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ORIGINAL ARTICLE

Ultrasonography and Nailfold Capillaroscopy Assessment in Patients With Systemic Sclerosis and Their Relation to Severity of Vasculopathy

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Abstract

Background: **Systemic sclerosis (SSc) is a disease of unknown etiology characterized by tissue fibrosis and microvascular abnormalities. Microvascular damage of the hand can easily be evaluated using nailfold capillaroscopy (NFC).**

Aim: **To evaluate the relationship between microvascular damages assessed by NFC and macrovascular features evaluated by Doppler Ultrasound to assess the severity of hand involvement and vasculopathy in SSc.**

Methods: **This study was carried out on 50 adult patients with progressive SSc.**

Results: **According to NFC findings, nine (18%) patients were diagnosed with early phase of SSc while 24 (48%) patients were in the late phase and 17 (34%) patients showed active disease. We used power doppler ultrasonography (PDUS) after NFC for detecting the incidence of pathological finger pulp blood flow (FPBF) and ulnar artery occlusion (UAO). We found that 24 (48%) patients showed UAO and 29 (58%) patients showed pathological FPBF. According to the association between ultrasonography/NFC findings and other parameters assess the severity of vasculopathy. There was a significant association between the presence of (Raynaud's phenomena, Pitting scar, Digital ulcer, acro osteolysis, and Calcinosis) and ultrasonography/NFC findings ($P < 0.05$).**

Conclusion: **NFC is a valuable complementary tools for evaluation of different aspects of SSc microangiopathy together with UAO and pathological FPBF as a main diagnostic parameters in PDUS may represent a relevant severity marker of vasculopathy in SSc.**

Keywords: Nailfold capillaroscopy, Systemic sclerosis, Ultrasonography, Vasculopathy

1. Introduction

Sustemic sclerosis (SSc) is an illness of obscure etiology described by tissue fibrosis and microvascular irregularities. Raynaud's peculiarity (RP) and computerized ulceration, as well as normal instances of microvascular illness. Patients with super durable hyperplasia or sinewy fibrosis of the computerized conduit and consequently the arteriole or arteriole of the organ can utilize it to cut the instinctive RP.[1]

Capillaroscopy is a painless or safe innovation that permits perception or estimation of early microvascular irregularities normal for optional RP. Capillaroscopy offers an extraordinary perspective on microcirculation and its utilization in illnesses where a microvascular part is thought of; it can likewise show me normal history and regular history.[2]

Ultrasonography (US) has fundamentally worked on the appraisal of joints and ligaments in rheumatoid joint pain and other rheumatic sicknesses. Hence, US hand evaluation is presently utilized in day to day practice for the majority rheumatic illnesses. Use B-mode investigation and Doppler assessment capability to translate.[3]

At the point when the lower arm holds the whole gadget, the gadget utilizes power doppler ultrasonography (PDUS) or nail-overlap capillaroscopy (NFC).[4]

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Therefore; this study we aimed to evaluate the relationship between microvascular damages assessed by Nailfold capillaroscopy (NFC) and macrovascular features evaluated by Doppler Ultrasound to assess the severity of hand involvement and vasculopathy in SSc.

2. Patients and methods

This study was carried out on 50 adult patients with progressive SSc diagnosed according to the 2013 American College of Rheumatology/European League Against Rheumatism SSc classification criteria (ACR/EULAR) of those attending the outpatient clinic and inpatient of Rheumatology and Dermatology Departments of Al-Azhar university hospital (Assuit) during the period from July 2019 to end March 2022. Informed consents were obtained from all patients, ethical committee of the institute approved the study.

We prohibited patients with other connective tissue illnesses, for example, B. rheumatoid joint pain, and foundational lupus erythematosus. Scleroderma copies, for example, eosinophilic fasciitis, reflex thoughtful dystrophy, morphea, straight scleroderma, diabetic kerioarthropathy, Burger infection, and other vasculitides were likewise rejected.

All patients went through a total clinical history, fitting clinical assessment and research facility tests (counting total blood count, ESR, C reactive protein, aspartate aminotransferase (AST), alanine aminotransferase (ALT), creatinine, urea, rheumatoid variable (RF), antinuclear antibodies (ANA), hostile to Scl-70 antibodies), hostile to centromere Stomach muscle (ACA), fasting blood glucose, complete urinalysis, cholesterol, low-thickness lipoprotein (LDL), high-thickness lipoprotein (HDL), and fatty oils. Echocardiography and blood glucose observation were performed. Procedure.[5,6]

2.1. Rodnan skin score (RSS)

Skin thickness was surveyed by clinical palpation of 17 body regions on a size of 0–3 (typical, gentle, moderate, serious). The adjusted Rodnan score for all-out skin thickness, got from the amount of scores of the 17 body regions.[7]

2.2. Ultrasound assessment

Blood stream in the ulnar corridor at the wrist was estimated utilizing a cross-segment of Guyon's waterway including the pisiform bone. Ulnar artery occlusion (UAO) was characterized as a complete shortfall of blood stream surveyed by DUS. The outspread corridor was likewise surveyed utilizing a palmar longitudinal view before entering the physical snuffbox. Ultrasound estimations were performed utilizing a gadget (Toshiba Xario200) furnished with a 10–12 MHz direct test.

2.3. Capillaroscopic assessment

NFC assessments were performed at confirmation by a similar inspector (AL) utilizing a DermLite dermatoscope (Heine Delta 20). Microvascular sores were reviewed utilizing the Cutolo order. We give specific consideration to the number of vessels/mm on the center finger of the prevailing hand. Serious going bald was characterized as less than 4 vessels/mm. The consequences of PDUS highlights were joined with NFC examination to choose patients with more serious vasculopathy portrayed by a relationship among large scale and microvascular sores.

2.4. Radiographic appraisal

All patients had standard posteroanterior radiographs of two hands taken at the hour of affirmation. Calcinosis was characterized by the presence of something like one delicate tissue calcification. Acroosteolysis was characterized as any bone resorption of something like one distal phalanx. The inspector who assessed the radiographs and NFC was likewise dazed by the aftereffects of the PDUS assessments.

The collected data was reviewed, organized, tabulated, and statistically analyzed using the Statistical Package for Social Sciences (SPSS 20, IBM Corporation, Armonk, New York, USA). Data were presented as mean ± SD and percentage and appropriate analysis was performed. The significance level was accepted if the P value was less than 0.05.

3. Results

This study included 50 patients with SSC, their mean age was 38.27 ± 3.91 years and 41 (82%) patients were females. The mean BMI was 28.3 and the age of onset of SSc was 26.3 years on average with a mean of 9.1 years of disease duration. Eight male patients of the studied sample were smokers 19 (38%) of them suffering from Hypertension six (12%) of them showed hyperlipidemia and only one patient has type 1 diabetes (Table 1).

According to Patients' Manifestations related to SSc, 38 (76%) patients showed Raynaud's phenomena, 27 (54%) patients with Pitting scars, and 17 (34%) with Digital ulcer. Two (4%) patients had previous Renal crises while two (4%) patients had advanced pulmonary hypertension and one of them

Table 1. Demographic data of studied patients.

Patients' parameters	Mean/N (±SD/%)
Age (year)	38.27 ± 3.91
Sex	
Male	9 (18)
Female	41 (82)
BMI	28.3 ± 6.4
Age of onset	26.3 ± 6.2
Disease duration	9.1 ± 3.6
Associated Morbidities	
Smoking	8 (16)
DM	1 (2)
HTN	19 (38)
Dyslipidemia	6 (12)

was complicated by lung fibrosis. Telangiectasia was found in 32 (64%) patients and 29 (58%) patients who suffered from Arthritis pain. The mean of the modified Rodnan Skin Score was 9.2 ± 6.8 (Table 2).

Regarding laboratory results, Hb was 10.9 gm/dl on average in all studied groups, white blood cell was 7.2 ± 1.9, Platelets was 261 ± 59.1 and ESR was 59.6 ± 17.4. 26 (52%) patients were positive for Anti-centromere, 23 (46%) of them were positive to anti-Scl70 and 47 (94%) were ANA positive.

According to NFC findings, nine (18%) patients were diagnosed with the early phase of SSc while 24 (48%) patients were in the late phase and 17 (34%) patients showed active disease (Table 3).

We used PDUS after NFC for detecting the incidence of pathological finger pulp blood flow (FPBF) and UAO. We found that 24 (48%) patients showed UAO and 29 (58%) patients showed pathological FPBF (Table 4).

A statistically significant difference between the incidence of UAO in patients with early or active stage patients and between late-stage patients. A statistically significant difference between the incidence of Pathological FPBF in patients with early/active stage patients and between late-stage patients (Tables 5 and 6).

Table 2. Patients' manifestations.

Manifestations	Mean/N (±SD/%)
Raynaud's	38 (76)
Pitting scar	27 (54)
Digital ulcer	17 (34)
Acroosteolysis	18 (36)
Calcinosis	20 (40)
Renal crisis	2 (4)
Pulmonary hypertension	2 (4)
Lung fibrosis	1 (2)
Telangiectasia	32 (64)
Arthritis/arthralgia	29 (58)
Modified Rodnan Skin Score (mean ± sd)	9.2 ± 6.8

Table 3. Nailfold capillaroscopy parameters in all studied patients.

	Mean/N (±SD/%)
Capillary density	7.4 ± 1.2
Capillary length	191.9 ± 31.7
Capillary width	39.4 ± 8.1
Capillary hemorrhage	17 (34)
Capillary shape	
Normal shape	9 (18)
Dilated-mega-capillaries	35 (70)
Tortuous capillaries	6 (12)
Sub-capillary venous plexus	33 (66)
NFC pattern	
Early	9 (18)
Active	17 (34)
Late	24 (48)
NFC score	
Score 0	16 (32)
Score 1	14 (28)
Score 2	11 (22)
Score 3	9 (18)

Table 4. PDUS findings in all studied patients.

PDUS findings	Mean/N (±SD/%)
Ulnar artery	
PSV, cm/s	38 ± 15
EDV, cm/s	3 ± 3.8
UAO	
Unilateral	15 (30)
Bilateral	9 (18)
Total	24 (48)
Pathological FPBF	
Unilateral	8 (16)
Bilateral	21 (42)
Total	29 (58)
Association of UAO and Pathologic FPBF	22 (44)

FPBF, finger pulp blood flow; PDUS, power doppler ultrasonography; UAO, ulnar artery occlusion.

Table 5. Value of UAO in diagnosis of systemic sclerosis stage.

NFC pattern	UAO presented ($N = 50$) [n (%)]	No UAO ($N = 50$) [n (%)]	χ^2	P
Early	1 (2)	23 (46)	33.90	<0.001
Active	1 (2)	8 (16)		
Late	15 (30)	2 (4)		

UAO, ulnar artery occlusion.

Table 6. Value of pathological FPBF in the diagnosis of systemic sclerosis stage.

NFC pattern	Pathological FPBF ($N = 50$) [n (%)]	Normal FPBF (N = 50) [n (%)]	χ^2	P
Early	2 (4)	7 (14)	15.29	0.005
Active	11 (22)	13 (26)		
Late	16 (32)	Stage1 (2)		

FPBF, finger pulp blood flow.

A significant association between the presence of (Raynaud's phenomena, Pitting scar, Digital ulcer, acroosteolysis, and Calcinosis) and US/NFC findings ($P < 0.05$). However, No significant association between Pathological FPBF and calcinosis.

4. Discussion

A few articles have featured the importance of Doppler ultrasound in evaluating the seriousness of hand contribution and computerized vasculopathy in SSc.[8] Specifically, these articles have expanded the comprehension that ultrasound can assist better with describing macrovascular sores. Even though microvascular changes have been widely portrayed, macrovascular harm is likewise normal in SSc. PDUS could likewise be a dependable instrument for evaluating finger mash blood stream (FPBF).[9] Few examinations have inspected the relationship between macrovascular harm surveyed by PDUS and microvascular harm evaluated by NFC.[10]

The point of this work was to assess the relationship between microvascular harm evaluated by NFC and macrovascular highlights surveyed by Doppler US to survey the seriousness of hand contribution and vasculopathy in SSc.

In this review, 38 (76%) patients had RP, 27 (54%) patients had a cut injury, and 17 (34%) had a computerized ulcer. Two (4%) patients had a past renal emergency, while two (4%) had progressed pneumonic hypertension and one of them had muddled aspiratory fibrosis. Telangiectasias were distinguished in 32 (64%) patients and 29 (58%) patients experienced arthralgias. The mean altered Rodnan skin score was 9.2 ± 6.8.

This is to some extent predictable with the examination of Elsayed *et al.*[11] Raynaud's side effects happened in 94% of patients, trailed by finger expanding in 78%, telangiectasia in 74%, cicatricial ulcers in 72%, computerized tuberculous ulcers in 68%, and negligible calcinosis in 32% of patients. Cutaneous signs were followed similarly (90%) by established, gastrointestinal, outer muscle, and neuropsychiatric appearances, trailed by aspiratory and cardiovascular indications in 88 and 40% of cases, separately; notwithstanding, renal signs were the most un-normal in 20% of cases.

As per the after effects of NFC, nine (18%) patients were determined to have beginning phase SSc, while 24 (48%) patients were in the cutting edge stage and 17 (34%) patients had a dynamic illness.

In the current review, patients with calcinosis and acroosteolysis were bound to have a late NFC profile, which was steady with Morardet *et al.*[12]

The rate of OUA in our review was 48%. Moreover, a profoundly huge measurable affiliation was found between late NFC vein design and ulnar course impediment in SSc patients, which is steady with Lescoat *et al.*,[3] They observed that UAO was altogether connected with the late example of NFC and weighty shagginess.

A review directed by Frerix and colleagues,[8] included 80 patients with SSc and 40 controls. Seventeen (21.5%) patients with SS had OAU (11 reciprocal patients), contrasted with none in the benchmark group. Patients with UAO had a fundamentally longer illness span (170 vs. 66 months, $P < 0.001$).

The critical relationship between the presence of (Raynaud's peculiarities, pitting scar, Telangiectasia, joint inflammation) and UAO/pathologic FPBF on PDUS assessment/NFC discoveries. This was in a similar line with study done by Lescoat *et al.*,[3] who found that a pathologic FPBF was related with extreme hairlike misfortune in NFC. Likewise, they observed that A background marked by computerized ulcer and pitting scars was essentially connected with UAO, pathologic FPBF on PDUS assessment, and late example and serious slim misfortune on NFC.

4.1. Conclusion

NFC is a valuable additional tool to evaluate various aspects of SSc microangiopathy with pathological UAO and FPBF, as key diagnostic parameters of PDUS may be a relevant indicator of the severity of vasculopathy in SSc. PDUS can be used in daily practice to identify patients with more severe vasculopathy.

Conflicts of interest

There are no conflicts of interest.

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