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Post-streptococcal scleredema an unusual rare mimicker of scleroderma: a case report

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SUMMARY

Scleredema of Buschke is a rare pathological disorder of connective tissue, which is characterized by a woody, diffuse induration of the skin, most often in the upper extremities. We report an extremely rare complication of post-streptococcal infection in a six-year-old male complaining of gradually progressing, painless skin thickening and tightness which was preceded by a one-month history of fever, cough, and tonsillitis. By reporting this case, we hope to contribute to the creation of a database for future research aimed at better understanding the incidence, pathophysiology, and management of this extremely rare complication.

Key words: *Scleredema, post-streptococcal, complication, skin thickening, infection.*

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INTRODUCTION

Scleredema of Buschke is a rare pathological disorder of connective tissue associated with diabetes mellitus, infection, particularly streptococcal infection of the upper respiratory tract, or monoclonal gammopathy. Scleredema is characterized by a woody, diffuse induration of the skin, most often in the upper extremities (1). Scleredema's histological feature is dermal fibrosis, which is characterized by thicker than usual collagen bundles and varying amounts of mucin deposits (2). Scleredema is equally prevalent in both sexes and across racial and ethnic lines. Although it can affect people of any age, the prevalence of different types of this condition tends to cluster in different demographic groups (3, 4). Approximately 29% of all scleredema cases appear in the first decade of life (5). We report this extremely rare complication of post-streptococcal infection, scleredema of Buschke. To the best of our knowledge, this is the first reported case in Iraq.

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CASE REPORT

A six-year-old male presented to our rheumatology outpatient clinic complaining of gradually progressing, painless skin thickening and tightness. This condition initially manifested on his neck and face, but within a month it had progressed symmetrically to involve his trunk, upper limbs, and lower limbs as well. He did not report any limitation in the range of motion in the trunk, limbs, shoulders, or neck. The condition was preceded by a one-month history of fever, cough, and tonsillitis treated conservatively at home with over-the-counter medications. There was no report of accompanying skin rash, dysphagia, or other systemic symptoms. His past medical history was negative. The physical examination revealed that his vital signs were all within normal ranges, including his blood pressure. The child's anthropometric measurements were consistent with his age. On examination, the skin of his face, upper torso, and upper and lower limbs, except for his

fingers, was notably stiff, with woody non-pitting induration, a loss of normal elasticity, and the absence of Raynaud's phenomenon (Figure 1). The examination of other systems was unremarkable. All laboratory and radiological tests, including complete blood count, erythrocyte sedimentation rate, C-reactive protein test, renal and liver function tests, electrolytes, urinalysis, chest X-ray, electrocardiogram, echocardiogram, and abdominal ultrasound, were all normal. Antistreptolysin O (ASO) titre was markedly elevated at 1600 IU/mL. The antinuclear antibody and extractable nuclear antigen panels were negative. A skin biopsy was performed revealing an unremarkable epidermis apart from basal melanosis with mild superficial and deep perivascular lymphocytic infiltrate and prominent dermal sclerosis (Figure 2). The history, clinical examination, and increased ASO titre were all suggestive of post-streptococcal scleredema, which was confirmed by the skin biopsy. Amoxicillin-potassium clavulanate was given at a dose of 45 mg/kg/day, every 12 hours for 10 days, to eliminate any residual streptococcal infection. Given the disease's benign nature, the family was reassured and regular monthly visits for follow-up were set up. Since his initial diagnosis four months ago, the patient has had monthly follow-up visits, but there has been no improvement yet. The plan is to administer him immunoglobulin intravenously if he doesn't regain his normal skin elasticity after one year of follow-up.

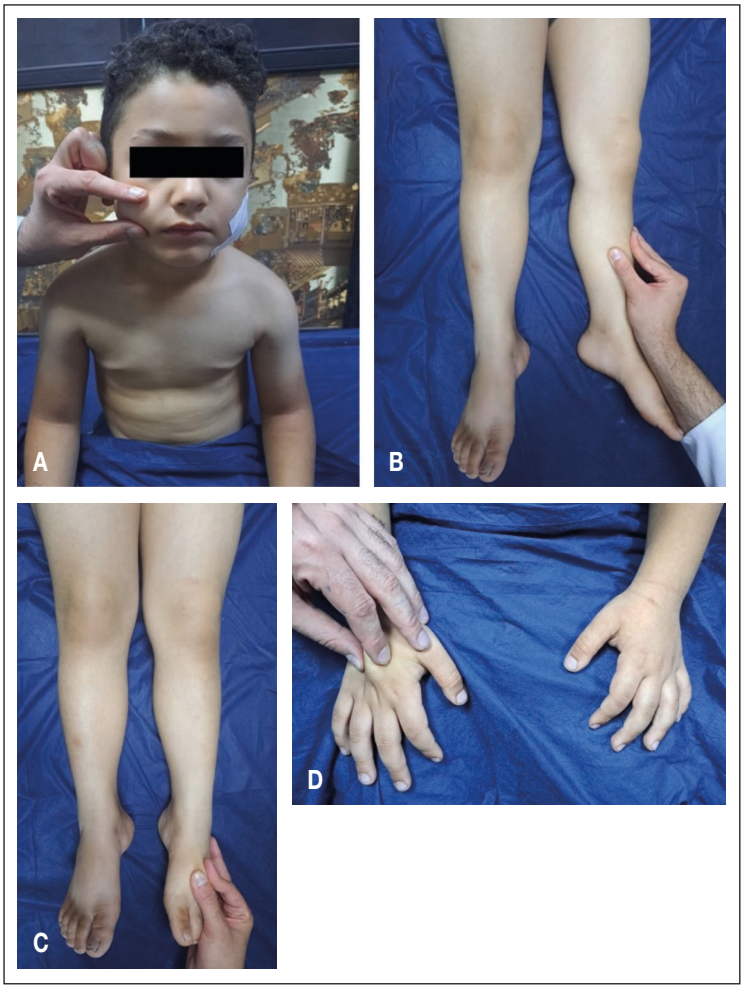

Figure 1 - Stiffness and a loss of normal elasticity of the skin on the face (A) and the shin (B) with relative sparing of the foot and the hand (C, D).

DISCUSSION

Scleredema of Buschke is a rare skin condition that is characterized by thickening and hardening of the skin (6). It is typically caused by a streptococcal infection, such as strep throat or scarlet fever, and can occur in individuals of any age (7).

The condition typically affects the neck, upper chest, and back, although it can also occur in the arms and legs. The skin appears swollen and firm and may have a woody or waxy appearance (8). In severe cases, the skin may become so thick and hard that it is difficult to move or bend the affected areas. Scleredema of Buschke is not contagious and does not usually cause any pain or discomfort (9). However, it can be cosmetically disfiguring and may cause emotional distress for those affected (10). The exact cause of the scleredema of Buschke is not fully understood, but it is believed to be an autoimmune response to the streptococcal infection.

There is no specific therapy for it and the treatment plan should focus on alleviating the symptoms and treating the underlying cause, depending on the specific type of scleredema. Type 1 is usually preceded by an upper respiratory infection. Antibiotics should only be administered to eradicate the underlying infection, as this type usu-

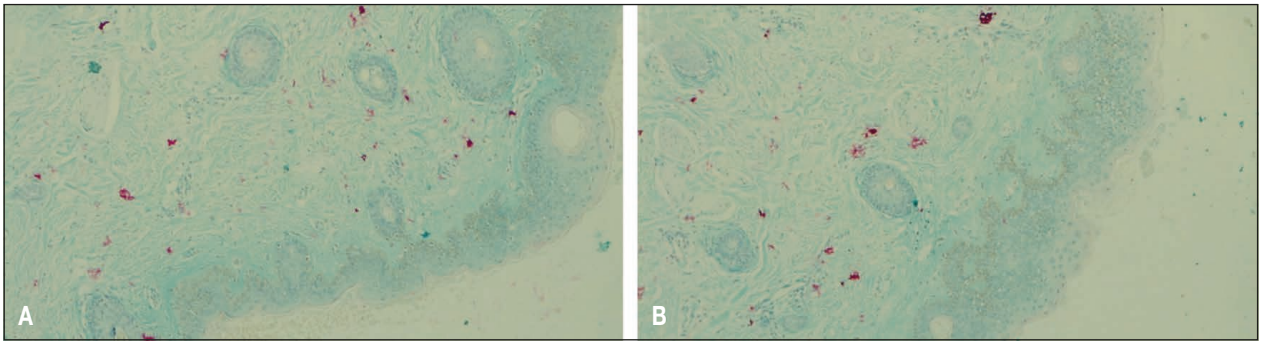

Figure 2 - Biopsy of the skin revealed unremarkable epidermis apart from basal melanosis with (A) mild superficial and deep perivascular lymphocytic infiltrate and (B) prominent dermal sclerosis.

ally resolves spontaneously. Type 2 usually occurs in patients without a preceding infection, and it is associated with paraproteinemias. Therefore, it requires management by a hematologist. Type 3 is associated with poorly controlled diabetes mellitus, so management should focus on the tight control of blood glucose (11-13).

The prognosis for the scleredema of Buschke is generally good, with most individuals seeing improvement in the appearance of their skin within several weeks to months of treatment (14).

While scleredema of Buschke is a rare condition, it is important for affected individuals to be aware of the symptoms and seek medical attention if they experience thickening or hardening of the skin following a streptococcal infection. Early treatment can greatly improve the appearance of the skin and prevent any potential complications (15).

It is also important to practice good hygiene and regularly wash hands to prevent the spread of streptococcal infections. This is important because, even if it occurs rarely, a streptococcal infection may lead to severe complications, especially in developing countries, and appropriate treatment is mandatory.

CONCLUSIONS

This case report aims to demonstrate and document a very rare post-streptococcal complication, which is rarely discussed or reported in the medical literature. By reporting this case, we hope to contribute to the creation of a database for future research aimed at better understanding the incidence, pathophysiology, and management of this extremely rare complication.

Conflict of interest

The authors declare no potential conflict of interest.

Funding

None.

Informed consent

Patient's legal guardian's consent was given to share this case for scientific purposes.

Availability of data and materials

Data and materials are available by the authors.

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