Volume : 3 | Issue : 5 | May 2014 • ISSN No 2277 - 8179

Scleroderma Associated with Rheumatic Fever : A Case Report

Medical Science

KEYWORDS : Scleroderma, Rheumaticfever, mitral stenosis, RheumaticHeartdisease

Om Shankar — Département of Cardiology, Institute of Medical Sciences, Banaras Hindu University, Varanasi-221005 Uttar Pradesh. * Corresponding Author

Vibha Pandey — Department of Anatomy, Institute of Medical Sciences, Banaras Hindu University, Varanasi-221005Uttar Pradesh.

Kapil Malviya — Department of Anatomy, Institute of Medical Sciences, Banaras Hindu University, Varanasi-221005 Uttar Pradesh.

ABSTRACT

We described a case of scleroderma associated with rheumatic fever. A 21-year-old female was admitted to our hospital because of polyarthralgia, cubital nodules with redness and pain of the elbows, and sclerodactyly. The diagnosis of rheumatic fever was confirmed by clinical features and high titer of ASK. After daily administration of 1.2 million unit of benzathine penicillin G was started, polyarthralgia and acute inflammatory reaction were improved but scrlodactyly continued. Scleroderma was diagnosed because the histological findings in skin biopsy revealed a striking increase of compact collagen fibers in the deeper reticular dermis. D-penicillamine of 100mg per day was then started, and sclerodactyly was gradually improved. There have been no reports India from of scleroderma with rheumatic fever. This case was thought to be interesting in order to consider the pathogenesis of scleroderma, because both scleroderma and rheumatic fever are considered to be based on immunological disorder.

Introduction

Scleroderma affects multiple organ systems, manifestations of the diseaseare diverse involving various systems,the reported organ systems involved are pulmonary,cardiovascular,renal etc. Heart is a major organ involvedin scleroderma and the presence of cardiac involvement generallyportends poorly for the patient. Cardiac involvement can either be direct myocardial effects or theindirect effect of other organ involvement (i.e. Pulmonary hypertension, renal crisis).The involvement of the heart in scleroderma was first identified in 1926 by Heine followedby Weiss et al, who described 9 cases of Scleroderma Systematic Sclerosis (SSc) with congestive heart failure and it was first postulated that cardiac fibrosis was theetiology only 5 Clinical association of scleroderma and mitral stenosis has been recorded by Crocker (1885),Ramond (1928), and Ramsey (1951),Evans & parker(1954). The account by Rake (1931) is the only one we can find in which the diagnosis was confirmed postmortem.

Clinical Presentation:

A 20 year old girl presented to our Medicine OPD, Sir Sunderlal Hospital, Institute of Medical Sciences, Banaras Hindu University with the chief complaint of maculopapular rashes on upper and lower extremities, dyspnoea and palpitation on exertion (Figure1A). She gaveHistory of nonmigratory symmetrical pain in bilateral knee jointwith feverforabout one month when she was 9 years old, but there was no history of rheumatic fever (sore throat, skin rashes, chorea). She also gave history of painful bluish discoloration of fingers and toes and chilblains on extremities since last 7 years.She was alert,conscious,slightly cyanosed,pale with cold extremities and ectomorphic built. Therewas increased pigmentation of the skin all overthe body, marked over the face legs and forearms, andslight bilateral ptosis(Figure1B). The skin of the face wasthin,pigmented, and tightly stretched over the bony structures.Lips were thin.She had stiff, claw-like hands with shallow ulcers and several scars and blunt terminal phalanges and deformed nails,The skin over the fingers and thumbs was hard,smooth, and fixed; that over the rest of the hands was tightly stretched (Figure 1 C&D). There was a gradual transition to normalskin in the forearm. Similar picturewas present on the feet and extendedup toankles.Feet and legsup to knee showed multiple healed scars. The skin of the neck and upper part of the chest was thin, taut and prominent veins were seen, with blood flow from down to up.

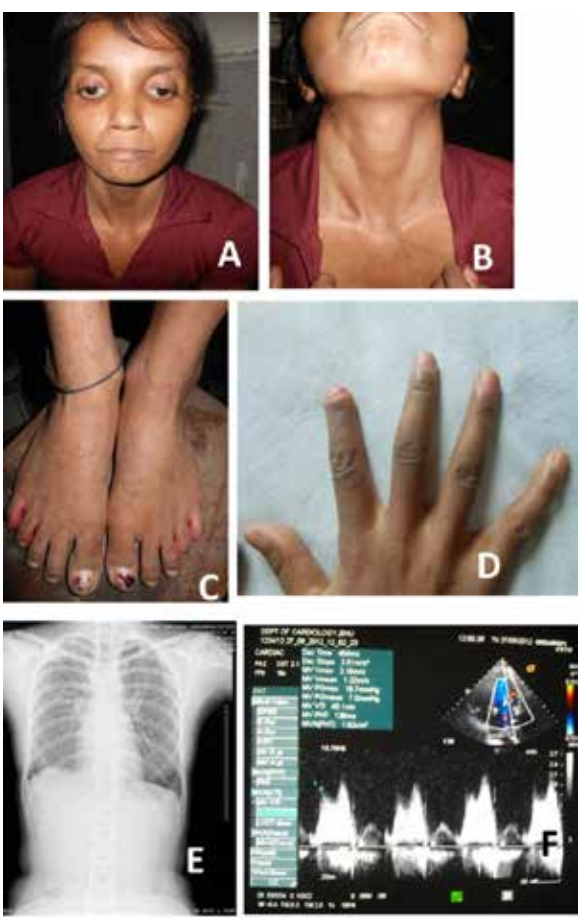

Figure 1: Figure showing (A), (B),(C) and (D)thin, pigmented, and tightly stretched skin

foot andhands with shallow ulcers and several scars and blunt terminal phalanges and deformed nails, The skin over the fingers and thumbs was hard, smooth, and fixed.(E). Chest skiagram showing no abnormality.(F). Echocardiogram showing mitral stenosis.

There was no rise of jugular venous pressure. The blood pressurewas 110/70 (mm. Hg).The apexbeat was feebly palpable in

fifth intercostal space 5 cm from mid-clavicular line. There was a loud rumbling mid diastolic murmur present in mitral area and audible in whole of precordium it radiated to back. Murmur increased on expiration. There was no opening snap or pre systolic accentuationheard. The murmur increased on physical exertion. The murmur found in this case is typical of rheumatic mitral stenosis.The lungs were resonant all over, and persistent basal crepitations were audible over both lungs. Movements of the left side of the soft palate were very limited, and this area was indurated.Ptosis, and marked muscle wasting wasnoted, no other significant abnormality was detected.Urine examination: 24 hour urine volume was 2.6 L. And urinary protein was 10 mg/dl.No RBC,WBC cast were seen.WBC count 9200 per cmm., and the E.S.R. was raised to 32 mm after one hour. The blood urea was 25.4 mg. perdl.; total serum proteins 7.5 g. per dl.(albumin 4.44 and globulin 3.06).Alkaline phosphatase 276.1 U/l.Sputumwas negative for tubercle bacilli. Electrocardiogramsshowed signs of left atrial enlargement.X-ray chest showed diffuse parenchymal infiltration and bronchovascular margins were prominentand left border of cardiac silhouettes was straightened,showing mitralisation of heart (**Figure 1 E**). Echo cardiogram suggest Mitral valve dysfunction as stenosis (**Figure 1 F**). Examination of the oesophagus by barium swallow did not show any abnormality.

Discussion:

Systemic sclerosis is a systemic autoimmune disease with fibrosis of the different tissues because of the collagen formation and other extracellular proteins. Its etiology is unknown but its pathophysiology involves microvascular abnormalities, secondary ischemia, and fibroblast overactivity. It has been lessthan a decade since when heart disease has been acknowledged as a serious and not an uncommon complication of systemic sclerosis. More recent studies suggest that clinical evidence of myocardial disease may be seen in 20% to 25% of patients with SSc Follansbee et al;1986,1990). The presence of clinical cardiac involvement in SSc is forerunner of a poor prognosis. (Medsger and Masi 1973) showed that clinical cardiac disease in SSc was associated with a 70% mortality at 5 years. Cardiac manifestationsin SSc vary from silent involvement to overt clinicalsigns associated with increasing mortality and morbidity (Kahan et al; 2009).The cardiac manifestations of systemic sclerosis could be in form of myocardial abnormalities, including segmental wall-motion abnormalities, and impaired coronary flow reserve in the absence of epicardial coronary artery disease, and coronary vascular diseases (Kahan et al., 1986;Vaccaet al; 2006). Microvascularabnormalities are base events in patients with systemic sclerosis.The stiffness of the microvascular andlarge arteries has been reported (Cheng et al., 2003) Primary valvulardisease is uncommon in patients with systemic sclerosis (Kahan et al., 2009). As concluded by (Kahan and Allanore 2006) in their study Primary myocardial involvement is likely to result from the general vasospastic mechanism that is thought to play a key role in this disease. Vasospasm of the small coronary arteries or arterioles would initially impair perfusion and function, with reversible involvement. This would be followed by structural coronary arteriolar lesion leading to irreversible abnormalities. Myocardial Raynaud's phenomenon (RP) has been postulated in scleroderma, but the availablefindings suggest that it is different from peripheral RP. Peripheral RP appears to involvesignificant anatomic narrowing of the vessels(Steen 1988, 2004) In contrast, SSc patients with myocardialischemia demonstrate only infrequent luminal narrowing of the small arteries in the heart (Steen 1988, 2004).Many studies have examined the effect of cold pressor provocation on myocardialfunction and perfusion in SSc patients to assess the presence of possible cold-induced coronaryvasospasm but the results of these studies are mixed and the clinical and prognosticsignificance require further investigation(Deswal et al.,1996, Steen et al., 2004).

The rarity of mitral stenosis (which is the commonest rheumatic valvular lesion) in patients with scleroderma supports our belief that the relationship, is just a chance.

REFERENCE

1.Cheng, K. S. Tiwari A. Boutin A. et al., "Carotid and femoral arterial wall mechanics in scleroderma," Rheumatology, vol. 42, 11,1299–1305, 2003 | 2.Deswal A, Follansbee W. Cardiac involvement in scleroderma. Rheum Dis Clin North Am | 1996;22:841. [PubMed: 8923599] | 3.Follansbee WP. The cardiovascular manifestations of systemic sclerosis (scleroderma).CurrProblCardiol 1986;11:241. [PubMed: 3487417] | 4.Follansbee WP, Miller TR, Curtiss EI, et al. A controlled clinicopathologic study of myocardial fibrosis in systemic sclerosis (scleroderma). J Rheumatol 1990;17:656. [PubMed: 2359076] | 5.Kahan and Y. Allanore, Rheumatology 2006;45:iv14–iv17 doi:10.1093/rheumatology/kel312 | 6.Kahan, A.Coghlan, G. and McLaughlin, V. "Cardiac complications of systemic sclerosis.," Rheumatology, 2009; 48,3,45–48. | 7. Kahan, A. Devaux, J. Y. and Amor, B. "Nifedipine and thallium-201 myocardial perfusion in progressive systemic sclerosis," New England Journal of Medicine, vol. 1986. | 314, 22, 1397–1401. | 8.Medsger TJ, Masi A. Survival with scleroderma. II. A life-table analysis of clinical and demographicfactors in 358 male U.S. veteran patients. J Chronic Dis 1973;26:647. [PubMed: 4587746] | 9.SteenV.Clinical manifestations of systemic sclerosis. SeminCutan Med Surg 1998;17:48. [PubMed:9512107] | 10.SteenV.The heart in systemic sclerosis. CurrRheumatol Rep 2004;6:137. [PubMed: 15016344] | 11.Vacca, A. Siotto, P. Cauli, A. et al., "Absence of epicardialcoronary stenosis in patients with systemic sclerosis withsevere impairment or coronary flow reserve," Annals of the | Rheumatic Diseases, vol. 65, no. 2, pp. 274–275, 2006. |