Radiological manifestations of scleroderma- A case report

S.G.Gandage*, V.D.Aironi**, Gauri Sawadkar***

Abstract

Scleroderma is a chronic multi-system disorder of autoimmune aetiology, characterised clinically by thickening of skin and structural and functional abnormalities of visceral organs. Here we present a case of scleroderma with emphasis on its radiological manifestations

Key words *- Scleroderma, barium swallow, acro-osteolysis*

Introduction

Scleroderma is a clinically heterogeneous generalised disorder which affects the connective tissue of the skin and internal organs such as gastrointestinal tract, lungs, musculoskeletal system.[1] It is characterised by alterations in microvasculature and massive deposition of collagen. Gintrac in 1847 introduced the term "scleroderma" as skin is most obvious organ involved.[2]

Case Report

We present classical radiological features of a case of scleroderma in a thirty five years old female patient, a diagnosed case of scleroderma, who was referred to the department of Radiodiagnosis for barium swallow examination for investigation of dysphagia.

Barium swallow examination findings

The pilot film showed reticulo-nodular infiltration in both lower lung zones and cardiomegaly. There is moderate dilatation of oesophagus with poor progression of peristalsis. There is delayed transit of barium across gastro-oesophageal junction with mild degree narrowing at distal oesophageal end. There is persistent hold-up of contrast in distal two-third of oesophagus due to sluggish peristalsis. (Fig1) There is no mucosal irregularity or gastro-oesophageal reflux. The findings of dilated oesophagus with slow progression of barium across gastro-esophageal junction and slow peristalsis are consistent with the history of scleroderma.

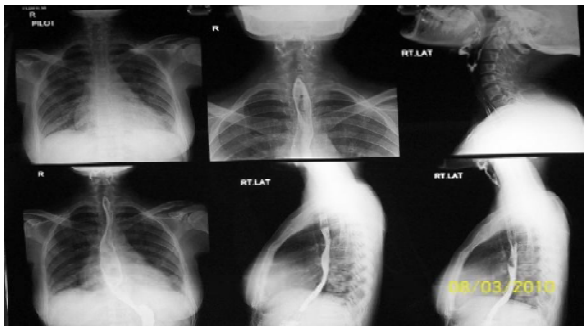

Fig 1: Barium swallow examination showing dilated aperistatic lower 2/3rd of oesophagus

The radiograph of both hands, antero-posterior view showed disintegration of tip of distal phalanx of index finger, suggestive of acro-osteolysis. (Fig 2).

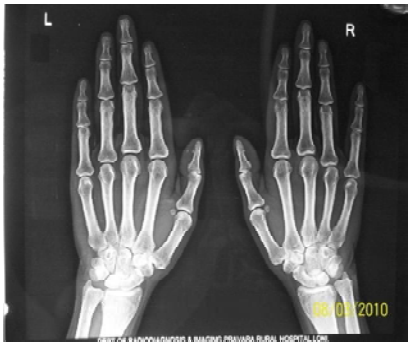

Figure 2: Radiograph of hands showing acro-osteolysis of distal phalanx of index fingers

*Professor and HOD, Radiology, Rural Medical College, Loni

**Associate Professor, Radiology, Rural Medical College, Loni

***Postgraduate Student, Radiology, Rural Medical College, Loni.

Corresponding Author:
Dr.Vyankatesh D. Aironi,
Associate Professor,Department Of Radiodiagnosis,
Rural Medical College,Loni-413736, Ahmednagar, Maharashtra, India
Email: v_aironi@yahoo.com

The photograph of the patient showed the nose with pinched or beaklike appearance. Wrinkles are seen around the mouth perpendicular to lips (Fig 3). Involvement of fingers leads to sclerodactyly, atrophy and resorption of soft tissue at finger tips as seen in the photograph of both hands (Fig 4).

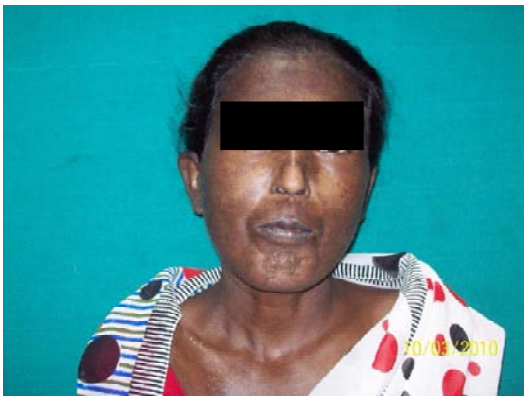

Fig 3: Photograph of the patient showing beak-like nose and expressionless face

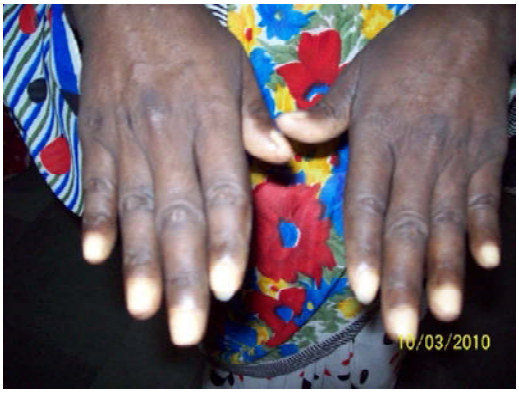

Fig 4: Photograph of hands of same patient showing sclerodactyly of index fingers

Discussion

Scleroderma is a multi-system connective tissue disorder of autoimmune aetiology characterised by widespread disorder of microvasculature and overproduction of collagen causing exuberant interstitial fibrosis with atrophy and sclerosis of many organ systems.[1] Peak age of occurrence is 30 to 50 years, with male to female ratio of 1:3.[3] It may be associated with other connective tissue diseases and many serological markers are used as adjuncts in its diagnosis.[1]

Raynaud's phenomenon, defined as episodic vasoconstriction of small arteries of fingers and toes is the first symptom in 95% of cases.[1] Involvement of skin leads to firm, thickened and eventually tightly bound skin to underlying subcutaneous tissue. Involvement of face results in thinning of lips, loss of skin wrinkles and facial expressions.[1] The nose takes on a pinched or beaklike appearance. Wrinkles appear around the mouth perpendicular to lips.

Majority of patients with scleroderma have GI tract involvement with oesophagus as first organ to be involved.[2] In oesophagus there is hypotonia or atony with hypokinesia or aperistalsis in lower two-third of oesophagus. Involvement of oesophagus may lead to complications like stricture formation, aspiration, Barrett's oesophagus and adenocarcinoma. Small bowel involvement leads to malabsorption due to delayed intestinal transit time and bacterial overgrowth. On barium meal follow through small bowel involvement in scleroderma has the appearance of "accordion pattern".[4]

Pulmonary scleroderma is seen as bibasilar pulmonary fibrosis with usual appearance on chest radiograph as basal reticulo-nodular shadowing with progressive pulmonary volume loss. More than 50% of patients with scleroderma have musculoskeletal features, majority of which are seen in hands. Involvement of fingers leads to sclerodactyly, atrophy and resorption of soft tissue at finger tips with soft tissue calcification.[5] On radiograph of hands, acro-osteolysis is noted which means resorption of distal phalanges of fingers.

Early diagnosis and individually tailored therapy by a team of specialists is needed to manage this disorder, which is treatable but not curable. Therapy involves immuno-modulation as well as the targeting of blood vessel mechanics and fibrosis. Physiotherapy and

psychotherapy are also important adjunctive therapies in this multifactorial disease.[2]

References

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