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EAR-NOSE-THROAT AND OCULAR MANIFESTATIONS IN PATIENTS WITH CONNECTIVE TISSUE DISORDERS

ENT

Dr.Ashish sawhney

Dr.Rohini Choudhary

ABSTRACT

Background- A variety of connective tissue disorders are known that produce different ophthalmic and ENT(ear, nose, throat) manifestations among individuals in different age groups that should be studied so that better monitoring and timely management of such manifestations can be done. **Aim-** To evaluate patients attending the eye and ENT OPD, presenting with ocular or ear-nose or throat complaints and having history (laboratory confirmed cases) of connective tissue disorder. **Material and method-** 52 patients of both genders, who attended our eye and ENT outpatient department with ocular or ENT complaints and had history of connective tissue disorder were examined, visual acuity, slit lamp examination, fundus examination, rhinoscopy, throat and ear examination was performed. **Results-** Majority of the patients presented with ophthalmological findings of uveitis followed by dry eyes and epscleritis. Among ENT findings, most patients had oral ulcers followed by dry mouth, hearing loss and sinusitis. **Conclusion-** Patients presenting with ocular or ENT symptoms associated with connective tissue disorders should be thoroughly evaluated and should be managed early to prevent long term complications.

KEYWORDS

Connective tissue, uveitis, oral ulcers, dry eye, scleritis, sinusitis

INTRODUCTION

Connective tissue disorder refers to a group of disorders that involve protein-rich tissues in body that support organs and other parts of the body. In addition to cutaneous, pulmonary, renal, and neurologic involvement, connective tissue disorders can also affect the eyes, nose, mouth and throat.[1]These connective tissue disorders include systemic lupus erythematosus, Sjögren's syndrome, localized scleroderma and systemic sclerosis, wegner's granulomatosis, sarcoidosis, systemic lupus erythematosis, ankylosing spondylitis, polyarteritis nodosa, dermatomyositis, giant cell arteritis, behcet's disease, relapsing polychondritis and rheumatoid arthritis. Ocular involvement can be the initial sign of certain systemic diseases and warrant further investigation and systemic treatment. The ocular immune process is mediated mainly by T helper cells 1 (TH1). It can be TH1 predominant with proinflammatory cytokines or antibody mediated through TH2 cells.[1] Systemic lupus erythematosus (SLE) is an autoimmune disease which is associated with presence of autoantibodies, that can potentially involve multiple body organs including the skin, brain, eyes and kidneys. Ocular involvement in SLE includes dry eyes, decrease visual acuity, keratoconjunctivitis sicca, keratitis, episcleritis, scleritis, ocular pain and rarely optic neuropathy.[2] Retinal vasculitis is the most severe ocular complication and presence of retinopathy is a poor prognostic factor.[3] Sjögren's syndrome is an autoimmune disease, where the lacrimal gland and major and minor salivary glands are affected causing dryness of the mucous membranes. It causes corneal and conjunctival xerosis, corneal scarring, thinning of cornea and recurrent corneal infections, xerostomia and less common findings include Raynaud's phenomenon, purpura and urticarial lesions. Immune mediated destruction of the exocrine glands results in decreased tear production by the lacrimal glands, also referred to as secondary Sjogren's syndrome.[4] Systemic sclerosis (SS) or scleroderma is a chronic multisystem disorder of idiopathic origin affecting skin along with involvement of other body organs including the lungs, kidneys, esophagus, ear and intestine. Lid stiffness, lagophthalmos, conjunctival vascularization, telengectasias, iris pigment epithelium defects and retinopathy are ocular features and sensorineural hearing loss, dry mouth, dysphagia are the ear and throat findings.[5] Rheumatoid arthritis is the most common autoimmune disease that affects cornea.[1] Scleritis and episcleritis are the second most common ocular manifestation of rheumatoid arthritis.[6,7] Dermatomyositis, polymyosistis and many other connective tissue disorders also present with variable ENT and ocular manifestations.[8,9] In this study we aimed at evaluating ocular findings in patients with connective tissue disorders presenting to our eye and ENT out-patient department (OPD).

MATERIALAND METHODS-

This study included 52 patients who had history of connective tissue disorder, out of which 38 patients presented to eye OPD with ocular complaints and 14 presented in ENT OPD. Inclusion criteria- any age group, both gender, history of connective tissue disorder, ocular or ENT complaints associated with these disorders. Exclusion criteria- Patients with other ocular or ENT conditions not related to connective tissue disorder, patients not willing to participate in the study.

All the patients with history of connective tissue disorder and were investigated and lab confirmed for any connective tissue disorder were included in the study. Thos who presented to eye OPD with ocular complaints underwent complete ocular examination including visual acuity, slit lamp and fundus examination. Patients presenting in ENT OPD also underwent detailed examination including ear, nose and throat examination. All the findings were recorded and tabulated in MS Excel spread sheet, different presentations in various connective tissue disorders were noted and data was represented in the form of mean, number and percentages.

RESULTS

Patients who presented to the OPD were in the age range of 18 to 64 years with a mean of 46±5.66 years. Among these 52 patients, 34 patients presented to eye OPD with ocular complaints and 14 presented in ENT OPD, and 4 presented with both ocular and ENT complaints, out of which, 32 were females and 20 were males. The demographic characteristics are given in the Table 1 below.

Table 1: Demographic characteristics of patients

Age range	Number of patients	Percentage
1-20 years	2	3.85%
21-40 years	18	34.61%
41-60 years	32	61.54%
Gender		
Male	20	38.46%
Female	32	61.54%

The clinical findings observed in patients diagnosed with different connective tissue disorders were noted and tabulated as given in Table 2. Majority of the patients presenting in Eye OPD had complaints of blurred vision or congested eyes which on examination revealed uveitis (16 patients), while others complained of dry eyes, episcleritis (Image 1) and chorioretinal lesions in decreasing frequency.

Table 2: Ocular findings in patients with different connective tissue disorders

Ocular finding	Connective tissue disease	No. of patients
Uveitis	Rheumatoid arthritis	7
	Behcet's disease	6
	Ankylosing spondylitis	3

Dry eyes	Rheumatoid arthritis	4
	Systemic lupus erythematosus(SLE)	3
	Sarcoidosis	2
	Systemic sclerosis	1
Episcleritis	Rheumatoid arthritis	4
	SLE	3
	Ankylosing spondylitis	2
Chorio-retinal lesions	Sarcoidosis	2
Retinal vasculitis	Behcet's disease	1

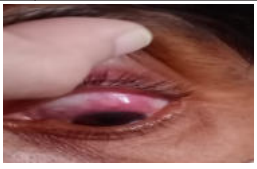

Image 1: Showing sectoral episcleritis in a patient with rheumatoid arthritis

The Patients who presented to ENT OPD who reported symptoms were thoroughly examined and findings were noted. Majority of the patients presented with recurrent oral ulcers (7, 38.8%), followed by dry mouth, vertigo and hearing loss, temporo-madibular joint stiffness and chronic sinusitis. The table 3 depicts various ENT findings in these patients.

Table 3: ENT (ear, nose, throat) findings in these patients

ENT findings	Connective tissue disorder	No. of patients
Oral Ulcers	Behcet's disease	7
	SLE	2
Dry mouth (xerostomia)	Sjogren's syndrome	2
	Sarcoidosis	1
Vertigo/ Sensorineural hearing loss	SLE	2
Chronic sinusitis	Wegner's granulomatosis	1
	SLE	1
Temporo-mandibular joint stiffness/ Trismus	Rheumatoid arthritis	2

The patients who had both ocular and ENT complaints (4 patients) had Sarcoidosis and behcet's disease.

DISCUSSION

Many patients with connective tissue disease can present with multiple system involvement. Ocular and ENT involvement has been known and studied earlier. In our present study, we tried to determine the frequency of eye and ENT involvement among connective tissue disorder patients. A total of 52 patients were included in our study, out of which 34 had ocular complaints, 14 had ENT complaints and 4 had both ocular and ENT complaints. The most frequent ocular symptom observed was uveitis and most common ENT symptom observed was oral ulcers. Ocular and Ear-nose-throat (ENT) manifestations of connective tissue disorders represent a diagnostic challenge for treating clinicians as they often constitute the initial sign of an otherwise asymptomatic autoimmune disease.[10] Such cases usually remain undiagnosed and are kept on symptomatic treatment only. So knowledge of these presentations in various organ systems is a must to timely diagnose and appropriately treat the condition. Our study also included less number of subjects because many patients remain undiagnosed and only those with diagnosed disease were included in our study. Gusmão *et al*, in their study performed clinical and complete otorhinolaryngological evaluations in patients with rheumatic disease and observed many ENT findings and stressed upon further evaluation of such patients.[11,12]Understanding various forms of ocular involvement in these conditions is important as untreated cases can develop severe vision loss.[13] Santoro *et al*, also studied about ocular involvement in connective tissue disorder and observed redness, ocular pain, dryness and vision loss, all to be associated with these disorders.[14] Many other studies have been done to evaluate ENT and ocular manifestations of various connective tissue diseases as their knowledge is important for treating clinicians for timely diagnosis and preventing life threatening complications.[15,16] Inflammation in these connective tissue disorders can affect any part of the eye, starting from the cornea anteriorly to the retina, uveal tract and sclera posteriorly.[10] These manifestations should be known to the ophthalmologist for timely diagnosis and management. Most of the patients remain undiagnosed until late and these symptoms when present if investigated promptly can help in diagnosing connective tissue disorders in such cases. Our study included less number of patients as majority of the patients presenting with such complaints in the OPD's remain undiagnosed which may be due to lack of patient's interest in getting tested or non compliance as most of the symptoms are mild and improve on symptomatic treatment.

CONCLUSION

Early diagnosis of connective tissue diseases can help in improving the patient's quality of life, timely treatment and prevention of life threatening complications. So whenever a patient presents with some ocular or ENT symptoms that could be associated with such disorder, he/she must be thoroughly examined and investigated for connective tissue disorders and record of their findings should be made for future prognosis and management.

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