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Case Report

Scleredema of Buschke in a Child: An Uncommon Poststreptococcal Complication

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Abstract

Scleroderma (Buschke's) is a rare disease of the scleromucous connective tissue of unknown etiology that is included in the groups of secondary cutaneous mucinosis. There are few reports of this condition in the pediatric literature. We report a case of scleroderma following streptococcal infection in a 9-year-old girl.

Keywords: Scleredema of Buschke; Child; Poststreptococcal; Mucinosis

Introduction

Scleredema (of Buschke) is a rare scleromucinous connective tissue disease of unknown aetiology which is characterized by non-suppurative, oedematous thickening of the skin, especially the face, neck, shoulders, thorax, proximal extremities but not the fingers [1].

Post-infectious sclerosis is a rare manifestation that may follow a common infection and should be considered in children presenting a typical swelling or induration of the face and/or neck. There are few reports of this condition in the pediatric literature. We report a case of scleroderma following streptococcal infection in a 9-year-old girl.

Observation

A nine-year-old girl, the second child of a non-consanguineous marriage, born at term, with no significant perinatal history or antecedents, presented with induration that started gradually and affected the entire body, including the face, trunk, and extremities. There was no associated rash. She had no other associated systemic symptoms. There was a history of an upper respiratory system infection 15 days before the onset of firmness of the skin, and the symptoms and signs of this infection were still present.

The physical examination showed symmetrical diffuse swelling and induration and loss of normal elasticity of the upper and lower extremities, face, neck and the trunck , sparing the hands and feet **(Figure1)**. There was a limitation of mouth opening and temporomandibular joint mobility **(Figure 2)**. Systemic examination was essentially normal. The patient did not show any abnormalities in the blood workup, which included thyroid stimulating hormone, fasting plasma glucose and antinuclear antibodies. A serum protein electrophoresis revealed normal total protein and a normal electrophoretic profile. The Antistreptolysin O (ASO) titer was elevated to 580 U/L (normal:<325 U/L). A throat culture grew β-hemolytic group A Streptococcus. Histology showed slight widening of collagen bundles with normal fibroblast number. The presence of mucin deposits between the dermal collagen fibers is highlighted by alcian blue. The diagnosis of the scleredema of Buschke following a streptococcal infection was confirmed and a spontaneous improvement was noted 4 months later. Considering the benign nature of the disease, the parents were counselled about the good prognosis of the disease. Over the span of six months, the skin has been gaining its normal elasticity and the child has improved symptomatically.

Figure 1

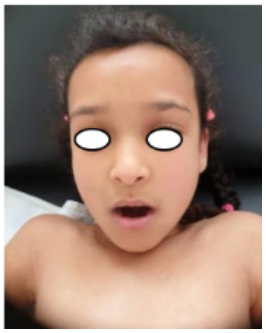

Figure 2

Discussion

Scleroderma (Buschke's) is a rare disease of the scleromucous connective tissue of unknown etiology that is included in the groups of secondary cutaneous mucinoses. Contradictory to Buschke's original description as 'scleredema adultorum', scleredema occurs in individuals of all ages, and more than 50% of patients are aged under 20 years. Its exact prevalence and incidence are unknown. There is no racial or ethnic predilection to the disease [2].

Three types of scleredema can been distinguished according to their association with preceding or underlying conditions: type

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1 usually occurs after a febrile illness, usually a streptococcal throat infection or a viral infection, and is the most common, with up to 55% of cases being postinfectious. Type 2 has no preceding febrile illness and is often associated with monoclonal gammopathies. This makes up approximately 25% of cases. The third subtype, accounting for 20% of cases, is termed as scleredema diabeticorum and is associated with poorly controlled diabetes. There are rare case reports of scleredema associated with solid organ malignancies [3].

The pathogenesis of scleredema remains unknown. The excessive production by fibroblasts of mucin (heavily glycosylated high-molecular weight proteins) and collagens in the reticular dermis may be provoked by diverse stimuli, including infections and inflammatory processes, drugs, toxins, immunoglobulins and cytokines, genetic factors, and hyperinsulinism, or chronic hyperglycaemia in case of type 3 scleredema [2].

The sclerosis is characterized by induration of the skin, especially on the face, neck, shoulder belts, and upper extremities, but spares the fingers. The abdomen and lower limbs are often affected, but to a lesser degree than the upper half of the body. The feet are never affected, but the dorsal surface of the hands is rarely affected (which differentiates scleroderma from sclerotherapy). Raynaud's phenomenon is absent, which is typically present in scleroderma.

The disease is usually self-limiting, with an active phase of 6 to 8 weeks and complete resolution within 6 months to 2 years. In rare cases, organs such as the eyes, tongue, esophagus are affected and the disease may result in pleural, pericardial or peritoneal effusions [1].

Histologically, scleredema is characterized by a normal epidermis and a thickened dermis filled with swollen collagen fibers and mucin that contains clear spaces.

Our patient was febrile and symptomatic with an elevated ASO titer and grew β-hemolytic group A Streptococcus from throat culture.

The differential diagnosis of scleredema in children includes scleroderma, eosinophilic fasciitis, and systemic amyloidosis and the other collagen vascular diseases [4].

Until the pathogenesis of scleredema is elucidated, treatment will remain solely symptomatic. This will include appropriate antibiotic for cutaneous infections to decrease the risk of poststreptococcal complications, physical therapy for those at risk for contractures, and maintenance of nutrition.

However, the number of patients reported in the literature that have received specific treatment is very small. Thus, it is very difficult to provide evidence based medical recommendations and conclusions on the optimal treatment regimens, optimal dose and long-term efficacy.

Conclusion

As a result of the growing evidence of the association between sclerosis and streptococcal infection, scleredema should be included in the medical literature along with rheumatic fever, glomerulonephritis, and erythema nodosum as a rare but significant complication of streptococcal infection.

Conflict of Interest: No

References

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