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AB0660 SEXUAL HEALTH IMPAIRMENT IN WOMEN WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Idiopathic inflammatory myopathies (IIM) are a group of disorders characterized by skeletal muscle inflammation that can cause functional impairment including sexual dysfunction.

Objectives: To assess sexual function, pelvic floor function and sexual quality of life of women with IIM compared to age-/sex-matched healthy controls (HC). To subanalyze sexual function in sexually active individuals.

Methods: In total 27 women with IIM [mean age: 54.2, disease duration: 7.3 years, dermatomyositis (DM, 10)/polymyositis (PM, 13)/necrotizing myopathy (IMNM, 3)/inclusion body myositis (IBM, 1)], who fulfilled the Bohan/Peter 1975 criteria for DM/PM, and 27 healthy women (mean age: 54.2) filled in 11 well-established and validated questionnaires assessing sexual function, pelvic floor function, quality of life, fatigue, physical activity and depression. Data are presented as mean±SEM.

Results: Compared to HC, patients with IIM had significantly higher prevalence and greater severity of sexual impairment (FSFI, BISF-W), dysfunction of pelvic floor (PISQ-12), and worse sexual quality of life (SQoL-F) (table 1). There were no significant differences in sexual function between PM and DM. Even sexually active IIM patients reported significantly greater sexual health impairment compared to sexually active HC. Sexual health impairment in IIM was associated with laboratory markers of disease activity, health status, physical activity, fatigue and depression (table 2).

Table 1.

Questionnaire: score range	IIM (n=27)	HC (n=27)	p-value	IIM SA (n=16)	HC SA (n=16)	p-value
FSFI :2(worst)-36 (best)	15.7 ±2.5	23.0 ±2.2	p=0.0296	23.8±2.6	29.6±0.8	ns
BISF-W:-16(worst)-75(best)	15.9 ±3.5	27.4 ±3.5	p=0.0212	25.2±4.3	39.2±2.3	p=0.0151
SQoL-F: 0(worst)-100(best)	21.5 ±4.5	83.3 ±2.9	p<0.0001	31.9±6.4	85.7±3.0	p<0.0001
PISQ-12: 0(best)-48 (worst)	14.0 ±1.0	8.7±0.9	p=0.0007	11.4±1.1	9.2±1.0	ns
PFIQ7: 0(best)-300 (worst)	20.5 ±6.9	6.0±2.1	ns	17.6±10.8	10.4±4.2	ns

Abbreviations: FSFI: Female Sexual Function Index; BISF-W: Brief Index of Sexual Function for Women; SQoL-F: Sexual Quality of Life Questionnaire; PISQ-12: Pelvic Organ Prolapse/Urinary Incontinence Sexual Questionnaire short form; PFIQ7: Pelvic Floor Distress Inventory Questionnaire, SA: sexually active; ns: not significant

Table 2.

Parameter: score range	Correlated parameters	r	p-value
FIS (fatigue): 0(best)-160(worst)	FSFI (sexual function)	-0.406	p=0.0353
	BISF-W (sexual function)	-0.424	p=0.0272
	SQoL-F (sexual quality of life)	-0.562	p=0.0027
	PISQ-12 (pelvic floor function)	0.459	p=0.0157
HAP (physical activity): 0(worst)-94 (best)	FSFI (sexual function)	0.429	p=0.0252
LD	FSFI (sexual function)	-0.411	p=0.0327
	BISF-W (sexual function)	-0.466	p=0.0141
BDI II (depression): 0(best)-63(worst)	PISQ-12 (pelvic floor function)	0.322	p=0.0400
HAQ (health status): 0(best)-3(worst)	PISQ-12 (pelvic floor function)	0.477	p=0.0117
	BISF-W (sexual function)	-0.411	p=0.0329

Abbreviations: FIS: Fatigue Impact Scale; BDI-II: Beck's Depression Inventory II; HAP: Human Activity Profile; HAQ: Health Assessment Questionnaire; LD: lactate dehydrogenase

Conclusion: Women with IIM reported significantly impaired sexual function, sexual quality of life and pelvic floor function than age-matched HC.

Worse scores in IIM were associated with disease activity/health status, physical activity, fatigue and depression.**Acknowledgement:** Supported by AZV-16-33574A, MHCR 023728.

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AB0661 ASSOCIATION OF ANTI-MDA-5 AUTOANTIBODY WITH AUTOIMMUNE ASSOCIATED HEMOPHAGOCYTIC SYNDROME IN DERMATOMYOSITIS

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Background: Autoimmune associated hemophagocytic syndrome (AAHS) is a rare complication in dermatomyositis (DM). We previously demonstrated by multivariate analysis that one of factors associated with mortality in AAHS is DM (OR 5.57 [95% CI 1.08–28.65], P 0.05) among connective tissue diseases (1).

Objectives: To find out underlying immunological characteristics, we examined the DM patients with AAHS.

Methods: We examined 31 new onset patients with idiopathic inflammatory myopathies (IIM) including clinically amyopathic dermatomyositis (CADM) admitted to our hospital between January 2009 and December 2018. Three patients had been diagnosed as AAHS proven by bone marrow aspiration. We examined these patient clinical and laboratory characteristics.

Results: Two patients were diagnosed as DM and one was CADM. We found that these 3 patients were associated with interstitial pneumonia. Laboratory tests of all 3 patients showed hyperferritinemia and high titer of anti-MDA5 antibody. All patients were diagnosed as AAHS by bone marrow aspiration smears. Two of them died on the 12th and on the 75th hospital day, respectively, in spite of intensive therapies.

	age/sex	diag.	CK (n)	ferritin (n)	antibodies			
					MDA5	Ro	PM-SCL100	others*
1	63/F	DM	8430 (36-216)	59,877 (5-120)	>150	+	-	-
2	50/M	CIDM	32 (59-248)	11,498(23-250)	>150	±	±	-
3	57/F	DM	277 (41-153)	4,876 (5-120)	>150	+	-	-

* examined antibodies against Jo1, SRP, Mi2, Ku, PL-7, OJ, EJ, Tif1-γ.

Conclusion: Anti-MDA5 may relate not only interstitial pneumonia but also AAHS.

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AB0662 **UTILITY OF NAILFOLD CAPILLAROSCOPY FOR THE EARLY DIAGNOSIS OF SYSTEMIC SCLEROSIS SPECTRUM OF DISEASES IN ASIA**

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Background: The utility of nailfold capillaroscopy (NFC) has largely been evaluated in patients with Raynaud's Phenomenon (RP). However, in equatorial countries, RP occurs in only 79% (1) and 84% (2) of patients in Singapore and Malaysia respectively. In the early stages of Systemic Sclerosis (SSc), patients may develop autoantibodies or puffy fingers before the onset of RP (3).

Objectives: There is no data on the utility of NFC in patients with non-RP features suggestive of the scleroderma-spectrum of diseases (SSc, mixed connective tissues disease, dermatomyositis). We aim to compare the utility of NFC for the early diagnosis of the SSc-spectrum of diseases in patients who present with and without RP.

Methods: Patients referred to our institution for NFC evaluation from March 2010 – March 2017 were recruited. Patients with confirmed diagnosis of a connective tissue disease were excluded. Eligible patients were divided into 4 groups: (I) RP with or without positive ANA; (II) Undifferentiated non-RP features to suggest SSc-spectrum diseases and negative ANA; (III) Undifferentiated non-RP features to suggest SSc-spectrum diseases and positive ANA $\geq 1/400$ and (IV) Positive SSc-associated autoantibodies without features to suggest SSc-spectrum diseases. Images were analysed at 200x magnification using a software analysis program. Patients were noted to have normal, aspecific or SSc pattern on NFC, defined by the presence of ≥ 1 giant capillary, avascular areas or >2 dilated capillaries with areas of hemorrhage.

Results: 133 patients were recruited (mean age 55 years, 87% female; 76% Chinese, 7% Malay, 10% Indian, 7% other ethnicity). ANA was positive in 74 (82.2%) patients (n=25 anti-centromere, n=3 anti-Scl70). Taking into consideration NFC patterns, the clinical outcomes were normal in 32 patients (35.6%), connective tissue disease (CTD) suspected in 33 patients (36.7%), and definite CTD in 25 patients (27.8%). Diagnostic yield for SSc-spectrum of diseases was highest in Groups I and III (Table 1).

Abstract AB0662 Table 1.

	Definite SSc-spectrum diseases n (%)	Suspected CTD n (%)	No CTD n (%)
Group I	24 (47.1%)	16 (31.4%)	11 (21.6%)
Group II	5 (31.2%)	5 (31.2%)	6 (37.5%)
Group III	14 (35.9%)	17 (43.6%)	8 (20.5%)
Group IV	2 (12.5%)	9 (56.2%)	5 (31.2%)

Conclusion: In patients without RP, NFC was most useful to confirm a diagnosis in those with undifferentiated features to suggest SSc-spectrum of diseases and positive ANA.

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AB0663 **ASSESSMENT OF HEALTH RELATED QUALITY OF LIFE AND FUNCTIONAL DISABILITY AMONG SCLERODERMA PATIENTS IN BANGLADESH**

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Background: Systemic sclerosis (SSc) is a complex autoimmune disorder that is characterized by overproduction and deposition of collagen in the skin and visceral organs, abnormalities of the microcirculation, and autoimmunity. SSc is responsible for skin, tendon, joint, and vessel damage, which leads to disability, handicap, and worsening of quality of life.

Objectives: The objective of the study was to assess the relationship between health-related quality of life (HRQoL) and functional disability among scleroderma (Systemic sclerosis, SSc) patients.

Methods: A cross-sectional study was conducted among 78 patients in Scleroderma clinic of BSM Medical University (BSMMU). Patients were selected according to American College of Rheumatology/European League Rheumatism (ACR/EULAR) classification criteria for SSc. The validated Bengali version of the Short Form 36 (SF-36) and Health Assessment Questionnaire (HAQ) were used to assess the HRQoL and functional disability respectively.

Results: The mean age of the patients was 36.47

11.28 years and most of them were female (91.0%). Majority of the patients (76.9%) had diffuse whereas 23.1% patients had limited SSc. Out of 78 patients, antibody profiles were available for 63 patients. The frequency of Antinuclear, Anti-topoisomerase and Anticentromere antibody were 58.7%, 38.1%, 3.2% respectively. Sclerodactyly was present in 84.6% patients and 78.2% had pinch nose and 60.3% had calcinosis. Pitting scars of finger tips was present in 67.9% patients. Raynaud's Phenomenon was present in 89.7% patients and Musculoskeletal manifestations was present in 47.4% patients. Among the patients, 78.2% patients had microstomia and 60.3% had esophageal involvement. The mean MRSS was 21.39 $\pm$ 13.02. SF-36 score showed that the mean Physical Component Summary (PCS) and Mental Component Summary (MCS) were 35.65 $\pm$ 23.65 and 41.36 $\pm$ 25.94 respectively. The mean HAQ-DI was found 1.40 $\pm$ 0.95. The PCS of the SF-36 was moderately correlated with the HAQ-DI ($r=0.641$, $p<0.001$) and the MCS of the SF-36 was weakly correlated with the HAQ-DI ($r=0.360$, $p<0.001$) (Spearman's Correlation).

Conclusion: Functional disability significantly affects health related quality of life. Treatment strategies should include multi-disciplinary approach such as physiotherapy, psychotherapy, social support and occupational therapy to reduce the functional disability which will further enhance the HRQoL of SSc patients.

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