Dermacase

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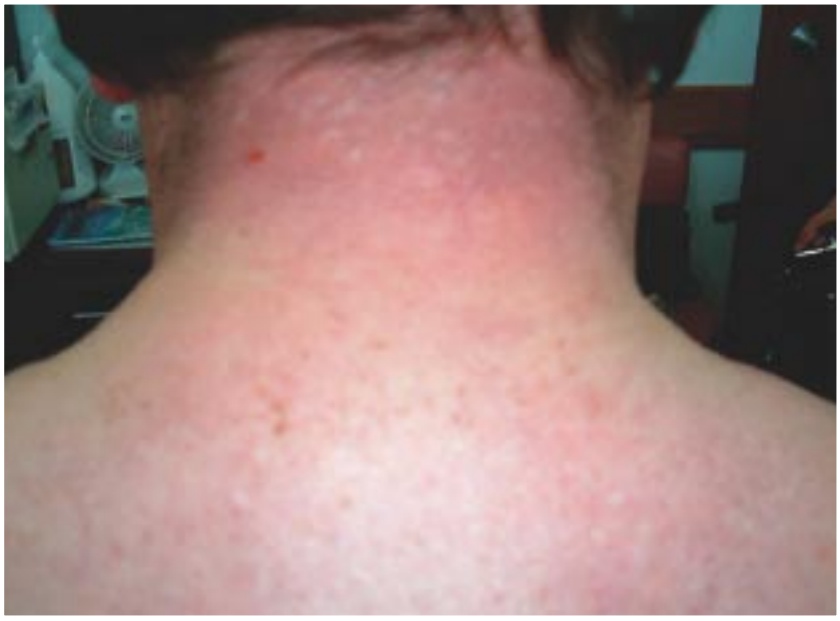

CAN YOU IDENTIFY THIS CONDITION?

A 46-year-old man with a history of type 1 diabetes presents with chronic poorly defined, woody, nonpitting, erythematous plaquelike lesions on his neck and the upper part of his back. Previous treatment with topical steroids did not improve the condition. Current medications include short- and long-acting insulin and angiotensin-converting enzyme inhibitors for blood pressure control.

The most likely diagnosis is:

1. Scleroderma
2. Scleredema adultorum, or Bushke disease
3. Amyloidosis
4. Myxedema
5. Cellulitis

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Answer to Dermacase

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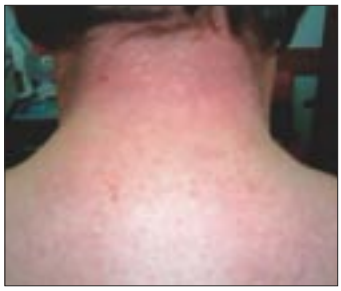

2. Scleredema adultorum, or Bushke disease

Scleredema adultorum, or Buschke disease, is a rare dermatologic condition associated with type 2 diabetes, previous febrile illness, or blood dyscrasias.[1] It is characterized by woody, nonpitting, erythematous or hyperpigmented, poorly defined infiltration of the skin. It is found most often on the face, neck, and upper back.

The cause of this condition is not understood. It is thought to arise from allergic sensitization or as an autoimmune phenomenon.[2] Procollagen and collagen synthesis is increased in fibroblast culture from affected skin.[3] Histologic examination shows a thickened dermis with mucin deposition between thickened collagen bundles.[4]

Three distinct forms of scleredema adultorum have been defined in the literature[1,5,6] (**Table 1**). In subtype 1, scleredema is preceded by a viral or bacterial infection (usually streptococcal) and most commonly occurs in children. Subtype 2 is usually associated with blood dyscrasias (eg, paraproteinemias). Patients with subtype 3 usually suffer from type 2 diabetes. Some studies suggest that scleredema is considerably more common than once thought and that diabetes is probably the most common cause.[7,8]

Table 1. Scleredema subtypes

ASPECT	SUBTYPE 1	SUBTYPE 2	SUBTYPE 3
Age of onset	Any age, more common in children	>15 y	>40 y
Associated diseases	Febrile illness: streptococcal pharyngitis, cytomegalovirus, influenza, measles, mumps, diphtheria, encephalitis, dental abscesses	Blood dyscrasias, araproteinemias, multiple myeloma	Type 2 diabetes
Onset	Days to 3 mo after infection	Insidious	Insidious
Sex affected	Women more than men (~2:1)	Women more than men	Men more than women
Duration	<2 y	Slow, progressive	Unremitting
Visceral involvement	Carditis, myositis, skeletal and ocular muscle, pharynx, liver, parotid glands, pleurae, peritoneum, spleen, upper esophagus	Occasionally	Rare, primarily diabetes-associated complications

History and physical examination suggest the diagnosis. Laboratory investigations include:

- throat culture for group A streptococcal (GAS) infection and antistreptolysin O titres to exclude recent GAS infection if you suspect type 1 scleredema,
- fasting blood glucose and glycosylated hemoglobin A_1C if you suspect type 3 scleredema,
- serum protein electrophoresis and immunoglobulin studies to exclude monoclonal gammopathy, paraproteinemias, and multiple myeloma if you suspect type 2 scleredema, and
- punch or incisional skin biopsy with inclusion of subcutaneous fat to confirm clinical diagnosis.

Differential diagnosis includes scleroderma, myxedema, amyloidosis, lymphedema, cellulitis, and dermatomyositis. Prognosis largely depends on the underlying etiology.

Scleredema in children and postinfection-onset scleredema in adults usually resolves within 2 years. Scleredema in patients with blood dyscrasias can resolve if treatment of the primary disease is successful. Scleredema in diabetic patients is usually progressive and unremitting.

Complications include limited range of motion, restrictive lung disease, dysarthria, dysphagia, skin infections, and poor wound healing.

Management of scleredema adultorum is often difficult. No dietary or activity restrictions are necessary. Patients with range-of-motion difficulties should be referred for physiotherapy. Antibiotics are recommended if GAS infection is confirmed, but antibiotics do not appear to shorten or cure skin conditions in scleredema. Tight glycemic control is recommended for diabetic patients, but has no effect on lesions once they exist. Intralesional steroids might be of some benefit. Adjuvant psoralen ultraviolet A–range therapy might be advised, and blood dyscrasias should be investigated and appropriately treated.

References

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