ORIGINAL RESEARCH PAPER

Otolaryngology

A RARE CAUSE OF DYSPHAGIA: SCLERODERMA

KEY WORDS:

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ABSTRACT

Scleroderma affecting 30 years old female will be discussed in this case report. Wide range of diagnostic modalities and system wise approach help to achieve confirmation of the disease and early intervention if needed. Ear, nose and throat symptoms give necessary vision required for investigative and surgical treatment.

INTRODUCTION

Scleroderma is a connective tissue disorder, of which skin thickening is a classical hallmark. Depending upon cutaneous, contribution the disease can be diffuse (diffuse cutaneous SS) or limited (limited cutaneous SS) or systemic sclerosis sine scleroderma or systemic sclerosis in overlap. Systemic scleroderma is a multisystem autoimmune disease with microvascular anomalies, excessive collagen fibers I & III production and deposition, fibrosis of skin and internal organs, with advanced progression and potentially fatal consequences[1]. The small vessels present fibrosis and peri vascular cellular infiltration with activated T cells. Its prevalence rises to approximately 200 cases per 1 million individuals; risk is 4 to 9 times higher among women than men, with age of onset at 30-50 years[2]. Ear, nose and throat (ENT) manifestations are an important hallmark of systemic scleroderma.

CASE REPORT

A 30 years old female patient came to the OPD of the ENT department with complaints of difficulty in swallowing since 1 month, difficulty in opening mouth since 2 years. Patient was not a known case of diabetes mellitus, bronchial asthma, hypertension, koch's and koch's contact. She denied history of tobacco chewing and cigarette smoking.

On Systemic Examination,

Cardio Vascular System: S1, S2 (+), No murmurs.
Respiratory System: Bilateral air entry equal, normal vesicular breath sounds.
Per abdomen: Soft
Central nervous system: no focal neurological deficit.

On Local Examination,

Oral cavity: trismus (+), hyperpigmented lips and angle of mouth, halitosis (+), dental caries (+). On video laryngoscopy: bilateral vocal cord- mobile.

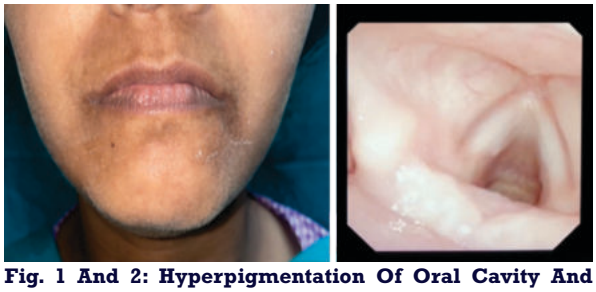

Fig. 1 And 2: Hyperpigmentation Of Oral Cavity And Laryngoscopy Image

Ear: Tympanic membrane bilaterally intact (+). Pure tone audiometry- Right sensorineural hearing loss.

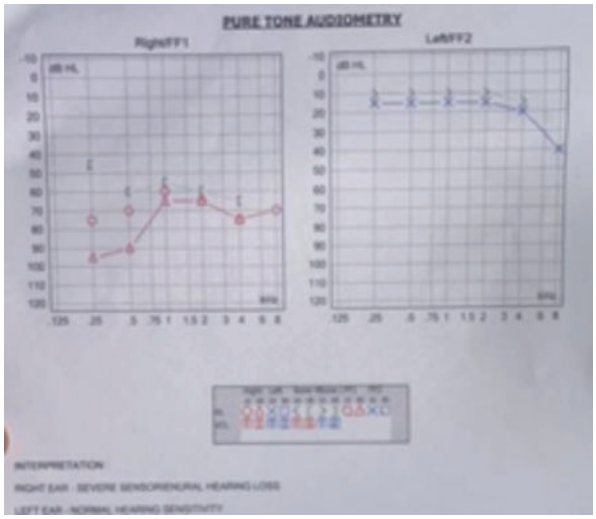

Fig. 3: Shows Pure Tone Audiometry

Evidence of skin features: focal patches of sclerosis over forehead, back, forearm, hand and perioral region.

Fig. 4, 5 And 6: Skin Changes Of Various Body Parts

ECG: within normal limit.
Chest Xray: NAD.

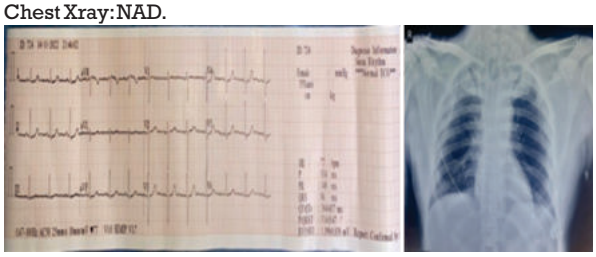

Fig. 7 And 8: Ecg And Chest Xray To Rule Out Pulmonary Hypertension

Cardiac MRI: NAD.
Pulmonary function test: NAD.
Barium swallow xray: showed esophageal stricture.

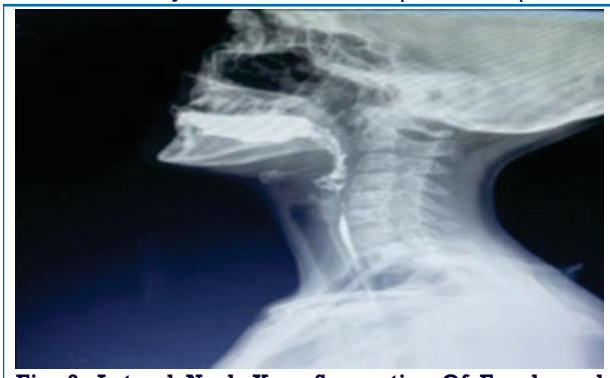

Fig. 9: Lateral Neck Xray Suggestive Of Esophageal Stricture.

Blood Reports:
SCL-70- 5.059
Antinuclear antibody- 3.93

On gatroesophagoscopy, narrowing of esophageal opening from cricopharynx to gastro-esophageal junction with mild fundal mucosal atrophy with mild antral gastritis.

Esophageal dilatation was performed in 2 sittings;
1st sitting: Dilatation were performed with Bougie no. 16 and 18.
2nd sitting: Dilatation were performed with bougie no. 26 and 28.

After consulting a dermatologist and rheumatologist, a combined therapy of oral corticosteroid, pentoxifylline, antacids and multivitamins was started and advised regular repeated follow ups.

DISCUSSION

Scleroderma is an autoimmune disease affecting various organs. ENT manifestations of scleroderma are rare but evident. cobblestone appearance due to pearly white plaque may found in esophageal epithium and subepithelium may show fibrosis[3]. Gastrointestinal disturbances such as acid reflux, dysphagia, foreign body sensation, change in voice or dyspnoea are infrequently the earliest features of the disease. Dysphagia, started by several abnormal swallowing sensation, is primarily caused by impaired esophageal motility but later can consequence from gastroesophageal reflux disease and secondary stricture formation[4].

Sensorineural hearing loss (SNHL) in association with increased inflammatory markers in the no infection features should aware otolaryngologists to the possibility of autoinflammatory diseases and its complications[5].

CONCLUSION

Autoimmune diseases that present wide spectrum of ENT manifestations. A high possibility of suspicion is required for timely intervention. ENT specialists have significant role in referring patients with possible inflammatory disease to rheumatologist and dermatologist. This case report of a patient with typical features of scleroderma provides therapeutic modalities with only temporary improvement.

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