High frequency skin ultrasonography in systemic sclerosis

Iffat Hassan, MD[1]
Peerzada Sajad, MD[1]
Majid Jahangir, MD[2]

1. Department of Dermatology, STD
   and Leprosy, Govt. Medical College
   Srinagar, University of Kashmir,
   J&K, India
2. Department of Radiodiagnosis and
   Imaging, Govt. Medical College
   Srinagar, University of Kashmir,
   J&K, India

Corresponding author:
Iffat Hassan, MD
Associate Professor & Head,
Postgraduate Department of
Dermatology, STD & Leprosy,
Government Medical College Srinagar,
University of Kashmir, J&K, India
E-mail: hassaniffat@gmail.com

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Background: Systemic sclerosis is an autoimmune connective tissue disorder which is characterised by cutaneous and internal organ fibrosis. Raynaud's phenomenon is the earliest feature and may precede the onset of the disease by months to years. The heart, lungs, gastrointestinal tract, kidneys and other organs may be involved. Our aim was to evaluate skin thickness and echogenicity in systemic sclerosis patients by ultrasound and compare it with the healthy age and sex matched controls.

Method: Our study included 15 successively admitted patients (14 females and 1 male) with systemic sclerosis and 15 healthy age and sex matched controls. All the patients met the ACR criteria of diagnosis. The age range of the patients was 25-55 years. The disease duration ranged from 1-8 years. A complete history and physical examination was done for each patient. Skin thickness and echogenicity were measured by a 20MHz ultrasound probe at 6 different anatomical sites which was compared with that of the controls.

Result: Skin thickness was more in systemic sclerosis patients (significant p value) as compared to controls, and echogenicity was inversely proportional to the skin thickness.

Conclusion: Skin ultrasonography is a noninvasive method which can reflect the severity of skin involvement and periodic assessment of skin thickness and echogenicity can help to monitor the progression of the disease.

Keywords: echogenicity, skin thickness, systemic sclerosis, ultrasound

INTRODUCTION

Scleroderma is a connective tissue disease that involves changes in the skin, blood vessels, muscles and internal organs. It is a type of autoimmune disorder, a condition that occurs when the immune system mistakenly attacks and destroys body tissues. The cause is not known [1]. Various etiological factors have been postulated like genetic, environmental, immune mediated, infective, toxic, etc. System sclerosis has three main hallmarks: cellular and humoral immunity activation, microvascular damage and widespread tissue fibrosis [2-6]. Fibrosis of the skin can be assessed by palpation and its extent predicts both internal organ involvement and survival.

The established method of skin assessment is the Rodnan's scoring system, where palpation is done and the anatomical sites are graded as mild (1), moderate (2) and severe (3). But its sensitivity is low and inter-observer variability is high. Various previous studies have shown that high frequency ultrasound offers a potential for quantitative assessment of skin thickness and echogenicity in systemic sclerosis with a very low inter- and intra-observer variability [7,8]. In this study, we aimed to measure the skin thickness and echogenicity in systemic sclerosis patients and to compare it with healthy age and sex matched controls.

PATIENTS AND METHODS

Our study included 15 successively admitted patients (14 females and 1 male) with systemic sclerosis and 15 healthy age and sex matched controls. All the patients met the American college of Rheumatology criteria of diagnosis. The patients ranged from 25 to 55 years in age. The disease duration ranged from 1-8 years. A complete history and physical examination was done for each patient. The disease was classified as limited and diffuse cutaneous systemic sclerosis according to the extent of skin involvement. Skin thickness and echogenicity were measured by a 20MHz ultrasound probe at 6 different anatomical sites (middle phalanx, dorsum of hand, forearm, forehead, sternum and legs) and the findings were compared with that of the controls. Two scans were obtained of the tissue: a one-dimensional A- mode image with different echoes defining the interface between epidermis, dermis and subcutis and a two-dimensional B-mode image with different colours reflecting the different echogenecities of the skin. All measurements were taken by the same observer. The anatomical sites which were used for measurement included the dorsal aspect of the proximal phalanx of the right second finger, dorsum of hand between the wrist and metacarpophalangeal joints, forearm 2 cm proximal to the wrist joint, forehead, sternum 2 cm distal to the manubrium sterni, and legs 3 cm proximal to the ankle joint.

An informed consent form was signed by each patient, and the study was approved by the local ethical committee. The statistical test for analysis was the one-sample t-test.

RESULTS

Of the 15 patients, 14 were female (94%) and one was male. The age range of the participants was 25-55 years and disease duration ranged from 1-8 years. This study revealed that the patients had maximum skin thickness over hands. The mean skin thickness at the examined sites were as follows: hands (3.00 mm vs. 1.15 mm in controls), middle phalanx (2.65 mm vs. 1.01 mm in controls), forearm (2.50 mm vs. 1.10 mm in controls), forehead (2.65 mm vs. 1.20 mm in controls), sternum (2.95 mm vs. 1.25 mm in controls) and legs (2.60 mm vs. 1.40 mm in controls). A significant difference of skin thickness was found between patient and healthy groups (p<0.05). The echogenicity was inversely proportional to the skin thickness. The patients with diffuse skin involvement displayed higher skin thickness and lower echogenicity as compared to the patients with limited skin involvement. There was an inverse correlation between skin thickness and echogenicity as measured by ultrasound at all sites, suggesting that the thicker skin was initially caused by oedema.

DISCUSSION

Systemic sclerosis is a connective tissue disease that involves changes in the skin, blood vessels, muscles and internal organs. It is a type of autoimmune disorder, a condition that occurs when the immune system mistakenly attacks and destroys healthy body tissues. The cause is not known. Various etiological theories which have been postulated include genetic, environmental, immune-mediated, infective, toxic, etc [1]. It usually affects people in the age group of 30-50 years with a male: female ratio of 3-4:1. It can occur with connective tissue disease like lupus erythematosus, dermatomyositis and polymyositis when it is called mixed connective tissue disease [3,4]. It is of two types: localized or limited cutaneous sclerosis and diffuse systemic sclerosis [5,6], depending on the extent of skin involvement. The symptoms of disease include Raynaud's phenomenon, binding down/hardness of skin, dyspigmentation, calcinosis, ulceration of the fingertips and toes, mask like facies, joint pains, numbness and pain in feet, stiffness and swelling of the fingers and joints, wrist pain, dry coughs, shortness of breath, wheezing, bloating after meals, constipation, diarrhoea, dysphagia and reflux/heartburn [9,10]. Treatment consists of vasodilators, corticosteroids, immunosuppressants, NSAIDS and symptomatic management of the internal organ involvement [11]. The causes of death are usually pulmonary fibrosis, heart failure, kidney failure, malabsorption and malignancy [12].

Various previous conducted studies have shown a strong inverse relationship between skin thickness and skin echogenicity measured by ultrasound in patients with systemic sclerosis and disease duration <2 years. This relationship indicates that the oedema that occurs early in the disease course results in both increased skin thickness and

decreased skin echogenecity; therefore, ultrasound may be useful in ascertaining whether a patient is in the early edematous phase. This relationship disappears when the edematous phase is replaced by the indurative phase [13]. In a longitudinal pilot study of 16 patients with early systemic sclerosis and 16 controls, diffuse but not limited sclerosis patients had higher skin thickness and lower skin echogenicity than controls. Unlike healthy individuals, these systemic sclerosis patients displayed large variations between different ultrasound measurements over time in the same individual. During the follow-up, skin thickness decreased and skin echogenicity increased. This study included 5 different anatomical sites and showed that high frequency ultrasound had a very low inter- and intra-observer variability [14,15].

The limitations of our study were the small number of the patients, and no follow-up period to monitor the progression of the disease. However, Skin ultrasonography seems to be a non-invasive method which can help to identify the early edematous phase, detect skin involvement early, and assess the severity of skin involvement. Moreover, periodic assessment of skin thickness and echogenecity, if done by ultrasound, can help to monitor the progression of the disease.

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