Unusual presentation of more common disease/injury

Face and neck swelling in a 16-year-old boy

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Summary

Scleredema adutorum is a rare condition characterised by progressive collagen and mucin deposition in the skin. While the aetiology has not been clearly delineated, the condition is often associated with common infections. The current report describes a previously healthy 16-year-old boy who presented with 3 weeks of progressive neck swelling and skin induration. He had evidence of both active streptococcal and Epstein-Barr virus (EBV) infections. Skin biopsy confirmed the diagnosis of scleredema. The patient was treated for his streptococcal infection, but otherwise managed conservatively. Clear improvement in the signs and symptoms was seen at a 3-month follow-up appointment. Scleredema can be a complication of streptococcal infection but to our knowledge has not been reported in association with EBV. It should be considered in the differential diagnosis of any patient presenting with cutaneous/subcutaneous induration and swelling of the face and/or neck.

BACKGROUND

Streptococcal and Epstein-Barr virus (EBV) infections are commonly seen in children. Scleredema is a rare manifestation that can follow a common infection and should be considered in patients with characteristic swelling of the face and/or neck. There are few reports of this condition in the paediatric literature.

CASE PRESENTATION

A 16-year-old boy presented to a paediatric hospital with a 3-week history of painless neck swelling which had progressed to involve his face, upper torso and proximal upper extremities. He complained of tightness of his face and neck causing reduced mobility. Despite this, he was in no distress, afebrile and otherwise well. There was no history of preceding trauma. A review of systems was unremarkable. He did not complain of any fatigue, weight loss or preceding upper respiratory tract infectious symptoms.

His medical history included a left hemi-hypertrophy since birth for which he was followed up yearly by an orthopaedic surgeon. He also had a history of non-alcoholic fatty liver disease (NAFLD) which was controlled with diet and weight loss. His only medication was milk thistle, which he took for his NAFLD. He had no known allergies.

Physical examination revealed a young man in no distress. His vital signs were stable and he was afebrile. He was slightly over weight with body mass index of 34.6. He was comfortable when speaking; however, mouth opening was limited as was facial expression. Head and neck exam revealed waxy, indurated, woody appearing skin covering the majority of his face from chin to forehead (figure 1). The skin changes extended inferiorly down his neck, to the lateral part of his upper arms and circumferentially affecting the upper torso above the nipple line. The skin was not erythematous, warm or tender. There was evidence of his longstanding mild left

side hemi-hypertrophy. He had no hepatosplenomegaly and his physical exam was otherwise unremarkable.

INVESTIGATIONS

Laboratory investigations revealed a normal complete blood count, with occasional atypical lymphocytes seen on smear. Urea and creatinine were normal as was a urinalysis. Transaminases were elevated with aspartate aminotransferase 44 units/l and alanine aminotransferase 96 units/l. This was similar to previous testing related to his known NAFLD. His antistreptolysin O (ASO) titre was elevated at 456 units/ml and his throat swab was positive for group A *Streptococcus*. His monospot was also positive. EBV antiviral-capsid antigen (anti-VCA) immunoglobulin G (IgG) and antinuclear-antigen IgG were both non-reactive. His anti-VCA IgM was reactive, confirming an acute EBV infection. His C reactive protein was slightly elevated at 11 mg/l. His erythrocyte sedimentation rate, C3 and C4 were within normal limits. An abdominal ultrasound revealed fatty infiltration of his

Figure 1 Patient at presentation with indurated skin and neck swelling.

Figure 2 Scleredema. Skin punch biopsy from neck shows intact epidermis and expanded dermis due to oedema (H&E stain, low power view).

liver and mild splenomegaly. The liver findings were consistent with a previous abdominal ultrasound in 2007. Results from a skin biopsy from the right posterior neck performed by dermatology confirmed the diagnosis of scleredema (figures 2–5).

DIFFERENTIAL DIAGNOSIS

A 16-year-old boy with progressive neck swelling and induration needs to have a thorough work-up to rule out lymphadenopathy, superior vena cava syndrome and thyroid mass. This can usually be done with history, physical exam and imaging. Other rare conditions

Figure 3 Skin oedema seen as fenestrations of collagen bundles in the dermis (H&E stain).

Figure 4 Skin oedema seen as fenestrations of collagen bundles in the dermis (H&E stain).

including systemic and localised scleroderma and scleromyxedema may have similar skin changes to scleredema, and should also be considered in the differential diagnosis. Scleroderma is a systemic autoimmune disorder characterised by skin and organ fibrosis. It can be associated with Raynaud's phenomenon and oesophageal dysmotility.¹ The localised scleroderma (morphea) is a fibrotic skin condition that has several subtypes, each with a unique appearance and distribution.¹ Scleromyxedema is a disease characterised by dermal deposition of mucin and fibroblast proliferation. Patients present most commonly in middle age with pale waxy papules on the hands, forearms, face, neck and upper trunk. This condition is associated with monoclonal gammopathies.² History and physical exam findings can be useful to differentiate between these disorders, but often the final diagnosis requires a skin biopsy. In all cases it is important to delineate the exact cause as the treatment and course for each is substantially different.

TREATMENT

The patient remained in hospital while awaiting biopsy confirmation of the diagnosis, during which time his streptococcal infection was treated with penicillin. As there was no further progression in the scleredema he was discharged home with a plan for outpatient follow-up.

Figure 5 Colloidal iron stain highlights the dermal accumulation of mucin filling the fenestration of collagen bundles in the dermis.

Figure 6 Patient in follow-up 3 months later showing near resolution of neck swelling and indurated skin changes.

OUTCOME AND FOLLOW-UP

He was seen in follow-up approximately 3 months later. The scleredema of his arms, chest and mid-scapular region had completely resolved. A significant reduction in neck and facial swelling was noted (figure 6). Repeat testing showed normalisation of his ASO titre.

DISCUSSION

Scleredema is a disorder of collagen and mucin deposition characterised by non-pitting oedema and sclerosis of the skin. The terms scleredema of Buschke and scleredema adultorum are used interchangeably to represent scleredema of any cause.³ Three subtypes have been recognised: type 1 usually occurs after a febrile illness, 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 diabetorum and is associated with poorly controlled diabetes.³ There are rare case reports of scleredema associated with solid organ malignancies.⁴ Types 1 and 2 seem to affect women approximately twice as often as men, while type 3 occurs more frequently in men.³ Infectious agents associated with scleredema include most commonly *Streptococcus*, but also influenza, varicella, measles and mumps.⁵

The woody induration of skin seen in scleredema typically involves the neck, back, interscapular region, face and chest. Typically the extremities, especially distally, are spared. There can be significant involvement of the face, in some cases causing ocular muscle palsy, diminished mouth opening or periorbital oedema. Systemic involvement is rare, but can involve the tongue, pharynx or upper oesophagus.⁵ There have been cases of cardiac dysfunction.⁶ Scleredema has been described in paediatrics, although the true frequency is difficult to elicit. A 1963 review by Greenberg *et al* reported 209 cases of scleredema of Buschke. In total, 29% of the cases occurred in children less than 10, and 22% in children aged 10–20 years.⁷

Histological features include a normal epidermis with minimal perivascular dermal infiltrate. The reticular dermis is significantly thickened, and swollen collagen fibres are seen with H&E staining. The secretory coils of the sweat glands lie in the middermis.⁸ There is often mucin deposition between the thickened collagen bundles.

The different subtypes of scleredema all have the same underlying histopathology.

The natural history depends on the underlying cause. Patients with scleredema diabetorum tend to have an insidious onset over many years. If glycemic control is improved, the scleredema may experience some improvement. In contrast, infection-related scleredema tends to have a rapid onset of symptoms over days to weeks after the infection. The scleredema in these cases tends to be self-limited with resolution up to 24 months.⁵ As the disease tends to be self-limited, there is little evidence for treatment. A course of penicillin should be prescribed if there is evidence of a recent streptococcal infection. Various immunosuppressants (methotrexate, corticosteroids) have been tried without clear benefit.⁵ Ultraviolet light has shown some benefit in diabetic scleredema.⁹

Learning points

- Our patient presented with evidence of concurrent group A streptococcal and Epstein-Barr virus (EBV) infections; either of these agents may have triggered the scleredema.
- To our knowledge there are no case reports of scleredema in association with an EBV infection.
- Scleredema is a rare complication that can follow common paediatric infections such as *Streptococcus*.
- Scleredema should be considered in the differential for any patient with (sub)acute skin induration of the face and/or neck, particularly in the context of a preceding upper respiratory tract viral or bacterial infection.
- While the skin changes of scleredema can be dramatic and worrisome, the patient can be reassured that the course is typically self-limiting, with a gradual but complete resolution.

Competing interests None.

Patient consent Obtained.

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