Original article

KNOWLEDGE OF MEDICAL SPECIALISTS ON ORAL MANIFESTATIONS OF SYSTEMIC SCLEROSIS IN A TERTIARY HEALTHCARE CENTRE IN NIGERIA

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ABSTRACT

OBJECTIVE: Due to the heterogeneous manifestation and multiple organ affectation associated with systemic sclerosis, a multidisciplinary management approach has been advocated. The aim of this study was to evaluate the level of knowledge of some medical specialists on oral manifestations of systemic sclerosis and the need for interdisciplinary/multidisciplinary collaboration with dentists in a tertiary hospital in the management of patients with this condition.

METHODS: This study was conducted at the University of Benin Teaching Hospital, Edo State. Data was collected using a self-administered questionnaire with a cover paged informed consent form and was distributed among some medical specialists in the departments of internal medicine, family medicine, mental health and ophthalmology. The information collected was analyzed using statistical package for social sciences SPSS for windows version 23.0 software (SPSS Inc. Chicago, IL, USA).

RESULTS: A total of 80 questionnaires were distributed among the physicians in UBTH, of which 50 were completely answered and returned, giving a response rate of 62.5%. Of these 50 respondents, 27 (54.0%) were male and 23 (46.0%) were female. On the grading of the oral manifestation of SSc knowledge, 19(34%) no knowledge, 12(24%) had poor knowledge, 9(18%) had 10(20%) had good knowledge. A total of 24 (48.0%) of the respondents strongly agreed while 18(36%) agreed for a multidisciplinary management with a dentist.

CONCLUSION: A good number of medical specialists from on this study had insufficient to poor knowledge of oral manifestations of systemic sclerosis. This has made collaboration with the dentist almost absent. A multidisciplinary collaborative interaction is encouraged to prevent worsening oral manifestations associated with systemic sclerosis.

Keywords: systemic sclerosis, oral manifestations, knowledge of medical specialists.

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INTRODUCTION

Systemic sclerosis (SSc) also known as Scleroderma is a rare, chronic, multisystem connective tissue disease characterized by microangiopathy leading to inflammation and fibrosis involving skin and internal organs.[1,2] Processes of fibrosis in the skin and internal organs, non-inflammatory vasculopathy of small blood vessels and specific autoantibodies which are not pathogenic, but are in correlation with activity of illness, are hallmark of this condition.[2,3]

The aetiology of systemic sclerosis is largely unknown with factors such as genetics, race, age, infection and environmental implicated.[4] Systemic sclerosis is divided into diffuse cutaneous (dcSSc) and limited cutaneous (lcSSc) forms based on the extent of skin involvement.[1] The prevalence ranges from 150 to 300 cases per million.[2,3] Worldwide distribution has been established with lower prevalence around the northern Europe and Japan and higher prevalence reported in southern Europe, Australia and North America.[3] Ethnicity has been shown to play a role in disease expression and mortality.[5]

In systemic sclerosis, a typical patient develops Reynaud's phenomenon (bluish discoloration of hands on exposure to cold), sclerosis in the hands, face, trunk, lungs fibrosis which can lead to pulmonary hypertension and cardiomyopathy[6]. The kidneys can also be affected resulting in renal hypertension; with all the above signs manifesting singly or in various combinations.[6] Ulceration of the finger tips due to abnormal collagen deposition is also seen. The skin develops a diffuse, hard texture which is difficult to pinch (hide bound skin) and its surface is usually smooth, taut, mask-like facies. Also, the nasal alae become atrophied, leading to mouse facies.[7]

Changes seen in the gastrointestinal tract in systemic sclerosis can span from mouth to the anus. Oral involvement may include microstomia which is due to skin fibrosis on the face that gives the look of "the bird's face" which is one of the characteristic features of patients with SSc.[8] Other changes seen include shortening of tongue frenulum, changes on the gingiva leading to generalized periodontitis, fibrosis of the uvula, fibrosis of salivary glands with associated hyposalivation leading to dry mouth (secondary Sjorgren syndrome) and rampant caries.[9,10] Other oral changes include reduced interincisal distance, missing teeth due to rampant caries, resorption of alveolar bone on radiograph and trigeminal neuralgia.[9,10]

The other oral manifestations include telangiectasia of oral mucosa, increased decayed, missing and filled teeth (DMFT).[7,11,12] The tongue can also become rigid, making speech and swallowing difficult. The soft tissues around the temperomandibular joint can also affected, which results in pseudo-ankylosis. The loss of attached gingiva and gingival recession may also occur. The radiographic findings include uniform widening of the periodontal ligament space, especially around the posterior teeth.[13] Also, the mandible shows varying degree of bone resorption.[13] The mechanism for this osteolysis is unknown. However, it has been postulated that vasculopathy associated with the disease process diminishes the blood supply to the mandible resulting in bone ischemia and necrosis.[14]

Fibrosis in lamina propria, submucosa layer, and muscular layer are seen on histology, with perivascular deposits of inflammatory cells and subsequent ulcerations, excoriations and strictures in mouth and digestive tube which diminish the gastric passage.[15,16, 17]

Ocular involvement in Systemic sclerosis has been widely documented[18,9,20] with great variations in the nature, frequency, and relevance of the ocular changes abounding in literatures. Ocular changes are thought to be the effects of systemic complications of scleroderma or adverse effects of therapy used in its control with these changes involving numerous ocular tissues and occurring at any stage of the disease process.[21] Their course can be clinically latent or very intensive like severe fundal changes in systemic sclerosis with acute terminal renal failure.[22]

Ocular changes which appear to be specifically related to systemic sclerosis include: telangiectasia and dermal sclerosis of the eyelids; a tear defect of varying severity, telangiectasia and sludging of the blood column in the

conjunctival vessels; possibly punctate defects of the iris pigment epithelium and posterior subcapsular lens opacities which have been described as a resultant effect of prolonged steroid therapy.[23] Other possible complications include conjunctival changes[23], Shallowing of the fornices, keratoconjunctivitis sicca,[24] eye dryness,[25] pinguecula and pterygium,[26] episcleritis, scleritis, uveitis, and peripheral ulcerative keratitis,[27,28,29] keratoconus and pellucid marginal degeneration,[30] cataract,[31] glaucoma,[32] refractive error,[33] and optic disk disease.[34]

Due to the heterogenous manifestation and multiple organ affectations associated with systemic sclerosis (SSc), interdisciplinary diagnosis and management have been recommended.[35] The therapeutic measures are not aimed at cure of the Scleroderma disease but are intended to greatly improve the patient's quality of life and should always apply. There is no specific treatment for oral changes in Scleroderma, but changes should be treated along with general treatment of SSc.[36]

The aim of this study was to evaluate the level of physicians' interdisciplinary/multidisciplinary collaboration with dentists in a tertiary hospital in the management of patients with systemic sclerosis.

MATERIALS AND METHODS

Ethical approval for this study was sought and obtained from the Ethics and Research Committee of the University of Benin Teaching Hospital (UBTH), Benin City, Nigeria before the study commenced. This study was conducted at the University of Benin Teaching Hospital, a leading tertiary health and training institution in the southern part of Nigeria.

Data was collected over a period of 2months by the researchers using a self-administered questionnaire with a cover paged informed consent form and was distributed among medical specialists in the departments of internal medicine, family medicine, mental health and ophthalmology. The questionnaires elicited information on their demographics, qualifications, stage of training and area of specialization. Other parameters that were assessed included their level of awareness, knowledge of systemic sclerosis, knowledge of oral manifestation of systemic sclerosis and level of collaboration with the dentist.

The information collected was analyzed using statistical package for social sciences SPSS for windows version 23.0 software (SPSS Inc. Chicago, IL, USA). Categorical data were reported as frequencies and percentages while numerical data were summarized as mean and standard deviation. Knowledge of the oral complications of systemic sclerosis will be graded into none, poor, good and excellent using Chi square statistics and independent t-test and also to determine the statistical significance of any difference in variables. The results of the statistical test were assumed to be significant where P value was less than 0.05 ($p < 0.05$)

RESULTS

A total of 80 questionnaires were distributed among the physicians in UBTH, of which 50 were completely answered and returned, giving a response rate of 62.5%. Of these 50 respondents, 27 (54.0%) were male and 23 (46.0%) were female physicians**.** Majority of the respondents were between the ages of 31 – 40 years of age and they accounted for 25 (50.0%) of the study population. Most of the respondents 45(90%) has less than 10 years of clinical experience except a few 5(10%) who has greater than 10years of clinical experience. Majority of the respondents 25 (50.0%) were internists followed by family physicians 20 (40.0%) while the ophthalmologists and mental health physician make up the remaining of the respondents (Table 1).

Concerning their knowledge of systemic sclerosis, 41 (82.0%) of the respondents were aware of the disease with only 14 (28.0%) opportune to have managed at least a case of systemic sclerosis. Among the respondents who had managed systemic sclerosis, 9 (18.0%), 3 (6.0%) and 2 (4.0%) had managed 2, 1 and 3 cases respectively. The respondents 2(12%) reported an incidence of 1 case per year while

2(4.0%) reported 2 case of Ssc per year (Table 2).

Greater than half of the respondents 28 (56.0%) in this study reported to be aware of the complications associated with systemic sclerosis, with majority 24(48%) pointing at dry mouth followed by gingival diseases 19 (38.0%). The least reported complication of Ssc was tooth decay 11 (22.0%).

Table 1: Socio-demographic characteristics of the respondents

	N	%
Gender		
Male	27	54.0
Female	23	46.0
Age group (years)		
24-30	19	38 .0
31-40	25	50.0
41-52	6	12.0
Area of Specialty		
Family Medicine	20	40.0
Internal Medicine	25	50.0
Mental Health	1	2.0
Others	4	8.0
Years of Experience		
1-10	45	90.0
11-20	3	6.0
21-30	2	4.0
TOTAL	**50**	**100.0**

Table 2: Knowledge/prevalence of systemic sclerosis

	N	%
Knowledge of systemic sclerosis		
Yes	41	82.0
No	9	18.0
Ever managed or been involved in the management of a case of systemic sclerosis		
Yes	14	28.0
No	36	72.0
Number of cases of systemic sclerosis managed		
Nil cases	36	72.0
1	3	6.0
2	9	18.0
3	2	4.0
Yearly diagnosis of new cases		
Nil	42	84.0
1	6	12.0
2	2	4.0

On examination of patients, only 6 (12.0%) of respondents admitted examining the oral cavity of systemic sclerosis patients for oral manifestations of the disease. Dry mouth 8 (16.0%), tongue fibrosis 7 (14.0%), temporomandibular joint ankylosis 7 (14.0%) were the most seen with the least being tooth decay 3 (6.0%). When asked for reasons for not examining the mouth, 8 16.0%) of the respondents reported that the patient didn't complain of any symptom from the mouth and 2 (4.0%) admitting to not knowing the oral manifestations of systemic sclerosis. Only one of the respondent didn't see examination of the mouth important. Among the respondents who have managed the disease, only 2 (4.0%) had referred their patient to a dentist for co-management (Table 3).

Table 3a – Knowledge of oral manifestations

	N	%
Awareness of the oral complications of systemic sclerosis		
Yes	28	56.0
No	22	44.0
Oral complications (n = 28)		
Tooth decay	11	39.3
Gingival diseases	17	60.7
Dry mouth	21	75.0
Bad breath	16	57.1
Poor oral hygiene	16	57.1
Reduced mouth opening	16	57.1
Tongue fibrosis	16	57.1
Resorption of the mandible	10	35.7
Temporomandibular joint ankylosis	14	50.0
Ever examined the oral cavity of a patient with systemic sclerosis?		
Yes	6	12.0
No	44	88.0
Oral features seen (n = 6)		
Tooth decay	1	16.7
Gingival diseases	2	33.3
Dry mouth	5	83.3
Bad breath	2	33.3
Poor oral hygiene	4	66.7
Reduced mouth opening	5	83.3
Tongue fibrosis	4	66.7
Resorption of the mandible	2	33.3
Temporomandibular joint ankylosis	4	66.7

A total of 24 (48.0%) of the respondents strongly agreed while 18(36%) agreed for multidisciplinary management with a dentist. On the need to immediately refer Ssc patients to the dentists at presentation 17 (34.0%) strongly,

20(40%) agreed while the remaining respondents are either undecided or totally disagree. When asked if multidisciplinary team approach could improve outcome and better quality of life of the patient 26 (52.0%) responded positively. On the need for interdepartmental seminars and joint clinical meetings, 27 (54.0%) and 24 (48.0%) respectively strongly agree. Using social media platforms for interactive sessions for better management of these patients, 24 (48.0%) of the respondents strongly agrees. (Table 4).

Table 3bThe maximum knowledge score attainable = 10.
Grading: 0 (None), 1 – 4 (poor), 5 – 6 (good), 7 – 10 (Excellent)

Levels of knowledge	Frequency	Percent
None	19	38.0
Poor	12	24.0
Good	9	18.0
Excellent	10	20.0
Total	50	100.0

Table 4-Test for associations between levels of knowledge on oral complications and age, sex, and years of practice

	Levels of Knowledge on Oral Complications					
	None	Fair	Good	Excellent	Total	Fishers
	n (%)	n (%)	n (%)	n (%)	n (%)	P-value
Sex						0.820
Male	9 (33.3)	7 (25.9)	6 (22.2%)	5 (18.5)	27 (100.0)	
Female	10 (43.5)	5 (21.7)	3 (13.0)	5 (21.7)	23 (100.0)	
Years in practice						
0 – 10	17 (37.8)	12 (26.7)	8 (17.8)	8 (17.8)	45 (100.0)	0.396
11 – 20	1 (33.3)	0 (0.0)	0 (0.0)	2 (66.7)	3 (100.0)	
21 – 27	1(50.0)	0 (0.0)	1 (50.0)	0 (0.0)	2 (100.0)	
Awareness of SS*						
Yes	11 (26.8)	12 (29.3)	8 (19.5)	10 (24.4)	41 (100.0)	**0.005****
No	8 (88.9)	0 (0.0)	1 (11.1)	0 (0.0)	9 (100.0)	
Ever managed SS*						
No	3 (21.4)	4 (28.6)	3 (21.4)	4 (28.6)	14 (100.0)	0.459
Yes	16 (44.4)	8 (22.2)	6 (16.7)	6 (16.7)	36 (100.0)	
Age group						
	7 (36.8)	4 (21.1)	4 (21.1)	4 (21.1)	19 (100.0)	0.950
	10 (40.0)	7 (28.0)	3 (12.0)	5 (20.0)	25 (100.0)	
	2 (33.3)	1 (16.7)	2 (33.3)	1 (16.7)	6 (100.0)	

SS* = systemic sclerosis ** = There was an association

Table 5 – Practice of multidisciplinary approach to management with the dentist

Need for multidisciplinary management	n (%)				
	Strongly agree	Agree	Undecided	Disagree	Strongly disagree
There is need for multidisciplinary management of a patient with systemic sclerosis with the dentist	24 (48.0)	18 (36.0)	7 (14.0)	0 (0)	1 (2.0)
There is need to immediately refer a patient with systemic sclerosis to a dentist for prevention/management of oral complications	17 (34)	20 (40)	11 (22.0)	1 (2.0)	1 (2.0)
Multidisciplinary management of a patient with systemic sclerosis with the dentist can improve patient quality of life	26 (52.0)	19 (38.0)	5 (10.0)	0 (0)	0 (0)
Collaborative efforts like imterdepartmental seminars can help improve management of patients with systemic sclerosis	27 (54.0)	16 (32.0)	6 (12.0)	1 (2.0)	0 (0)
Joint clinical meetings and reviews of patients with systemic sclerosis can help improve patient's quality of life	24 (48.0)	19 (38.0)	7 (14.0)	0 (0)	0 (0)
Multidisciplinary interactive sessions using social media platforms can help in new information dissemination and management of systemic sclerosis patients	24 (48.0)	17 (34.0)	9 (18.0)	0 (0)	0 (0)

DISCUSSION

Recent studies on systemic sclerosis have estimated the prevalence to be 150-300 cases per one million adult patients.[2,3] Because of this rarity, many primary physicians (including the rheumatologists, dermatologists, ophthalmologists, family physicians and dentists) may only interact with very limited number of systemic sclerosis all through their career.[37] Morbidity and mortality have often been associated with multiple internal organ involvement which can present as a nonspecific symptoms organ failure.[17]

The result of this study showed that most of the physicians who participated in the study have insufficient knowledge of the disease and its associated complications though overwhelming majority 41(82%) are aware of systemic sclerosis. This is consistent with the study by Distler et al,[38] who reported that lack of disease awareness in dcSc patients and inadequate knowledge among the primary physicians as the cause of delayed referral and mitigation of disease progression. It is also of note that a statistically significant proportion of the physician 22(44%) who participated in this study are also unaware of the oral manifestation of systemic sclerosis. Of the physician who reported to be aware of the oral manifestation of systemic sclerosis, only 6 (12%) has ever bothered to examine the oral cavity, therefore consultation and referral to the dentist and other specialist is almost not available to the patients. Thus programmes geared towards improved awareness among primary care physicians is necessary to facilitate diagnosis, early referral and appopriate intervention.[8,12,27,38]

On the concept of multidisciplinary practice and collaborative interactions, majority of the respondents agrees with the need for interactions with the dentist to manage the patients, prevent worsening of the oral manifestations, and improve the patients' quality of life.[37,38] Opinions were high on the need for interdepartmental seminars /reviews and joint clinical meetings among the respondent as most agree that it will improve management, clinical outcome and the overall impact on the systemic sclerosis patients. Multidisciplinary interactive sessions using the social media was also strongly advocated, as it can help in new information dissemination and management of systemic sclerosis patients. It is therefore our opinion that efforts toward this should be strongly recommended

Furthermore, establishing a specialist Centre with multidisciplinary care may help promote such changes and improve treatment outcomes.[38]

Conclusion: From this study it can be concluded that most primary care physicians have insufficient knowledge of oral manifestations systemic sclerosis. This has made collaboration with the dentist almost absent. We therefore suggest that multidisciplinary collaborative interactions be encouraged to prevent worsening oral manifestations, allow early intervention and improve patients' quality of life.

No conflict of interest declared.

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