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STUDY OF CLINICAL PROFILE OF CONNECTIVE TISSUE DISORDERS

Medicine

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ABSTRACT

INTRODUCTION: Connective tissue disorders include inherited as well as autoimmune disorders like SLE, RA, MCTD, Scleroderma, Sjogren syndrome etc. Individuals with autoimmune diseases may have numerous antibodies which eventually lead to multiorgan involvement including musculoskeletal, cutaneous, renal, cardiopulmonary, neurological involvement. The main stays of management remains glucocorticoids and immunosuppressants.

METHODOLOGY: A one year cross sectional study was conducted in the Guru Gobind Singh Hospital, Jamnagar. 50 patients of connective tissue disorders fulfilling the inclusion criteria were included in the study.

RESULTS: In our study, the highest prevalence was noted for SLE ~ 36% followed by RA ~24. We found that the most common presentation of these disorders was constitutional and musculoskeletal features and the least common being neurological features. We also found an association between CTDS in females and adverse pregnancy outcomes.

KEYWORDS

Connective Tissue Disorders, Anti-nuclear antibodies

INTRODUCTION

Connective tissue disorders refer to group of disorders involving the protein rich connective tissue that supports organs and other parts of the body. The group includes both Inherited (eg. Ehler danlos syndrome, marfans syndrome, osteogenesis imperfecta) as well as autoimmune diseases (eg. SLE, rheumatoid arthritis, scleroderma, sjogren's syndrome, MCTD etc).This study will exclude the inherited ones as they mostly present in the pediatric age group.

SLE is one of the most common autoimmune disease in women during their child bearing age. The peak incidence being around the age of 15-40 years with female:male ratio of 6-10:1.

Diagnosis: ANA is positive in >98% of cases specifically Anti dsDNA and Anti-smith antibody.

Clinical features: Apart from constitutional symptoms, the most common manifestations are of musculoskeletal and cutaneous origin including myalgia, photosensitivity, malar rash(typical butterfly shaped rash over cheek and nasal bridge), discoid rash, alopecia, polyserositis (pleural/pericardial effusion/ascites), pancytopenia, lupus nephritis, seizures etc. Management options include glucocorticoids plus immunosuppressants like cyclophosphamide/ mycophenolate mofetil/ azathioprine.

Rheumatoid Arthritis is a chronic inflammatory disease of unknown etiology marked by symmetric peripheral polyarthritis which may or may not be associated with extra articular manifestations

AIMS AND OBJECTIVES

1. To study the clinical presentation and physical findings of newly diagnosed as well as known cases of connective tissue disorders.
2. To study spectrum and magnitude of different systems getting involved in different connective tissue disorders.

MATERIALS AND METHODS

This is a one year observational cross sectional study conducted in the Guru Gobind Singh Hospital, Jamnagar.

Data was collected from the patients presenting to department of internal medicine at M.P.SHAH Medical college, Guru Gobind Singh Hospital, Jamnagar, fulfilling inclusion criteria. Duration of study was one year in the Period of September 2016 to September 2017 with sample size of 50.

After history and clinical examination, patients underwent routine blood tests like CBC, Sr. Creatinine, Urine Microscopy and other tests needed for the study like ANA Profile, RA Factor, Thyroid Profile, Imaging according to the requirement.

Data was analyzed by descriptive statistics and calculated as percentages and presented by using table, bag graph etc

RESULTS

Table - 1 : Demographic Characteristics Age at the onset of disease

Age group	No. of pts.	%
< 20	4	8%
21-40	27	54%
41-60	17	34%
>60	2	4%
Total	50	100%

Sex Distribution

Sex	No. of pts.	%
Female	42	84%
Male	8	16%
Total	50	100%

Table - 2 : Profile of Connective Tissue Disorders

Name of Disease	No. of pts.	%
SLE	18	36%
RA	12	24%
MCTD	10	20%
SS	8	16%
Others	2	4%
Total	50	100%

18 out of 50 cases (i.e. 36%) were of SLE which makes it the most prevailing CTD among all. The next highest prevalence was noted for RA (12 out of 50 patients i.e. 24%) followed by MCTD and Systemic sclerosis around 20% and 16% respectively.

Table - 3 : Different System Involvement in CTDS

System Involved	Number of cases					Total No. Pts.	%
	SLE	RA	MCTD	SS	Others		
Hemato-logical	15	7	9	6	2	39	78%
Musculo-skeletal	13	9	5	7	2	36	72%
Cutaneous	16	0	1	4	1	22	44%
Cardio-pulmonary	5	2	5	6	1	19	38%
Renal	5	0	3	1	0	9	18%
Neuro-logical	2	0	2	0	0	4	8%

Most commonly involved system was haematological, getting involved in around 78% of patients. The next most commonly involved system is musculoskeletal (72%) followed by cutaneous (44%), cardiopulmonary (38%), renal (18%) and least involved system was nervous system (8%).

Table - 4 : Adverse Pregnancy Outcomes in CTDS

Adverse Pregnancy Outcomes	No. of pts.	%
Preterm labour	3	30%
IUFD / SFD	2	20%
PIH	3	30%
Recurrent abortion	2	20%
Total	10	100%

Above table showed that 10 out of 42 (i.e. 24%) females in the study had met with adverse pregnancy outcome in form of preterm labour, IUFD, spontaneous abortion, PIH.

Table - 5 : Immunological Profile

Antibodies	No. of pts.	%
ANA	48	96%
ANTI dsDNA	26	52%
ANTI-SMITH Ab	6	12%
ANTI SmRNP/U1RNP	28	56%
ANTI SSARo/SSBRo	16	32%
ScL-70	9	18%
Antiphospholipid Ab	0	0%
Other like Anti-RBC/Anti-platelet	0	0%

ANA to be the most commonly occurring antibodies (96%) followed by RNP (56%), ANTI ds DNA (52%) then SSARo/SSBRo (32%).

DISCUSSION

Over the time many studies have been carried out to detect prevalence and clinical profile of CTDs. We compared data from our study with other two similar studies. One conducted in Western India and other one in Port Harcourt, Nigeria.

All the three studies were having maximum prevalence in the age group 21-40 years. Prevalence in Females was 84% v/s. 91% in Western India study and 92% in Port Harcourt, Nigeria. We found that frequency of different system getting involved in CTDS was quite similar among three studies with most frequently involved system in our study was haematological 78% v/s 60% and 52% in Western India and Port Harcourt, Nigeria respectively followed by musculoskeletal 72% in our study v/s. 56 and 42% in Western India and Port Harcourt, Nigeria respectively and then cutaneous 44% in our study v/s. 52% and 30% in Western India and Port Harcourt, Nigeria respectively.

ANA was consistently present in the maximum percentage of cases in all the three studies i.e. 96%, 98% and 75% in present study, Western India and Port Harcourt, Nigeria study respectively.

We found association between CTDS and adverse pregnancy outcome, to be 18% in our study whereas 10% and 14% in Western India study and Port Harcourt, Nigeria study respectively.

Our study has some limitations as it is a single centre study with small sample size so it is difficult to generalize result on all Indian Population.

Thus we concluded from our study that connective tissue disorders are multisystem involving autoimmune disorders, most common in the 2nd to 4th decade of life with female preponderance, presenting commonly as haematological or musculoskeletal manifestations and also associated with adverse pregnancy outcomes. Most of them are ANA positive and responds to steroids and immunosuppressants.

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