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Medical Science

STEROID RESPONSIVE SICK SINUS SYNDROME IN LIMITED CUTANEOUS SYSTEMIC SCLEROSIS

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ABSTRACT We report a south-asian woman with skin thickening limited to her hands and a pulse rate of 56 per minute which lead to a diagnosis of sick sinus syndrome with possible limited systemic sclerosis. Observation of moderate thrombocytopenia resulted in evaluation with autoimmune antibodies. She was found positive for anti-nuclear antibody and anti-centromere antibody. Echocardiography showed moderate pulmonary artery hypertension. Treatment with low dose oral gluco-corticoids resulted in restoration of sinus rhythm. Cardiac involvement in limited cutaneous systemic sclerosis is very rare and clinically silent. It often remains undiagnosed until heart failure supervenes. This case illustrated the possible benefit of systemic steroids in sick sinus syndrome associated with limited form of systemic sclerosis.

KEYWORDS : CONNECTIVE TISSUE DISORDER,SYSTEMIC SCLEROSIS,SICK SINUS SYNDROME

BACKGROUND

Systemic Sclerosis is an uncommon connective tissue disorder which has varied systemic involvement. It is a rare disease with an incidence of about 20 cases per million per year and more common in females. The peak age of onset is 45–60 years and prognosis is worse in older individuals, especially increased risk of pulmonary hypertension in disease onset after 65 years [1,2].It is unique among the rheumatic diseases because it presents the challenge of managing a chronic multisystem autoimmune disease with a widespread disrupting vasculopathy of small arteries that is associated with varying degrees of tissue fibrosis. The limited form of systemic sclerosis (lSSc) has a strong female predominance, with a female-to-male ratio of 10:1.[3] In limited systemic sclerosis Raynaud's phenomenon is most common and often first sign of disease. Other signs usually appear some years later. Visceral organ involvement shows insidious progression and occurs as late complication. This case highlights the importance of evaluating a patient with asymptomatic sick sinus syndrome for possible connective tissue disorder.

CASE PRESENTATION

A 53 year old female with no prior medical comorbidities visited our outpatient department with symptoms of heartburn, early satiety, reflux of food contents ,nausea and vomiting for 5 days. She also gave history of experiencing fatigue while doing her day today physical activities. She had no history of palpitations, syncope , dyspnea, skin changes, polyarthralgia or other features of connective tissue disorder. There was no past history of drug intake or sudden cardiac death in the family. On examination, her general condition and appearance was normal. Pulse rate was 56 beats per minute which was irregular, blood pressure -130/80mm Hg. She had skin thickening limited to both her hands. Systemic examinations were normal except for minimal tenderness over the epigastrium.

INVESTIGATIONS

ECG showed atrial fibrillation with marked sinus bradycardia and a 24-hour holter monitoring showed evidence of sick sinus syndrome She also had an incidental finding of isolated thrombocytopenia and trace albuminuria. With background finding of skin thickening of hands, autoimmune workup was done which showed positivity for anti-nuclear antibodies and anti-centromere antibody suggesting the presence of limited cutaneous systemic sclerosis. Echocardiography showed severe tricuspid regurgitation and pulmonary artery pressure of 38mmhg. Hemoglobin-10.2 gm%, WBC count-6700 cells per cubic mm, platelet count of 68,000 per cu mm. ESR – 60 mm at one hour. Renal function test and liver function tests were normal. Trace albuminuria was present. Ultrasonography of abdomen was normal.

TREATMENT

We initially treated her with proton pump inhibitors for GERD symptoms. Sildenafil 25mg twice daily was given for the pulmonary artery hypertension. Since she had asymptomatic bradycardia placement of a pacemaker device was deferred, yet she was advised about necessity for permanent pacing considering the poor outcome of sick sinus syndrome in systemic sclerosis. She was started on oral predinisolone 10 mg per day.

OUTCOME AND FOLLOW UP

During follow-up after 12 weeks her pulse rate was 72 beats per minute. Her follow-up ECG showed normal sinus rhythm . Review echocardiography at 8 months did not show any signs of pulmonary artery hypertenssion.

DISCUSSION

Sick sinus syndrome is an abnormality of cardiac rhythm which occurs as a result of primary pathology of sinus node or external causes which secondarily affect the sinus node function.[4] Sick sinus syndrome often has no identifiable cause and can occasionally be multifactorial. [5] Sick sinus syndrome is frequently the result of malfunction of SA node due to degenerative fibrosis which occurs as a result of inflammation, infiltration , fibrosis or atherosclerosis.

Sick sinus syndrome can clinically present with non-specific dizziness or alarm symptoms like syncope, dyspnea and altered cognition. In the absence of ongoing symptoms, clinical examination is unremarkable except for bradycardia in a small proportion of patients. Sick sinus syndrome produces a spectrum of electrocardiographic changes which include tachy and bradyarrhythmias of atrial or atrio-ventricular origin. [6] SSS is often confirmed with a 24 hour holter monitoring. Symptomatic bradyarrthythmia is treated with pacemaker implantation while if asymptomatic, watchful waiting should suffice. Our patient recovered with systemic steroids alone.

Isolated thrombocytopenia could be occasionally the only clue to certain diseases like systemic lupus erythematosus, anti-phospholipid antibody syndrome, HIV and lymphoma.[7] Current literature is less clear in the extent of evaluation for isolated thrombocytopenia.

Limited form of systemic sclerosis often presents with skin changes distal to elbow and knees. Heart may be affected in systemic sclerosis in the form of myocarditis, pericarditis and conduction abnormality. Symptomatic cardiac involvement is associated with poor prognosis. Supraventricular tachyarrhythmia is more frequent than ventricular tachyarrhythmia and sick sinus syndrome is rare.[8]

Presence of anti-centromere antibodies and ANA is associated with limited cutaneous form of systemic sclerosis. Anti-centromere antibody is associated with involvement of pulmonary vascular bed, oesophagus, lung and kidneys.[9] The occurrence of steroid responsive SSC along with limited form of systemic sclerosis is rare.

Though low dose glucocorticoids may benefit systemic sclerosis, Higher doses are often avoided due to fear of precipitating renal crisis.

Our case highlights a possible benefit of steroids in the setting of SSC. Long term favourable prognosis in limited cutaneous form changes to unfavourable prognosis when organ involvement occurs.

This case illustrates the need for screening for possible systemic cause in patients with sick sinus syndrome. Steroid responsive sick sinus syndrome in limited systemic sclerosis need further confirmation in future clinical observation.

LEARNING POINTS

1. Proper systemic assessment is beneficial in a patient with Sick sinus syndrome, especially examination focusing features of autoimmune diseases.
2. Isolated thrombocytopenia is an important clinical clue to autoimmune disease
3. Sick sinus syndrome associated with limited form of systemic sclerosis may benefit from systemic steroids.

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