Case 14

Thickened skin on the posterior neck

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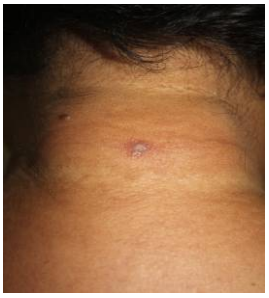
Patient: A 50-year-old Thai woman from Samutprakarn

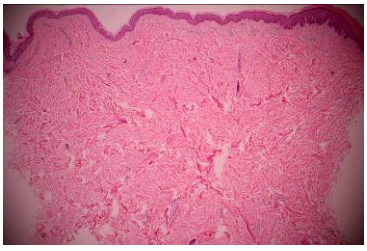
Chief complaint: Large skin induration on the posterior neck for 20 years

Present illness: The patient presented with asymptomatic, tightness, thickening and hardening of skin over the posterior neck for 20 years and gradually progressed larger. She was treated by topical medications without any improvement.

Past history:
- Underlying diseases: Diabetes mellitus, dyslipidemia and schizophrenia
- On metformin 2,000 mg/day, simvastatin 40 mg/day

Skin examination:
- A waxy, ill-defined, non-pitting induration of posterior neck skin with peau d' orange appearance
- Other physical examination was unremarkable





Histopathology: (S13-025390, posterior neck)

There is proliferation of thick and coarse collagen bundles, arranging in haphazard array with a hint of mucinous deposition in the lower reticular dermis in association with epidermal cyst in the overlying dermis.

Diagnosis: Scleredema Adultorum (of Buschke)

Investigation:
- Fasting blood glucose 108 mg/dL
- HbA1C 6.89%
- Cholesterol 254 mmol/L, triglyceride 104 mmol/L

Treatment:
- Advice diet control
- 10% Urea cream apply twice daily

Discussion:

Scleredema Adultorum of Buschke (SAB) is an infrequent disorder characterized by non-pitting induration of skin due to excessive mucin and collagen deposition in the reticular dermis. The etiology of SAB is unknown. The skin changes of SAB were described by Dr. Abraham Buschke who characterized the development of a hardness of the skin in 44-year-old man following Influenza infection in 1902. The relation of Diabetes Mellitus was established in 1970 and the term "Scleredema Diabeticorum" is often used in this setting.[1]

Characteristically, the affected skin appears smooth and waxy, with tense dermal induration and prominent follicular ostia called" Peau d' Orange" appearance. In general, however, skin changes are better felt on palpation than seen. The involved skin gradually transition into normal skin with no clear demarcation. The areas occurred at the neck, face, spreading symmetrically to involve arms, shoulder, back and chest. They are usually confined to the upper part of the body. Hands and feet are typically spared. However, a case of SAB localized on the thigh has been reported.[2]

Systemic manifestation included dysphagia and dysarthria due to tongue involvement; ocular involvement including exophthalmos, chemosis, induration of conjunctiva and eyelid, ophthalmoplegia were reported.[3] Also bone marrow, nerve, liver and salivary gland might be involved. Another serious manifestation is cardiac involvement eg. Friction rubs, gallop, arrhythmia, ST segment elevation, inverted T wave, right-sided heart failure have been reported.[4] Pulmonary fibrosis and pleural effusion have also been shown to correlate with cutaneous disease.

Scleredema Adultorum of Buschke has 3 clinical variants.[5]

Type I SAB is the classic type. The development of fine induration of the skin over posterior neck spreads upward to the head and face. And downward on the trunk and proximal upper extremities after a febrile prodrome of acute streptococcal respiratory tract infection.[6] The tongue and pharynx maybe involved leading to difficulty opening the mouth and swallowing. Most common in children and adolescence completely resolve without intervention in 6 months to 2 years.

Type II SAB shares similar clinical feature to Type I. However, Type II SAB has an insidious onset without preceding febrile illness. It usually appears several years after the onset of scleredema and is frequently associated with monoclonal gammopathy.[7]

Type III SAB occurred particularly in middle-age obese individual more often in men, with insulin dependent Diabetes Mellitus and is refer to Scleredema Diabeticorum. The onset is subtle with a slowly progressive, the upper back typically demonstrates erythema and induration, usually scleredema refractory to treatment. Complications of diabetes tended to be the case of mortality before skin involvement becomes widespread.

There is no racial or ethnic predilection. Women are affected twice as frequently as men in Type I and II while men are predominant in Type III. Rare associations include pituitary-adenocortical neoplasm[8], carcinoid tumor[9], gall bladder carcinoma[10], malignant insulinoma, hyperparathyroid, connective tissue diseases and HIV infection.[11]

The term "Scleredema Fulminans"[12] is utilized in severe progressive cases of SAB with rapid onset and life- threatening such as with HIV infection, DM poor controlled, severe hypertension. Accompanying the skin changes, worsen cough, dysphonia resulting in a " Kermit-the-frog-like" voice and shortness of breath developed to dyspnea.

Histology, the biopsy reveals that thickening of reticular dermis with large collagen bundles separated from another by clear space filled with mucin, resulting in fenestration of the dermis. Fibroblasts are normal in number and morphology. The Alcian blue stain revealed mucin between collagen bundles.

Treatment of SAB is quite difficult. No single therapy has been consistently effective for treatment of SAB. However, there have been case reports of successful treatment with Psoralen+Ultraviolet A (PUVA), UVA1[13], narrowband UVB,[14] cyclosporine, allopurinol, high dose penicillin, localized electron beam therapy and IVIG.[15] Treatment with methotrexate and systemic corticosteroid has been disappointing. The patient who was experiencing worsening dyspnea and functional disability requiring immediate therapeutic intervention was reported.

In this patient, she refused the phototherapy treatment because she is inconvenient to go to the hospital. The disease is asymptomatic and does not disturb her quality of life. So the follow-up and observation is suitable for this patient.

Reference:

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