SCLEREDEMA
(Case reports with follow up)

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Summary

Four cases of scleredema are reported. Two cases showed high serum cholesterol level and were put on eltroxin and one patient showed some response. Two cases were below 20 years of age and other two above 20 years. The ratio of male to female was 3 : 1.

Scleredema is a rare entity of unknown cause characterised by diffuse induration of skin. The onset is sudden, usually preceded by some infectious episode commonly of streptococcal origin. The induration is wooden-like, non-pitting, usually starting from the sides of the neck or the face and progressing symmetrically to the upper limbs and trunk. The whole process reaches its peak of intensity usually in 1–2 weeks. The disease is commonly limited to the skin though in rare cases internal involvement like pleural and pericardial effusions, cardiovascular disease, diabetes mellitus and occular involvement have been reported[1,2]. Four cases followed up for a long period (1½ years to 3½ years) are being presented.

Case No. 1

A 10 years old female child visited Skin and V. D. O. P. D. of Medical College Hospital, Rohtak on 23-3-1974 with the complaints of stiffness of the skin over the face, neck, upper arms, chest and back of 6 months' duration. The process initiated from the face and within 3–4 weeks spread downwards involving neck, upper arms, chest and back.

There was history of fever, sore throat and living infection for about 2 weeks preceding the onset of skin involvement.

Examination of the skin showed woody hard skin over the face, neck, upper arms, chest and back. The induration was non-pitting and skin could not be pinched. Opening of the mouth was restricted and wrinkling on the forehead could not be demonstrated. The movements of the neck were limited.

Investigations

Routine investigations on blood, urine and stool were normal. S. T. S. was negative. Serum cholesterol, blood sugar and serum calcium were within normal limits. Vital capacity was normal. E. C. G. and X-ray chest did not show any abnormality. L. E. cell phenomenon was negative.

Histopathology

Biopsy of the skin supported the clinical diagnosis. Staining for mucopolysaccharide was negative.

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Follow-up :

Patient was put on systemic steroid, resulting in gradual regression of stiffness after 6-8 months of therapy and within 1½ years there was complete relief. She has been attending the follow-up clinic regularly for the last 3½ years. One residual feature that has been noticed in this case is that the patient develops slight stiffness of the cheeks on exposure to cold in the morning hours and in winter, which softens on exposure to sunshine or warmth.

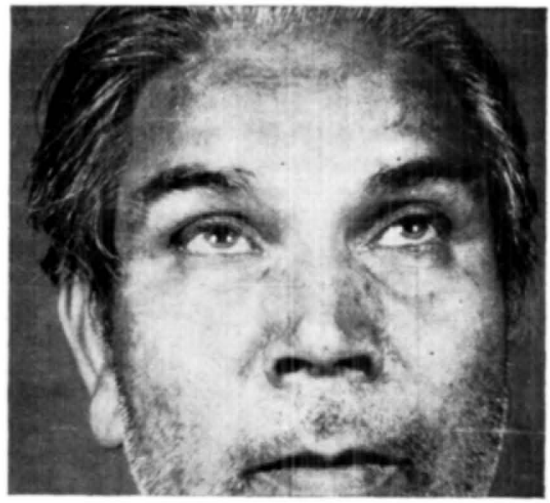

Fig. 1 Showing. lack of wrinkling over forehead.

Case No. 2

A 31 years old male patient attended the skin out patient department of Medical College and Hospital, Rohtak, on 28—5—1976 with complaints of stiffness of skin over the face, neck, upper extremity and trunk of one month's duration. The process initiated from the back and side of the neck and within 10-12 days progressed rapidly upwards and downwards involving face, trunk and upper extremity, limiting the movements of the neck and shoulders. There was no difficulty in swallowing.

There was history of fever with cough and expectoration before the onset of the present symptoms. These prodromal symptoms were relieved by some treatment nature of which was not known.

Examination of the skin revealed the skin over the face, posterior and lateral side of the neck, back, chest, abdomen and upper extremity excluding the hands and distal half of the forearms, hard and bound down while genitalia and lower extremity were normal. The induration was non-pitting and skin could not be pinched. Full opening of mouth was not possible and wrinkling on the forehead could not be demonstrated (Fig. 1). The movements of the neck and shoulder were restricted but there was no inflammation atrophy or loss of hair and sensations were normal.

Investigations

Routine blood and urine examination was normal. S. T. S. non reactive, serum calcium - normal but serum cholesterol was slightly high (325 mg%), L. E. cell phenomenon - negative. PBI-normal. Sputum for A. F. B. negative. Vital capacity slightly reduced (2100 cc). E. C. G. – N.A.D. X-ray chest– N. A. D. Blood sugar normal.

Histopathology

Biopsy of skin supported the clinical diagnosis. Section did not show positive staining for mucopolysaccharides.

Follow-up

Patient was put on combination of systemic steroids and thyroxine (eltroxin) for about o months. No improvement was seen. Thyroxine was withdrawn. Patient is still under observation and treatment and regularly attending the follow-up clinic. So far the only improvement noticed is slight increase in the movements of the shoulders.

Case No. 3

A 33 years old male patient attended the Skin O. P. D. of Medical College and Hospital, Rohtak on 3—7—76 with the complaints of hard skin around the neck, chest, and back of 1½ years' duration. There was history of recurrent episodes of sore throat and cold preceding the skin complaints. About 5-6 days after such an episode feeling of tightness around the neck and some difficulty in movements of neck was observed but there was no difficulty in deglutition. The tightness then spread upwards and downwards involving the face, back, upper arms, chest and abdomen and reached its peak within a few weeks. There was occasionally difficulty in breathing at the onset but there was no restriction of movement of shoulder or elbow joints.

Patient was earlier admitted into A. I. I. M. S., Delhi with a diagnosis of scleredema and was put on steroid therapy for over a year. With 8 months of treatment, there was gradual regression of the disease and when it became stationary for 4–5 months, he was admitted into Medical College and Hospital, Rohtak. Examination of the skin showed hard and bound down skin over the neck and some stiffness over the forehead and around shoulders, and neck movements were restricted but the skin had become normal over the face, lower back, abdomen, and upper extremity. He had no difficulty in breathing.

Investigations

All the investigations as mentioned in case 2 were done in this patient also, and revealed normal values except mild elevation of serum cholesterol (350 mg%).

Histopathology

Skin biopsy supported the clinical diagnosis of scleredema.

Follow-up

With systemic steroids, the patient showed initial regression of the disease and then remained stationary for about 6 months before his visit to this hospital. Combination of steroid and eltroxin was administered for about 4 months with some improvement in stiffness on the face and neck movements. Patient was discharged with advise of regular follow-up but subsequently defaulted.

Case No. 4

A 17 years old male first visited on 22—10—1977 with the complaints of fever, sore throat and stiffness of skin around the neck. It started with fever and sore throat 2 months earlier. Simultaneously stiffness of neck was noticed and progressed on to face within a few days. And later to upper extremities and trunk.

Examination revealed wood like stiffness of skin which was bound to the underlying structures. The induration was non-pitting and skin could not be pinched. Full opening of the mouth was not possible and wrinkling on the forehead could not be demonstrated. Movements of the neck were also restricted.

Investigations

All the investigations as mentioned in earlier cases were done in this patient also, and revealed no abnormalities.

Discussion

Although this condition was originally described by Piffard in 1876 it was recognised as a distinct entity by Buschke[8]. The term scleredema adultorum is a misnomer as it has been reported to occur in quite a number of cases before the age of 20 years; about 29% before the age of 10[4]. It is more prevalent in females. In our series of 4 cases, one was a normal female child

of 6 years and three were males aged 35 years, 40 years and 17 years. All four cases gave history suggestive of upper respiratory infection preceding the disease.

The disease is predominantly cutaneous although associated internal involvement like pericardial and pleural effusion, cardiovascular disease and maturity onset diabetes mellitus, have been reported by some workers[1,2,5]. None of our cases showed any evidence of associated conditions, except that two males were slightly obese with marginal increase in serum cholesterol.

Prognosis is usually good as far as life expectancy is concerned. Condition usually improves within 2 years though in some the disease is known to persist for many years[1]. There is no effective treatment for this disease. Various treatments including corticosteroids have been tried with different claims. The female child who was only on corticosteriods was cured in $1\frac{1}{2}$ years. The two males, being slightly obese with raised serum cholesterol were given thyroid hormones in addition to corticosteroids. Case No. 2 still under observation and treatment for nearly 2 years has shown no improvement while case No. 3 showed initial improvement with corticosteroids and further improvement on addition of thyroid hormones to the corticosteroids. The latter case did not turn up for further check up while case No. 1 and 2 are attending the follow up clinic regularly.

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