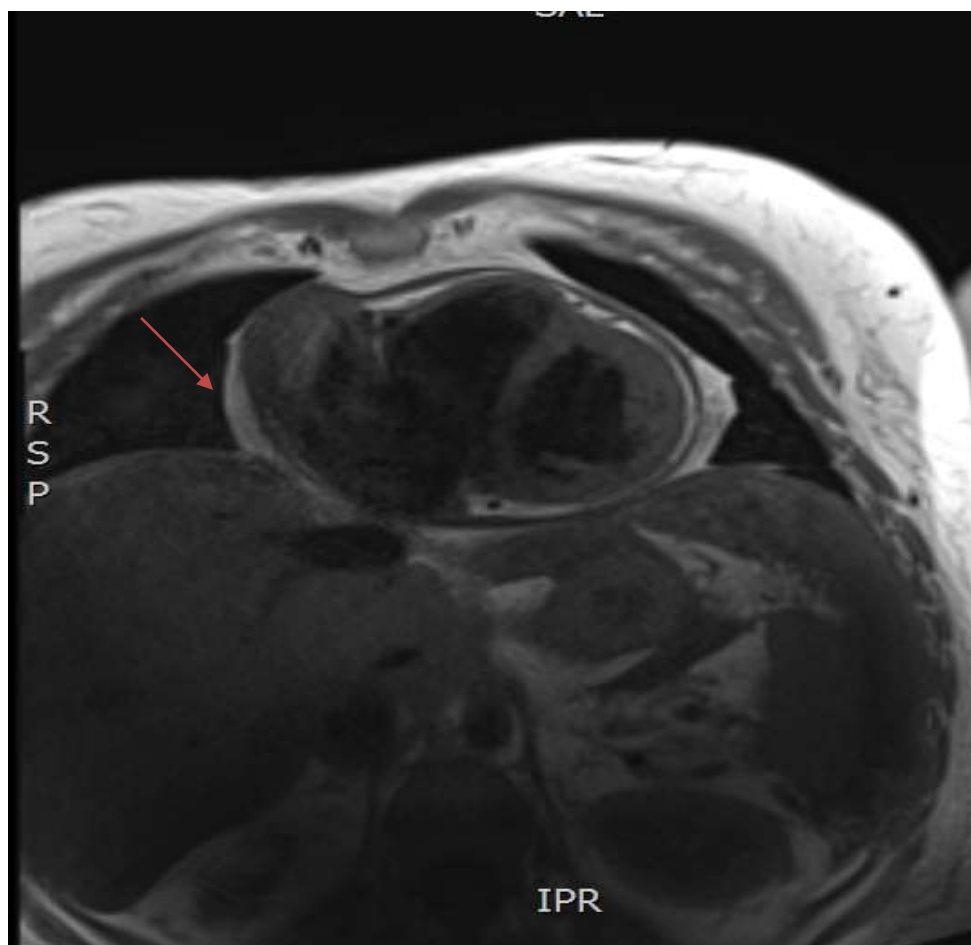


Figure 3:- Cardiac MRI shows thickening overlying the lateral wall of the right atrium (*red arrow*).

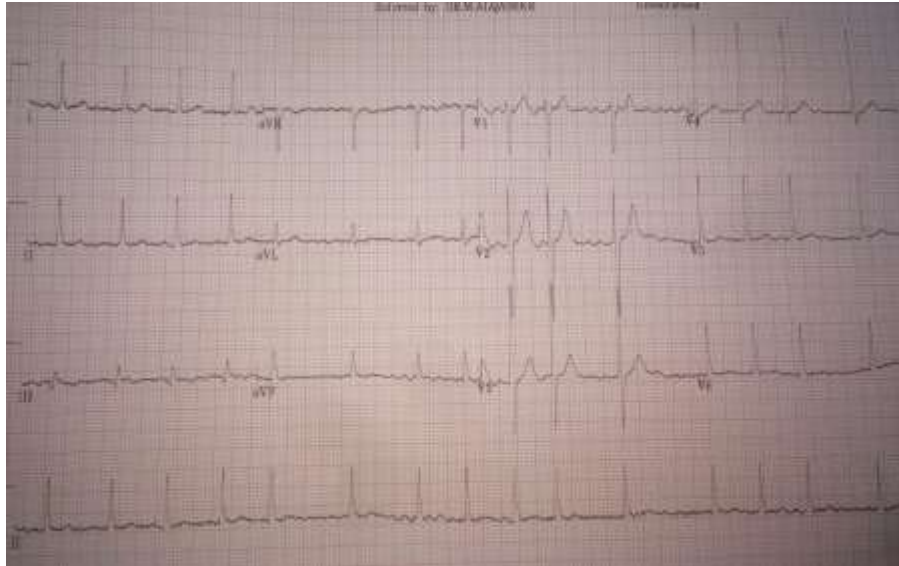


Figure 4:- ECG shows Atrial Fibrillation with ventricular rate 73 bpm.

Discussion:-

Constrictive pericarditis can occur in virtually any pericardial disease process e.g.

Idiopathic/viral (42-61%), post-cardiac surgery (11- 37%), post-radiation therapy primarily in Hodgkin's disease and breast cancer (2- 31%), connective tissue disorder (3-7%), post infectious; tuberculous or purulent pericarditis (3 -15%) percent and other miscellaneous causes.¹

Patient with constrictive pericarditis usually present with symptoms and signs of volume overload due to right and left cardiac failure.²

Several diagnostic tests are available for diagnosis of pericarditis. Chest X-ray occasionally shows pericardial calcification. Trans esophageal echocardiogram is an essential diagnostic test to evaluate constrictive pericarditis. MRI or CT of pericardium is needed to evaluate the patient prior to surgery and are fast tools for diagnosis.

Symptoms of constrictive pericarditis are usually permanent and often progressive requiring pericardiectomy. Steroid therapy gives only temporary relief before pericardiectomy.^{3,4}

Patients with RA occasionally develop constrictive pericarditis requiring pericardiectomy as definitive treatment. Most studies have shown that rheumatoid constrictive pericarditis is commoner in males who tend to have subcutaneous rheumatoid nodules and chest radiograph showing a considerably enlarged heart.⁴

Our case is the first in the literature of a patient with rheumatoid arthritis with Severe constrictive pericarditis early in the disease process presenting with right sided heart failure and congestive hepatopathy causing liver cirrhosis within months of diagnosis. Liver cirrhosis precluded the use of methotrexate and Plaquenil and Prednisone were used instead.

Conclusion:-

Pericarditis in rheumatoid arthritis is associated with poor prognosis. This is a rare case of a young patient with [constrictive pericarditis](#) associated with RA. There is a growing population of early RA patients with cardiac involvement. Early cardiac screening is thus recommended.

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