A Rare Case of Tumoral Scleromyxedema

Ali Sadeghinia, Mohammad Al Salman, Jafar Taghizadeh Fazli, Pedram Normohammadpour, Amir Hooshang Ehsani, Seyed Naser Emadi

Abstract
Scleromyxedema is an uncommon disease, affecting the skin mainly and other internal organs sometimes, characterized by fibroblasts proliferation, fibrosis, and mucous deposition in the absence of thyroid disorder. It is associated with monoclonal gammopathy in most cases. We are reporting a case with a rare presentation of tumoral scleromyxedema in the neck, with a mass mimicking other tumoral lesions, highlighting the importance of diagnosis and histopathologic correlation.

Key Words: Lichen myxedema, paraproteinemia, scleromyxedema, sclerosis, tumoral scleromyxedema

Introduction
Scleromyxedema is a chronic disease characterized by marked mucin deposition and fibrosis in the dermis presented with multiple firm waxy papules and plaques, systemic manifestations and monoclonal gammopathy. The purpose of this article is to focus on an unusual and confusing presentation that was not discussed in previous publication.

Case Report
We report a case of a 55-year-old male patient with an infiltrative slowly progressive firm, reddish, nontender pruritic waxy plaque with a cobblestone appearance on the neck extending to the mandible and upper chest with some minimal prominent lymph nodes in bilateral jugular chains from 9 months ago. The patient has no history of weight loss or any other systemic symptoms [Figures 1 and 2].

Spiral CT scan of the neck, chest, abdomen, and pelvis showed the vascular soft-tissue mass with 82 mm × 50 mm size anterior to the thyroid and trachea with some minimal prominent lymph nodes in bilateral jugular chains from 9 months ago. The patient has no history of weight loss or any other systemic symptoms [Figures 1 and 2].

Spiral CT scan of the neck, chest, abdomen, and pelvis showed the vascular soft-tissue mass with 82 mm × 50 mm size anterior to the thyroid and trachea with some minimal prominent lymph nodes in bilateral jugular chains from 9 months ago. The patient has no history of weight loss or any other systemic symptoms [Figures 1 and 2].

Immunohistochemistry (IHC) was done to support the diagnosis and to rule out other suggested diagnosis such as inflammatory myofibroblastic tumor and dermatofibrosarcoma protuberans (DFSP); Ki67: 3%–5% in inflammatory cells, rare in spindle cells [Figure 4]; Smooth muscle actin (SMA): positive in spindle cells of the tumor [Figure 5]; Desmin: negative in tumor cells [Figure 6]; CD34: positive in vascular walls and focally in spindle cells [Figure 7]; CD31: positive in vascular walls [Figure 8]; Immunoglobulin G (IgG) 4: negative; Anaplastic lymphoma kinase (ALK): negative; CD99: negative; Vimentin: positive. Linear endoscopic ultrasound examination was performed for the mass around the gastric artery showing ill-defined hypoechoic soft-tissue closed to the celiac region. Fine-needle aspiration was performed, but no malignant cells were found in the cytology.

Discussion
Scleromyxedema is an uncommon disease. It affects the middle-aged adults without sex predilection. It is characterized by a widespread symmetric eruption as interlacing bundles between collagen fibers, in a marked mucinous background which was compatible with the diagnosis of scleromyxedema [Figure 3]. Immunohistochemistry (IHC) was done to support the diagnosis and to rule out other suggested diagnosis such as inflammatory myofibroblastic tumor and dermatofibrosarcoma protuberans (DFSP); Ki67: 3%–5% in inflammatory cells, rare in spindle cells [Figure 4]; Smooth muscle actin (SMA): positive in spindle cells of the tumor [Figure 5]; Desmin: negative in tumor cells [Figure 6]; CD34: positive in vascular walls and focally in spindle cells [Figure 7]; CD31: positive in vascular walls [Figure 8]; Immunoglobulin G (IgG) 4: negative; Anaplastic lymphoma kinase (ALK): negative; CD99: negative; Vimentin: positive. Linear endoscopic ultrasound examination was performed for the mass around the gastric artery showing ill-defined hypoechoic soft-tissue closed to the celiac region. Fine-needle aspiration was performed, but no malignant cells were found in the cytology.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Sadeghinia A, Al Salman M, Fazli JT, Normohammadpour P, Ehsani AH, Emadi SN. A rare case of tumoral scleromyxedema. Indian J Dermatol 2020;65:310-2

Received: September, 2018. Accepted: November, 2018.

From the Department of Dermatology, Razi Hospital, Tehran University of Medical Sciences, Tehran, Iran

Address for correspondence:
Dr. Mohammad Al Salman, Razi Hospital, Vahdate-Eslami Square, 11996, Tehran, Iran.
E-mail: al_salman85@me.com
of small, waxy, and firm papules that are closely spaced and often arranged linearly. The papules are typically 2–3 mm, dome-shaped, or flat-topped. The most commonly affected areas include the face, neck, distal forearms, and hands, with sparing of the palms, scalp, and mucous membranes. Itching is not rare. The affected skin may exhibit diffuse erythema, edema, and a brownish-discoloration. As the condition progresses, erythematous and infiltrated plaques may occur with skin stiffening, sclerodactyly, and decreased mobility of the mouth and joints.[1] The systemic manifestations of scleromyxedema are serious and have been fatal in some cases. Mucin has been identified in adventitia around vessels in many systems, including the heart and lungs.[2] Up to 83% of patients with scleromyxedema...
have been described as having a paraproteinemia, predominantly of the IgG-λ subtype. The disease may progress to multiple myeloma in 10% of cases.[3] Other systemic manifestations include myositis, disturbances in the central nervous system and peripheral neuropathy, coma (preceded by flu-like illness), arthralgia and arthritis, renal disease, lung involvement, dysphagia, and laryngeal involvement. Intravenous immunoglobulin (IVIG), alone or in combination with systemic medications, has gained widespread acceptance as first-line therapy for both the cutaneous involvement and associated systemic manifestations, including the dermato-neuro syndrome.[4,5] Melphalan used to be the main treatment for scleromyxedema; however, it may have contributed to patients deaths from the inducement of hematologic malignancies.[6] Other therapies include steroids, methotrexate, cyclophosphamide, thalidomide, cyclosporine, oral retinoids, extracorporeal photopheresis, and plasmapheresis.[7]

In such cases with growing infiltrative tumoral hard plaque, scleromyxedema could mimic other entities and the diagnosis becomes difficult, revealing the importance of clinicopathologic correlation. Scleromyxedema could be mistaken with nephrogenic systemic fibrosis (NSF), scleredema, localized myxedema, mycosis fungoides, nodular amyloidosis, metastasis, DFSP, other mesenchymal tumors, and inflammatory myofibroblastic tumor. It is easy to rule out nephrogenic systemic fibrosis (NSF) clinically, as it develops in patients suffering from renal failure exposed to gadolinium-containing contrast media, and it usually tends to be distributed symmetrically on extremities. Scleredema lacks fibroblast proliferation despite mucin deposition and fibrosis, and myxedema usually appears on the shins with a previous history of Grave's disease or treatment of the thyroid disease. Mycosis fungoides, nodular amyloidosis, and metastasis are easily differentiated on the basis of histologic findings, and general workup for any malignancy source as was done for this patient. Immunohistochemistry (IHC) is sometimes necessary, especially for DFSP to be ruled out. DFSP usually starts as a single slowly growing plaque on the trunk, and histologic findings show storiform fascicles of atypical spindle cells and involvement of the subcutaneous tissue in a pattern seems like a honeycomb, but in early stages, there might be low-cellularity and minimal atypia. Spindle cells are strongly positive for CD34 and negative for factor Xlla. Inflammatory myofibroblastic tumor also called inflammatory pseudotumor is a rare tumor found mostly in internal organs, such as lung, abdomen, mediastinal, genital, and soft tissues, but it can involve the skin rarely. Spindle cell proliferation (predominantly myofibroblastic) and a prominent inflammatory infiltration mostly composed of plasma cells forms the main pathologic findings. Its IHC is positive for the Desmin, SMA, and ALK-1.

Unfortunately, patient’s compliance with any treatment modality was poor.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. Rongioletti F, Rebora A. Updated classification of papular mucinosis, lichen myxedematous, and scleromyxedema. J Am Acad Dermatol 2001;44:273-81.
2. Cokonis Georgakis CD, Falasca G, Georgakis A, Heymann WR. Scleromyxedema. Clin Dermatol 2006;24:493-7.
3. Dinneen AM, Dicken CH. Scleromyxedema. J Am Acad Dermatol 1995;33:37-43.
4. Blum M, Wigley FM, Hummers LK. Scleromyxedema: A case series highlighting long-term outcomes of treatment with intravenous immunoglobulin (IVIG). Medicine (Baltimore) 2008;87:10-20.
5. Rey JB, Luria RB. Treatment of scleromyxedema and the dermatoneuro syndrome with intravenous immunoglobulin. J Am Acad Dermatol 2009;60:1037-41.
6. Brunet-Possenti F, Hermine O, Marinho E, Crickx B, Descamps V. Combination of intravenous immunoglobulins and lenalidomide in the treatment of scleromyxedema. J Am Acad Dermatol 2013;69:319-20.
7. Allam M, Ghozzi M. Scleromyxedema: A case report and review of the literature. Case Rep Dermatol 2013;5:168-75.