

Case Report

A rare cause of dysphagia: CREST syndrome

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ABSTRACT

Reporting a case of a 29-year-old female patient who came to the outpatient department with features of CREST syndrome. This case report illustrates the ENT presentation of CREST syndrome. A 29 year female with features of CREST syndrome (Calcinosis+, Raynaud's phenomena +, oesophageal dysmotility+ sclerodactyly+ and Telangiectasias+, with no pulmonary hypertension) with ANA titres positive. Complete blood count, serum electrolytes, renal function tests, liver function tests, chest X-ray, barium swallow, ECG was done. Rigid oesophagoscope was used for the bougie dilatation of the stricture of oesophagus and was conservatively treated for her symptoms and being followed up at present. GI disturbances such as heartburn, dysphagia or respiratory complaints. Example: dyspnoea are occasionally the first manifestations of the disease. Dysphagia, manifested by various abnormal swallowing sensation, is initially caused by impaired oesophageal motility but later can result from gastroesophageal reflux disease and secondary stricture formation. Hereby concluding that any patient coming in OPD with similar features as mentioned above we should have a differential diagnosis of CREST syndrome and limited cutaneous scleroderma, yet it is a rare case.

Keywords: Oesophageal stricture, Limited scleroderma, ANA titres

INTRODUCTION

Systemic scleroderma is a multisystem disease with microvascular abnormalities, autoimmune disorders, excessive collagen production and deposition, fibrosis of skin and internal organs, with progressive course and potentially fatal outcome.

LeRoy et al. proposed the simplest classification of systemic sclerosis: diffuse cutaneous SS (diffuse systemic scleroderma), and limited cutaneous SS (limited systemic scleroderma). This is a case report of a patient with presentation of Limited scleroderma.¹

CASE REPORT

A 29 year old female came with complaints of: difficulty in swallowing associated with vomiting since 1 month.

Patient was apparently normal 1 month back following which she developed, difficulty in swallowing– it was gradually worsening resulting in intake of a single chappati for more than 1\2 hour.²



Figure 1: Patient's brother.

History of difficulty in swallowing first to solids and then to liquids associated with 2 episodes of vomiting. No history of fever\cold\cough\dyspnoea\trauma. Not a known case of diabetes mellitus \systemic hypertension\pulmonary tuberculosis \bronchial asthma \epilepsy. Also adds with history of skin lesions since childhood and similar history found in her younger brother. (Figure 1).



Figure 2: Malformed dentition and sclerosed lips.



Figure 3: Resorbed digits and sclerosed foot.



Figure 4: Barium swallow PA.

On examination

Conscious, oriented, afebrile.

Evidence of skin features: focal patches of sclerosis over the chest abdomen and limbs, resorbed fingers with raynouds phenomenon. Loss of hair over the forehead (Figure 2 and 3). Pulse rate: 86 \min. Blood pressure: 110\80 mmHg.



Figure 5: Barium swallow- lateral view. Contrast with? oesophageal stricture.

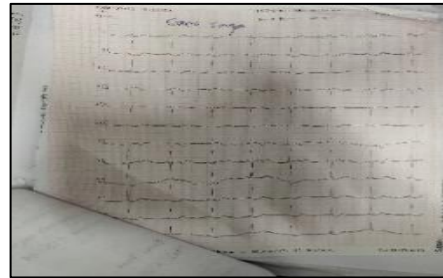


Figure 6: ECG of the patient showing normal sinus rhythm (to rule out pulmonary hypertension).

Systemic examination

Cardio vascular system: s1s2+ no murmurs, respiratory system: bilateral air entry + normal vesicular breath sounds, per abdomen: soft, central nervous system: no focal neurological deficit, ENT examination: oral cavity: malformed teeth +, trismus+, mild sclerosis over the lips, halitosis+, dental caries+ ,no e\o mass\growth seen, on video direct laryngoscopy: bilateral vocal cords mobile and normal, nose: depressed nasal bridge+, ear: right: tm intact and left: wax+, patient was worked up for oesophagoscopic dilatation.

ECG: normal sinus rhythm within normal limits (no evidence of pulmonary hypertension) (Figure 6)

Chest X-ray: NAD

Barium swallow: suggestive of oesophageal stricture. (Figure 4, 5).

All the blood reports were within normal range.

Anti-centromere antibody titres: (1:160)³

Intraoperative findings: A pin hole lumen of oesophagus from cricopharynx till gastroesophageic junction. With the help of bougie dilatation was done in 2 sittings with a gap of 1 month.⁴

1st sitting: bougie dilatation of size 16 & 18 was done.

2nd sitting: bougie dilatation of size 26&28 was done.

And patient was advised for repeated follow-ups and corticosteroids were given for scleroderma as advised by dermatologist.

DISCUSSION

Crest syndrome incidence: 1/12,500 adults. Males: females 1:4. Although patients with Scleroderma present with changes in the entire gastrointestinal tract, esophageal involvement is most common.⁵ The esophageal epithelium may show a cobblestone appearance due to pearly white plaques; apart from sub-epithelium, fibrosis may also affect regions with muscle atrophy.⁶ Apart from esophageal aperistalsis, spasms and esophageal stricture have rarely been reported. It has been pointed that symptoms of esophageal reflux are twice as common as dysphagia. Gastro intestinal disturbances such as heartburn, dysphagia or respiratory complaints example: dyspnoea are occasionally the first manifestations of the disease. Dysphagia, manifested by various abnormal swallowing sensation, is initially caused by impaired oesophageal motility but later can result from gastroesophageal reflux disease and secondary stricture formation. This case report includes a rare presentation of crest Syndrome in a male with typical features.

CONCLUSION

When classifying systemic scleroderma into two groups - diffuse and limited, with CREST syndrome as a variant of the latter, it should be considered that both types represent clinical variants of systemic sclerosis, share similar visceral involvement, laboratory abnormalities and the course which is not invariable, as shown in our patient. This case report of a patient with typical features of limited SSc, and all the symptoms of CREST syndrome,

therapeutic modalities provided only temporary improvement.⁶

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REFERENCES

1. LeRoy EC, Black C, Fleischmajer R, Jablonska S, Krieg T, Medsger TA, et al. Scleroderma (systemic sclerosis): classification, subsets and pathogenesis. J Rheumatol. 1988;15:202-5.
2. Nisa L. Otolaryngol Head Neck Surg. 2012;146(4):6767.
3. Respaldiza N, Ocana WC, Hernandez FJ, Castillo MJ, Magarino MI, Magarino R et al. Scand J Rheumatol. 2006;35(4):290-4.
4. Gut. 2018 Jun;67(6):1000-1023. doi: 10.1136/gutjnl-2017-315414. Epub 2018 Feb24.
5. Yoon JC, Elston DM. CREST syndrome treatment and management. Medscape Drug, Disease and Procedures.
6. Goodfield MJ, Jones SK, Veale DJ. Connective tissue diseases. In: Burns T, Breathnach S, Cox N, Griffiths C, editors. Rook's textbook of dermatology. 8 ed. Oxford: Blackwell Publishing; 2010;51.1-51:138.

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