DOI: 10.1515/sjdv-2016-0003

Scleromyxedema (Arndt-Gottron Syndrome): a Case Report

Danijela POPOVIĆ*, Mirjana PARAVINA, Dragan JOVANOVIĆ, Vesna KARANIKOLIĆ, Dragana LJUBISAVLJEVIĆ

Clinic of Skin and Venereal Disease, Clinical Center Niš, Serbia
Medical Faculty, University of Niš, Serbia

*Correspondence: Danijela Popović, e-mail: danijelapopovicnis@gmail.com

DE GRUYTER OPEN

UDK 616.5-004

Abstract

Lichen myxedematosus, also known as papular mucinosis, is a primary diffuse cutaneous mucinosis. It is a rare cutaneous myxedematous condition characterized by formation of numerous lichenoid papules. Scleromyxedema, also known as Arndt–Gottron syndrome, is a rare, confluent, papular and sclerotic variant of lichen myxedematosus, characterized by diffuse thickening of the skin underlying the papules. The condition is associated with systemic, even lethal manifestations, fibroblast proliferation and accumulation of acid mucopolysaccharides of the hyaluronic acid. Serum IgG class paraproteinemia is always present and it can be detected in all patients if appropriate or even repeat testing is used.

Herein, we present a 67-year-old patient with a 2-year history of skin problems. He had no health problems other than hypertension and diabetes, wich were both diagnosed 15 years before. On examination, the patient exhibited sclerodermoid lesions with diffuse pseudo-sclerodermatous thickening of the exposed skin, microstomia and sclerodactyly-like changes; on the face, there were numerous solid, shiny 2 - 4 mm in diameter skin-coloured lichenoid papules, scattered across the forehead, glabellar area, nasolabial folds, perioral region, ear lobes and the neck. Histopathological examination revealed: highly distinctive collagenosis and fibrosis in the middle dermis, increased fibroblasts; collagen bundles with irregular arrangement and fragmentation; alcian blue-positive deposits with appearance consistent with acid mucins. Serum protein electrophoresis detected IgG lambda paraproteinemia. The patient was treated with systematic corticosteroids during 9 months with subsequent introduction of methotrexate and showed satisfactory results.

The etiology of scleromyxedema remains unknown, since the purified IgG paraprotein itself has no direct effects on fibroblast proliferation. In scleromyxedema, numerous therapeutic modalities are proposed, unfortunatelly with limited effects.

In colclusion, we report a case of an adult male with lichenoid papules; after a two-year progression, they evolved into scleromyxedema and exhibited well response to conventional therapy.

Key words

Scleromyxedema; Skin Diseases; Diagnosis; Therapeutics; Case Reports; Treatment Outcome

Lichen myxedematosus, also known as papular mucinosis, is a primary diffuse cutaneous mucinosis. It is a rare cutaneous myxedematous condition characterized by formation of numerous lichenoid papules. Scleromyxedema, or Arndt–Gottron syndrome, is a rare, confluent, papular and sclerotic variant of lichen myxedematosus, which is characterized by diffuse thickening of the skin that underlies the papules. The condition is associated with systemic, even lethal manifestations, fibroblast proliferation and accumulation of acid mucopolysaccharides of the hyaluronic acid. Serum IgG class paraproteinemia is always present and it can be detected in all patients after appropriate or repeat testing (1, 2, 3).

A case of an adult male with IgG paraproteinemia, in whom lichenoid papules evolved to scleromyxedema after two years, is reported.

Case Report

A 67-year-old patient was referred with a 2-year-long history of skin lesions that initially started on the forehead and earlobes, and then subsequently spread to the trunk and extremities. The patient was previously treated as an out-patient with the following diagnoses: steatocystoma multiplex< xanthelasma< erythroderma. He had no health problems other than hypertension and diabetes mellitus, wich were both diagnosed 15 years before.

On admission, the patient exhibited sclero-dermoid lesions, with diffuse pseudosclerodermatous thickening of the exposed skin, microstomia and

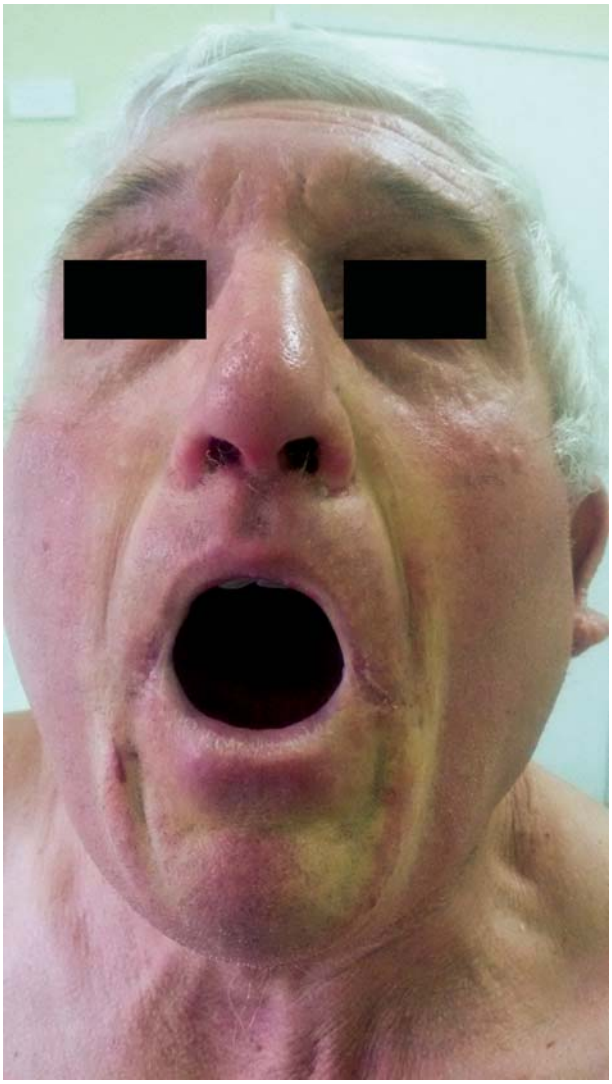

Figure 1. Persistent skin induration on the face, forehead, nasolabial area, and chin with microstomy

Figure 2. Diffuse thickening of the skin of the hands

sclerodactyly-like changes (Figures 1, 2, 3), as well as elephatiasis thickening on the trunk and extremities (Figures 4, 5, 6, 7) There were numerous lichenoid, solid, hemispherical, shiny, skin-coloured papules 2 - 4 mm in diameter, scattered across the forehead, glabellar area, nasolabial folds, perioral region, ear lobes, with linear distribution on the neck (Figures 8, 9, 10).

Laboratory test results

Laboratory tests revealed the following abnormal test results: elevaed erythrocyte sedimentation rate - 19, C-reactive protein - 15,5 mg/L (normal range 0 - 5 mg/L), fibrinogen - 6,1 g/L (normal range 1,86 - 3,86 g/l), glucose - 9,2 mmol/L, (normal range 3,9 - 6,1 mmol/l), glycated hemoglobin (HbA1c) level - 9,30% (normal range 4.8 - 5.9), b2 microglobulin - 3,27 (normal range 0,97 - 2,64 mg/l). Serum protein electrophoresis revealed "M spike" present in the lambda fraction, and immunofixation showed IgG lambda paraprotein; urinalysis for free light chains (Bence-Jones proteins) was negative. All other findings were within normal limits including the following: baseline laboratory tests such as complete blood count,

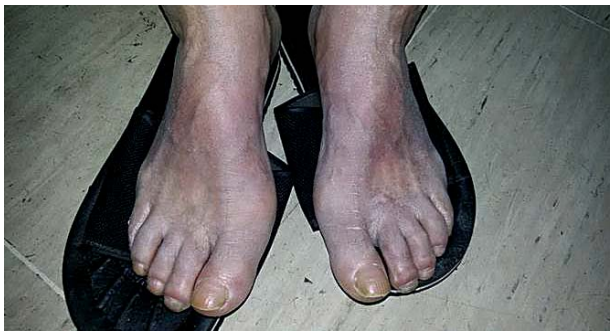

Figure 3. Diffuse hyperpigmentation and induration of the skin of the legs

Figure 4. Elephantiasis thickening of the skin of the trunk

Figure 5. Skin thickening on the trunk with coarse folds

serum electrolytes, blood urea nitrogen and creatinine, aspartate aminotransferase, alanine aminotransferase, gamma-glutamyl transpeptidase, amylase, lipase, creatine phosphokinase, lactate dehydrogenase, alkaline phosphatase, rheumatoid factor, antistreptolysin titer, lipid status, ferritin, thyroid hormones - thyroxine, triiodothyronine and pituitary thyroid stimulating hormone, as well as antinuclear antibodies, antibodies to double-stranded DNA (dsDNA), antibodies to Sjogren's Syndrome related antigen A (anti-Ro/SS-A), antibodies to Sjogren's Syndrome related antigen B (anti-SS-B/La), antibodies to topoisomerase (anti-Scl-70), anticentromere and anticardiolipin antibodies.

Histopathological analysis

Histopathological analysis of skin biopsy specimens showed: profound epidermolytic hyperkeratosis in the epidermis; discrete lymphocytic perivascular infiltrate in the upper dermis, flattening of the epidermal dermal junction, atrophy of the adnexal structures; highly distinctive collagenosis and fibrosis; increased fibroblasts; collagen bundles exhibiting irregular arrangement and fragmentation; alcian blue-positive deposits in reticular dermis, with appearance consistent with acid mucins (Figure 11).

Histopathological analysis of bone marrow biopsy specimens was normal.

Figure 6. Numerous lichenoid papules on the arms

Nailfold capillaroscopy

Nailfold capillaroscopy showed a significant decrease in the number of capillaries, which were fragile and irregularly distributed but without markedly dilated capillary loops.

Radiography

Chest and esophageal passage x-ray (including barium swallow) were normal.

Bone redaiographs of the hands revealed osteodegenerative changes with narrowing of the distal interphalangeal spaces.

Ultrasonography

Ultrasound of the upper abdomen revealed a cyst, 10 mm in diameter, in the right kidney.

Figure 7. Diffuse hyperpigmentation and induration of the skin of the legs

Specialist consultations

Specialist consultations established the diagnosis of insulin-dependent diabetes, therefore insulin therapy was indicated.

Therapy

Apart from insulin, systemic methylpred-nisolone was initiated parenterally at a dose of 60 mg daily, with gradual dose reduction and conversion to oral therapy. Two months later, the clinical status showed considerable improvement: the number of lichenoid papules on the face (Figure 12) and on the neck was reduced; elephantiasic skin folds and hyperpigmentation dramatically decreased, especially on the trunk and extremities (Figures 13,

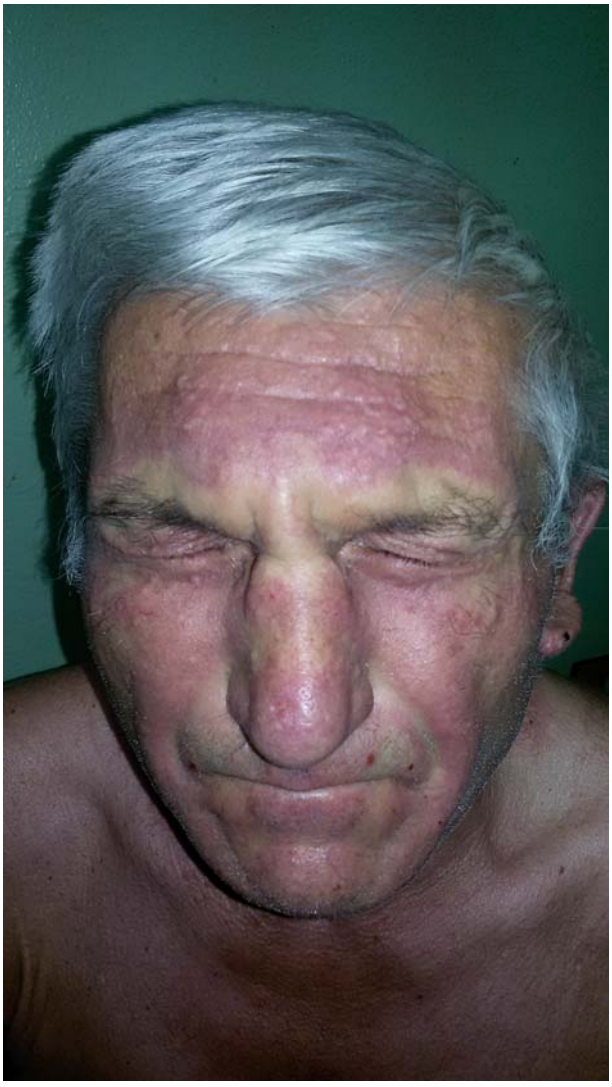

Figure 8. Numerous lichenoid papules - especially on the forehead and in the glabellar region

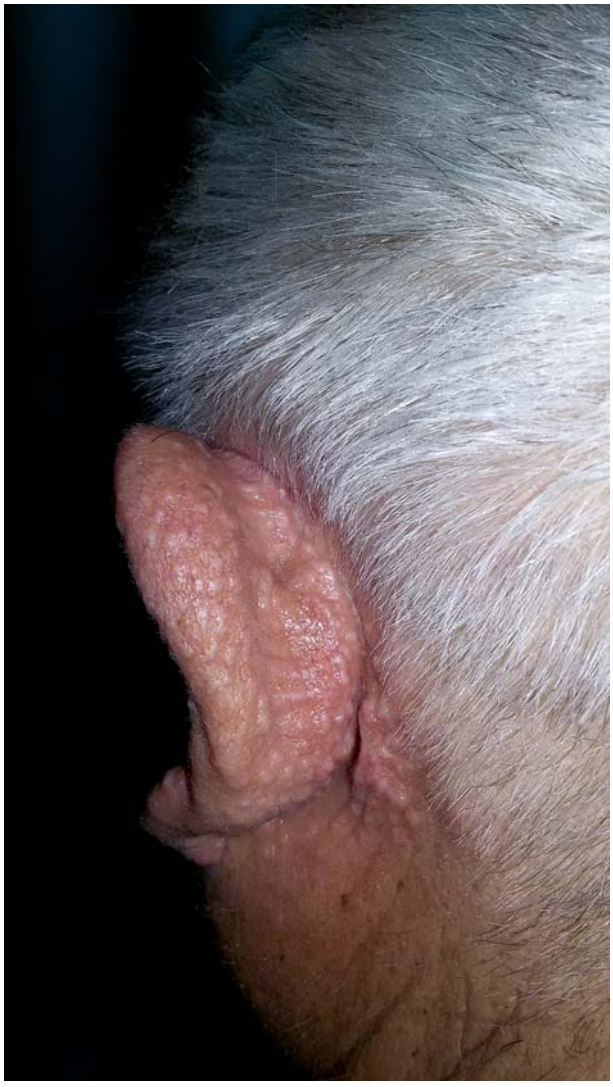

Figure 9. Solitary, hemispherical, solid papules on the earlobes

14). The most resistant to therapy were nasolabial folds, earlobes and hands, where induration, papules and sclerodermoid appearance were still present, but with a subjective feeling of better mobility of fingers. After being dismissed from the hospital, the patient was treated with prednisone tablets - 30 mg per day. Six months later, due to persistent skin induration of the face, cheeks, chin and microstomy, the dose was increased to 40 mg per day and methotrexate was introduced at a weekly dose of 12,5 mg as an adjunct therapy, which resulted in marked improvement once again.

Discussion

Scleromyxedema (papular mucinosis, lichen myxedematosus, lichen fibromucinodosis, lichen myxedematosus generalisatus et sclerodermoides, Arndt Gottron) is a variant of cutaneous mucinosis (1, 2). The original description of cutaneous mucinosis was given by Dubreuilh in 1906 and Reitmann in 1908 (4, 5).

All cutaneous mucinoses are divided into primary and secondary. Primary mucinoses can be subdivided into diffuse degenerative inflammatory mucinoses, focal hamartomatous neoplastic mucinoses and

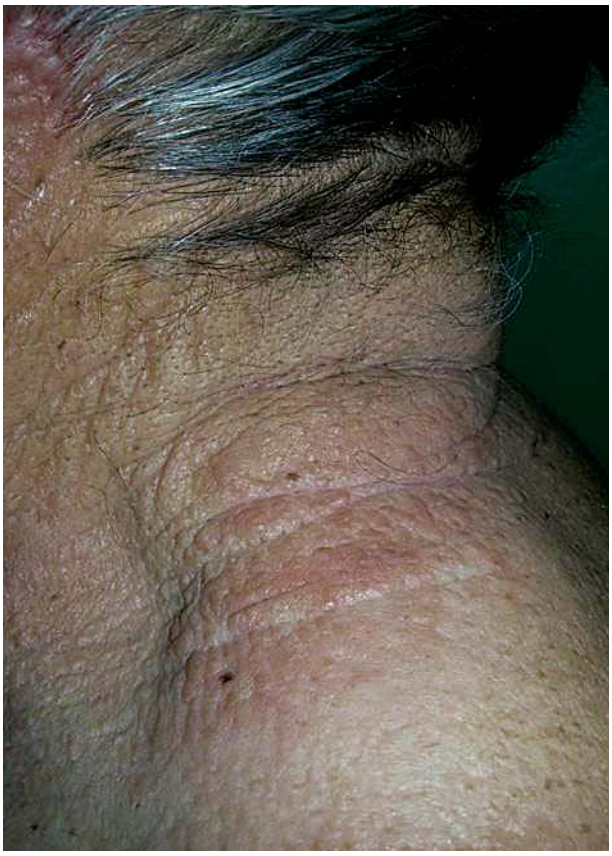

Figure 10. Linear papules on the neck

follicular mucinoses. The group of degenerative inflammatory mucinoses comprises several different conditions, such as generalized and several localized forms of lichen myxedematosus (LM) (1).

In 1953, Montgomery and Underwood distinguished 4 types of LM: 1) generalized lichenoid eruption, later called scleromyxedema; 2) discrete papular form; 3) generalized or localized lichenoid plaque form; and 4) urticarial plaque form (6). The term "scleromyxedema" was coined by Gottron in 1954 to signify the generalized lichenoid papular eruption with sclerodermoid appearance (7). In the literature, terms LM, papular mucinosis, and scleromyxedema have been often used indiscriminately as synonyms, but in 2001 based on personal experience and literature review, Rongioletti concluded that most reported cases of LM or papular mucinosis published without indication of the subtype, appeared in fact to be cases of scleromyxedema. He also recognized two clinico-pathological subsets of LM, a generalized papular and a sclerodermoid form (also called scleromyxedema) with systemic, even lethal, manifestations, and a localized papular, more benign form, without a demonstrable paraprotein, with 5 subtypes: 1) a discrete papular form involving any site; 2) acral persistent papular mucinosis involving only the extensor surface of the hands and wrists; 3) self-healing papular mucinosis, of a juvenile and adult type; 4) papular mucinosis of infancy, a pediatric variant of the discrete form or of acral persistent papular mucinosis; and 5) nodular form (8).

Scleromyxedema is a rare chronic cutaneous myxedematous condition of the connective tissue (1, 9, 10) which occurs in middle-aged individuals (11), most commonly between the ages of 30 and 80, in all ethnic groups and equally in both sexes (10, 11). The etiopathogenesis of scleromyxedema hasn't been fully clarified. There is hyperproliferation of dermal fibroblasts which produces mucin in quantities larger than normal fibroblasts, with increased collagen deposition (3). The serum shows the ability to stimulate fibroblast proliferation in vitro, thus indicating the role of monoclonal paraproteinemia. The presence of monoclonal component (M component) or paraprotein in sera of patients with lichen myxoedematosus and scleromyxedema has been noted in nearly all cases. However, this ability of the serum remains even after the removal of IgG, which indicates that in addition to paraproteinemia, another circulatory factor is responsible. Thus, the role of serum paraproteins in scleromyxedema pathogenesis remains unknown (1, 2, 3, 12). It has been suggested that innate altered regulation of dermal fibroblast growth might play a role in pathogenesis of sclermyxedema (3). Paraproteinemia, or monoclonal gammopathy, delineates an immunoproliferative disorder manifested by the presence of excessive amounts of one monoclonal gamma globulin in blood. There are three types of paraproteins: light chain, heavy chain, and whole immunoglobulin, each of them can be present alone, or combined. Light chains can be excreted through urine, and then they are called Bence-Jones proteins. Potential causes of paraproteinemia include the following: leukemia and lymphatic myeloma (in 8.7% of the cases it is combined with multiple myeloma), plasmacytoma, or it is manifested as in our patient idiopathically as monoclonal gammopathy of unspecified cause. As far

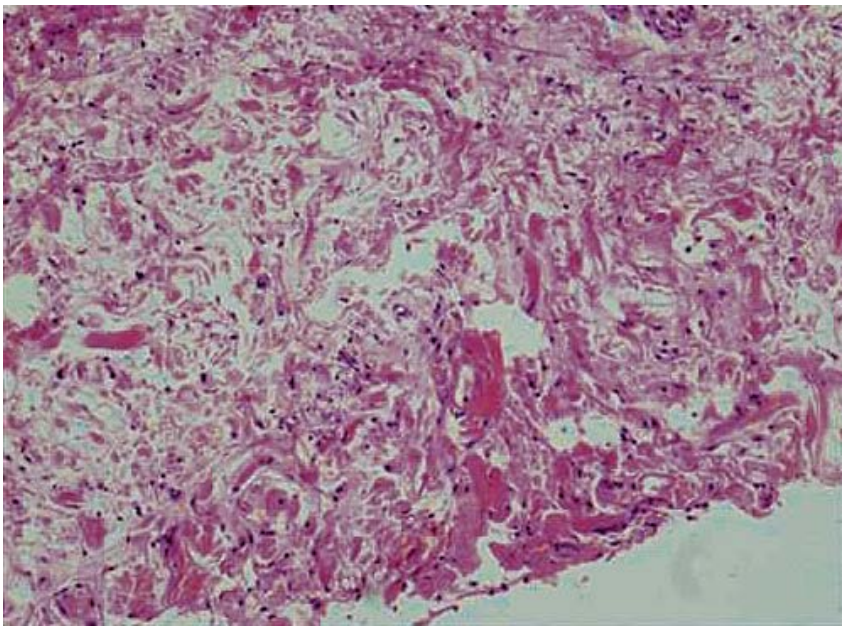

Figure 11. Histopathological analysis of the skin biopsy specimens showed: highly distinctive collagenosis and fibrosis, as well as an increased number of fibroblasts; irregular arrangement and fragmentation of collagen bundles (HE stain, x 100)

as scleromyxedema is concerned, the serum protein electrophoresis most frequently detects a monoclonal type IgG paraprotein of lambda light chain isotype, while IgA and IgM proteins are less common. Paraprotein IgG lambda, also present in our patient, has a molar eght of 110.000 daltons, whereas normal IgG weighs 160.000 daltons. These findings suggest that paraprotein IgG is an incomplete IgG molecule which lost a significant antigen part of Fc fragment. The IgG of lichen myxedematosus differes from the IgG of multiple myeloma not only by usually showing slower electrophoretic migration, but also by the fact that its IgG molecules nearly always possess light chains of the lambda type. The role of the monoclonal IgG in lichen myxedematosus is not clear. Although the serum from patients with lichen myxedematosus stimulates fibroblast proliferation *in vitro*, the purified IgG paraprotein itself has no direct effects on fibroblast proliferation (3).

Scleromyxedema, a generalized papular form of LM is characterized by the presence of the following criteria (13): 1) multiple cutaneous manifestations (papules, nodules, plaques) and sclerodermiform diffuse thickening of the skin of the elephantiasis type; 2) specific histologic findings, including mucin deposits, fibrosis and fibroblast proliferation; 3) monoclonal paraproteinemia; 4) absence of thyroid disease; 5) potential systematic spreading, sometimes with lethal outcome. In scleromyxedema, these characteristics represent crucial criteria in diagnosing the generalized disease (1). However, in order to diagnose localized LM, the presence of the first and second of the aformentioned characteristics is required, as well as the absence of the third, fourth, and the fifth one. Our patient met 4 of 5 aforementioned criteria: skin lesions, specific histologic findings, monoclonal paraproteinemia and absence of thyroid disease.

There is a triad of histopathologic characteristics of scleromyxedema (8, 3): 1) diffuse mucin deposition in the upper and mid reticular dermis; 2) increase collagen deposition, 3) significant proliferation of irregularly distributed fibroblasts. The epidermis may be normal or thickened due to mucin pressure and fibrosis, hair follicles may be atrophic, as in our patient; slight, perivascular, superficial lymphoplasmacytic infiltrate is often present, as it was in our patient; the elastic fibers are fragmented and decreased in number; in systematic spreading of the disease, mucin may fill the walls of myocardial blood vessels, as well as the interstitium of kidney, pancreas, adrenal glands and

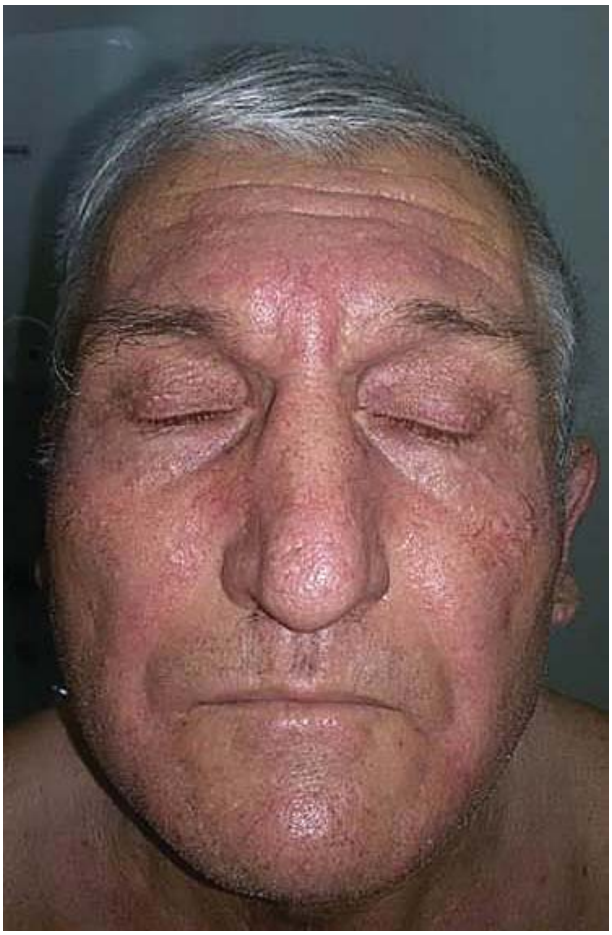

Figure 12. Reduced papular lesions on the skin of the face after 2 months of therapy

nerves (2). The mucin present in mucinous stains: light blue in sections stained with hematoxylin-eosin; alcian blue at 2.5 ph but negative at 0.5 pH and shows metachromasia with toluidine blue at 7.0 pH and 4.0 pH but no metachromasia below 2.0 pH< it is PAS negative, indicating absence of neutral mucopolysacharides, and aldehyde-fuchsin negative (indicating absence of sulfated mucopolysacharides). Regular demonstration of the presence of mucin in the dermis is possible only in pretibial myxedema, in self healing-juvenile cutaneous mucinosis and in lichen myxedematosus (3). There were alcian blue positive-deposits detected in the lesions of our patient.

Our patient presented without a thyroid disease, but he had diabetes mellitus and hypertension. Although no endocrine abnormalities have been found, there are numerous extracutaneous manifestations that can be present in patients with

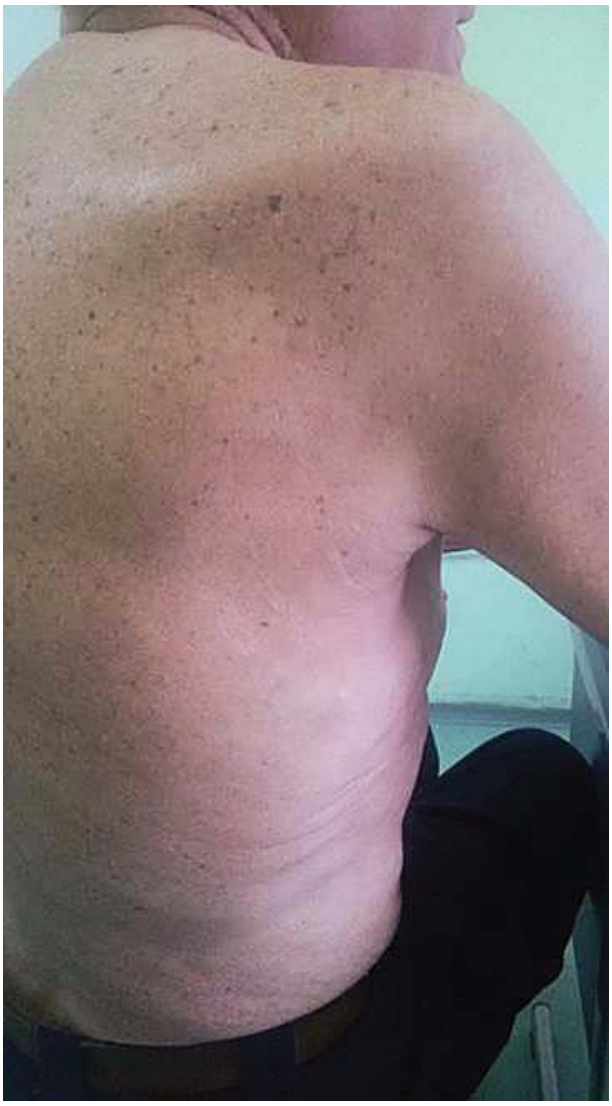

Figure 13. A dramatic improvement of skin lesions over the trunk after 2 months of treatment

scleromyxedema, eg. hematological alterations such as eosinophilia, neurological manifestations (confusion, dysarthria, ascending paralysis, convulsions and coma; combination of high temperature, convulsions and coma), proximal myopathy, inflammatory polyarthritis, laryngeal alterations, esophageal dysfunction, pulmonary restrictive disease, and in 10% cardial alterations (1). Occasionally, systemic involvement may occur in other internal organs. There are reports of hepatitis C virus infection (9), bilateral scleromyxedema of the eyelids (14), and dermato-neuro syndrome (10); malignant hematological neoplasia (multiple myeloma, acute leukemia and

T-cell lymphoma) as well as cancers, including thymic carcinoma (15).

Scleromyxedema can be clinically differentiated from scleroderma by the presence of papules and the absence of telangiectasias: scleromyxedema, presents with generalized eruption of papules as in lichen myxedematosus and diffuse erythematous tickening of the skin which is movable over the subcutis, not bound down as in scleroderma (1, 3).

No established standard therapy exists for systemic treatment of scleromyxedema. Various medications and methods are used with varying, mostly insufficient therapeutic effects (11). These include: topical application and intralesional injection of hyaluronidases, topical, intralesional and systemic administration of corticosteroids, radiotherapy, psoralen and ultraviolet A (PUVA) phototherapy, plasmapheresis combined with pulsed corticosteroid and/or immunosuppressive therapy, intravenous immunoglobulin combined with thalidomide, extracorporeal photochemotherapy (12, 16, 17, 18), retinoids (11), peripheral blood autologous stem cell transplantation (16). Aggressive surgical interventions may be indicated for palliative care, esthetic corrections, and functional training. Melphalan, as a cytoreduction chemotherapeutic agent, has been considered as the first line therapy for decades (16, 18). Our patient was treated with systemic corticosteroids continuously during 9 months, with subsequent introduction of methotrexate. Satsifactory results have been achieved, which was also reported by other authors (19).

The disease has a chronic progressive course. Spontaneous improvement is possible, but extremely rare. The possibility of other organs being affected must also be taken into consideration (11). The prognosis is rather poor. Lethal outcome is also possible as a result of non-specific complications, but also hematological malignancy (1). Clinical staging of scleromyxedema has been proposed as follows: 1) limited cutaneous papular mucinosis; 2) generalized cutaneous mucinosis and/or extracutaneous manifestations; 3) generalized cutaneous mucinosis and/or extracutaneous manifestations with a Karnofsky performance status less than 50% (20, 21). The Karnofsky Performance Status (KPS) is a widely used method to assess the functional status of patients.

Full staging investigations should always be undertaken before making any decisions concerning the treatment strategy. Our patient presented with the first limited cutaneous stage of the disease.

Conclusion

We reported a case of an adult male in whom lichenoid papules of lichen myxedematosus evolved into scleromyxedema after a two-year pregression with no extracutaneous manifestations, and good response to conventional therapy.

Abbreviations

LM - lichen myxedematosus
HbA1c - glycated hemoglobin A1c
ds DNA - double stranded deoxyribonucleic acid
Ro/SS-A - Sjogren's Syndrome-related antigen A
SS-B/La - Sjogren's Syndrome-related antigen B
Scl-70 - topoisomerase 1
KPS - Karnofsky performance status

References

1. Breathnach SM. Mucinoses. In: Burns DA, Breathnach SM, Cox NH, Griffiths CEM, editors. Rook`s textbook of dermatology. 8th edition. Oxford: Blackwell Publishing Ltd; 2010. p. 59.21-59.30.
2. Rongioletti F, Rebora A. Mucinosis. In: Bolognia JL, Jorizzo JL, Rapini RP, editors. Dermatology. 2nd edition. London: Mosby Elsevier; 2008. p. 611-21.
3. Maize J, Maize J Jr, Metcalf J. Metabolic diseases of the skin. In: Elder DE, Elenitsas R, Johnson BL Jr, Murphy GF, editors. Lever's histopathology of the skin. 9th ed. Philadelphia: Lippincott Williams & Wilkins; 2005. p. 435-67.
4. Dubreuilh W. Fibromes miliares folliculaires: sclerodermie consecutive. Arch Dermatol Syph 1906;37:569-72.
5. Reitmann K. Uber eine eigenartige, der sklerodermie nahestehende affection. Arch Dermatol Syph 1908;92:417-24.
6. Montgomery H, Underwood LJ. Lichen myxedematosus (diferentiation from cutaneous myxedemas or mucoid states). J Invest Dermatol 1953;20(3):213-36.
7. Gottron HA. Skleromyxödem; eine eigenartige Erscheinungsform von Myxothesaurodermie. Arch Klin Exp Dermatol 1954;199(1):71-91.
8. Rongioletti F, Rebora AN. Updated classification of papular mucinosis, lichen myxedematosus, and scleromyxedema. J Am Acad Dermatol 2001;44(2):273-81.
9. Azfar NA, Khan AR, Zaman T, Jahangir M. Scleromyxedema in an HCV- positive male. Journal of Pakistan Association of Dermatologists 2008;18:116-8.
10. Savran Y, Akarsu S. Dermato-neuro syndrome in a case of scleromyxedema. Eur J Rheumatol 2015; DOI:10.5152/eurjrheum.2015.0116.
11. Serdar ZA, Altunay IK, Yasar SP, Erfan GT, Gunes P. Generalized papular and sclerodermoid eruption: scleromyxedema. Indian

J Dermatol Venereol Leprol 2010;76(5):592.
12. Weening RH, Pittelkow MR. Scleredema and scleromyxedema. In: Wolff K, Goldsmith L, Katz S, Gilchrest B, Leffell D, Paller A, editors. Fitzpatrick`s dermatology in general medicine. 7th edition. New York: Mc Graw Hill Medical; 2008. p. 1562-6.
13. Salas-Alanis JC, Martinez-Jaramillo B, Gomez-Flores M, Ocampo-Candiani J. Scleromyxedema, a therapeutic dilemma. Indian J Dermatol 2015;60(2):215.
14. Shams M, Javadi MA, Bidaki R. Bilateral scleromyxedema of eyelids: a rare report in psychodermo-ophthalmology: clinical image. JSM Ophthalmol 2015;3(2):1030.
15. Chan JC, Trendell-Smith NJ, Yeung CK. Scleromyxedema: a Cutaneous Paraneoplastic Syndrome associated with thymic carcinoma. J Clinical Oncol 2012;30(3):e27-9.
16. Laimer M, Namberger K, Massone C, Koller J, Emberger M, Pleyer L, et al. Vincristine, idarubicin, dexamethasone and thalidomide in scleromyxoedema. Acta Derm Venereol 2009;89(6):631-5.
17. Efthimiou P, Blanco M. Intravenous gammaglobulin and thalidomide may be an effective therapeutic combination in refractory scleromyxedema: case report and discussion of the literature. Semin Arthritis Rheum 2008;38(3):188-94.
18. Meigel W. Mucinoses. In: Burgdorf WH, Plewig G, Wolf HH, Landthaler M, editors. Braun-Falco`s dermatology. 3th ed. Munich: Springer; 2009. p. 1263-73.
19. Mehta V, Balachandran C, Rao R. Arndt Gottron scleromyxedema: successful response to treatment with steroid minipulse and methotrexate. Indian J Dermatol 2009;54(2):193-5.
20. Donato ML, Feasel AM, Weber DM, Prieto VG, Giralt SA, Champlin RE, et al. Scleromyxedema: role of high-dose melphalan with autologous stem cell transplantation. Blood 2006;107(2):463–6.
21. Péus D, Newcomb N, Hofer S. Appraisal of the Karnofsky Performance Status and proposal of a simple algorithmic system for its evaluation. BMC Med Inform Decis Mak 2013;13:72.

Skleromiksedem (Arndt–Gottron syndrome) – prikaz slučaja

Sažetak

Uvod. Miksedematozni lihen (lat. *lichen myxoedematosus*) takođe poznat pod nazivom papulozna mucinoza, klasifikovan je kao primarna kutana difuzna mucinoza. Predstavlja retko miksedematozno stanje u koži koje karakteriše formiranje brojnih lihenoidnih papula. Skeromiksedem, poznat i kao Arnt–Gotronov (*Arndt-Gottron)* sindrom, retka je papulozna sklerodermiformna varijanta miksedematoznog lihena, u kojoj aglomerirane papule rezultuju difuznim zadebljanjem kože. Stanje je povezano sa sistemskim, čak i letalnim, manifestacijama, tako da se razlikuju tri stadijuma skleromiksedema: 1) limitirana, kutana papulozna mucinoza; 2) generalizovana kutana mucinoza i/ili ekstrakutane manifestacije; 3) generalizovana kutana mucinoza i/ili ekstrakutane manifestacije uz *Karnofski status* manji od 50%.

U osnovi patogenetskog mehanizma skleromiksedema je proliferacija fibroblasta i akumulacija kiselih mukopolisaharida tipa hijaluronske kiseline. Paraproteinemija IgG klase je prisutna u serumu i može biti detektovana kod svih pacijenata ukoliko se sprovodi na adekvatan način, ponekad u više ponovljenih pokušaja.

Dijagnoza bolesti se postavlja na osnovu kliničke slike, histopatološkog nalaza depozita kiselih mucina u retikularnom dermisu, proliferisalih fibroblasta i fragmenacije umnoženih kolagena vlakna, uz prisustvo u serumu monoklonalne gamapatije. U diferencijalnoj dijagnozi potrebno je razlikovati sklerodermiju, u kojoj nema papula, a zadebljala koža nije čvrsto srasla za potkožno masno tkivo.

Prikaz slučaja. Prikazujemo pacijenta starosti 67 godina, sa dvogodišnjom evolucijom promena na koži. Pacijent nije imao drugih zdravstvenih tegoba osim hipertenzije i dijabetesa melitus, koji su dijagnostikovani 15 godina ranije.

Tokom ispitivanja, pacijentova koža je imala sklerodermoidni izgled sa difuznim pseudo-sklerodermatskim zadebljanjem, ”mikrostomijom” i promenama sličnim sklerodaktiliji; na licu, u predelu glabele na čelu, nazolabijalnim brazdama, perioralnoj regiji, ušnim školjkama i vratu, bio je prisutan veliki broj sjajnih, uniformnih, 2–4 mm u dijametru lihenoidnih papula, normalne boje kože.

U isečku ledirane kože, histopatološkom analizom otkrivene su sledeće promene: kolagenoza i fibroza u retikularnom dermisu; između kolagenih vlakana povećan broj fibroblasta; nepravilan raspored i fragmentacija kolagena usled alcijan plavo-pozitivnih depozita koji bi mogli odgovarati kiselom mucinu. Elektroforezom serumskih proteina detektovana je IgG lambda paraproteinemija.

Pacijent je lečen sistemskim kortikosteroidima tokom 9 meseci; uveden je i metotreksat, što je dovelo do

privremene ali ne i trajne remisije.
Diskusija. Etiologija skleromiksedema ostaje nepoznata pošto za razliku od seruma obolelih, prečišćen IgG paraprotein samostalno nema direktni efekat na proliferaciju fibroblasta.
I pored brojnih terapijskih modaliteta, prognoza oboljenja je loša, a prisustvo paraproteina uvek može značiti uvod u malignu hemopatiju.
Zaključak. U radu je prikazan slučaj odrasle muške osobe, kod koje su lihenoidne papule nakon dvogodišnje progresije evoluirale u generalizovane, difuzne sklerodermoidne lezije skleromiksedema, bez zahvatanja drugih organa, sa dobrim odgovorom na primenjenu kovencijalnu terapiju.

Ključne reči

Skleromiksedem; Kožne bolesti; Dijanoza; Terapija; Prikazi slučajeva; Ishod terapije