ARNDT-GOTTRON SCLEROMYXEDEMA: CASE REPORT

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Abstract

Data on the etiology, pathogenesis and clinical picture of a rare dermatosis of the mucinosis group are presented. Typical clinical manifestations of Arndt-Gottron scleromyxedema in a 61-year-old patient who consulted a dermatologist are described. The diagnosis was confirmed by pathomorphological examination of the biopsy taken from the affected skin.

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skin mucinosis,
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diagnostics,
clinical manifestations.

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Introduction

The clinical feature of any dermatosis diagnosis is based on the analysis of the existing changes in the patient's skin and the identification of the primary morphological element by the dermatovenerologist. The presence of exophytic elements (nodule, tubercle and papule) on a patient's skin inevitably triggers the algorithm of differential diagnostics with skin malformations, benign and malignant skin tumors, neoplastic syndromes, skin lesions in diseases of visceral organs and systemic diseases, including rare metabolic dermatoses (1-4).

Arndt-Gottron scleromyxedema is a rare chronic progressive disease of the mucinosis group which is not associated with thyroid function abnormality. The disease is characterized by mucin precipitation in the skin, sebaceous glands, hair follicles and visceral organs. Mucin is a gel-like substance consisting of proteins, carbohydrates and acid glucosoaminoglycans present in metabolic disorders and produced by dermal fibroblasts. The etiology and pathogenesis of Arndt-Gottron scleromyxedema have not been fully studied, but it is assumed that the key role in the stimulation of fibroblast proliferation can be played by paraproteinemia,

which was detected in 80% of patients, an internal defect of fibroblasts or another not yet identified circulating factor (5, 6).

The disease is a rare dermatosis that occurs mainly in women aged between 40-50 years. Clinical symptoms are characterized by slow formation. The most common localization of the first rashes is the extensor surfaces of the forearms, the back surfaces of the wrists, the face and the auricles. The process begins with the appearance of numerous shiny (side lighting) hemispherical nodules, 3-5 mm in diameter. When they merge, foci with thickened skin are formed, which do not fold. Progressive infiltration of the skin in the face can lead to the distortion of facial expressions, which is why the face acquires a masklike appearance with thickened lips and nose, swollen eyelids. At the same time, the formation of dense edema of the dorsal aspects of the hands can lead to movement restriction of the fingers in the interphalangeal joints and limb dysfunction. Many authors report frequent development of visceral and neurological complications, which worsens the prognosis of the disease and can lead to death due to the development of acute cerebrovascular disease (7-11).

Differential diagnostics of nodular skin lesions in Arndt-Gottron scleromyxedema is carried out with ring-shaped granuloma, dermatomyositis, scleroderma, Scleredema adultorum of Buschke, lupus erythematosus, eosinophilic fasciitis, sarcoidosis and amyloid lichen (7, 9).

Pathomorphological examination of the skin in the upper and middle layers of the dermis reveals mucin precipitations between the split collagen fibers, as well as infiltrates, mainly consisting of fibroblasts and mast cells (9, 10).

To attract the attention of dermatovenerologists to the problem of diagnostics of rare dermatoses in nodular skin lesions, we give a description of a rare clinical case of Arndt-Gottron scleromixedema in a patient.

Case report

We present the case of a 63 years old male patient consulted by a dermatovenerologist of the clinic of Ural Research Institute for Dermatovenerology and Immunopathology for complaints of rashes on the back surface of both hands, extensor surfaces of the elbow joints and the auricles.

Anamnesis morbi: According to the patient, he had been sick for 2.5-3 years, when he first noticed the appearance of rashes on the skin of the hands, not accompanied by any subjective sensations. The amount of rashes gradually increased, similar elements began to appear in other areas of the skin too: on the ear auricles and on the extensor surfaces of the elbow joints. The patient began to notice difficulties in moving the fingers of the interphalangeal joints. He did not consult a dermatologist, no treatment was received.

Anamnesis vitae and occupational history of the patient are unremarkable. There are no skin diseases and oncopathology in the family history.

The patient's general condition was satisfactory. The breathing was vesicular, without wheezing. The heart sounds were clear, rhythmic. The abdomen was soft and painless in palpation. The liver was at the costal arch. Bowel and bladder functions were normal.

Local status

The unaffected skin areas had normal color, normal temperature, moisture and turgor. The visible mucous membranes were of normal color. The skin lesions were affecting the skin of the ear auricles, extensor surfaces of the elbow joints and the back surfaces of both hands.

The rash was characterized by very dense nodules, with a diameter of 6-7 mm, with flesh and whitish-yellowish color, hemispherical in shape,

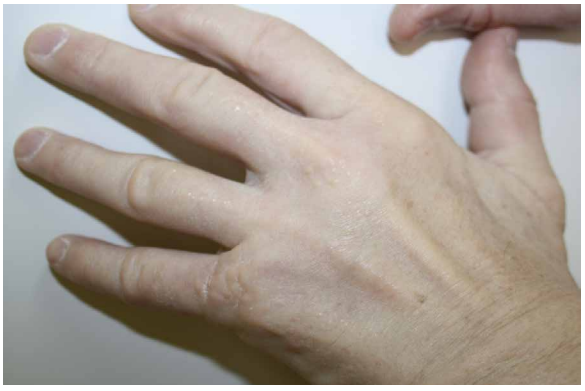


Figure 1. Patient M. Arndt-Gottron Scleromyxedema – numerous nodules on the skin of the back surface of the left hand

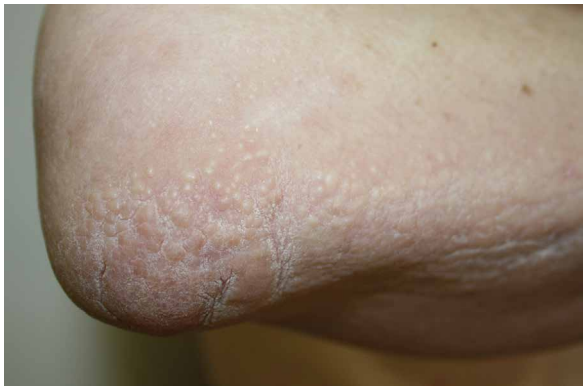


Figure 2. The same patient, numerous nodules and compaction of the skin of the extensor surface of the right elbow joint

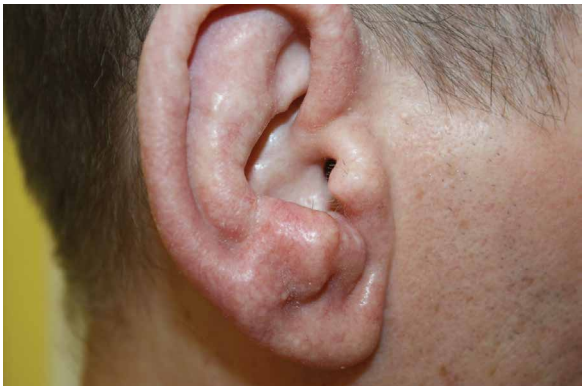


Figure 3. The same patient, deformity of the right auricle

with a waxy sheen. The nodules had a tendancy to merge, forming lesions with dense and thick skin, hard to form a crease, and when pressure was applied, without forming a hole. As a result of dense edema of the skin, the range of motion in the metacarpophalangeal and interdigital joints was limited. The auricles were thickened, had a dense consistency and a bumpy surface due to numerous shiny nodules (Figures 1, 2 and 3). The hair and nail plates were not changed. Red dermographism was present.

Laboratory data

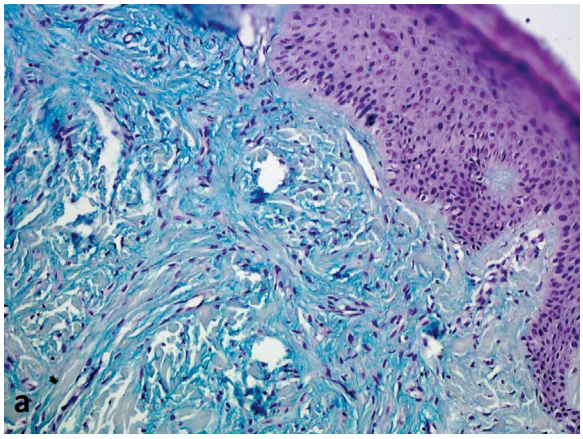
Clinical blood analysis and urinalysis were unremarkable. Serological *Treponema pallidum* test was negative. Antibodies to HIV, hepatitis B and C have not been detected. The biochemical study of blood revealed no pathology of lipid and calcium metabolism.

The results of pathomorphological studies of biopsy material of skin are as follows: in the epidermis there is mild hyperplasia, hyperkeratosis; in the papillary and reticular layers of the dermis, a well-defined nodular focus of loosening and thinning of multidirectional collagen fibers is determined by the accumulation of mucin and proliferation of fibroblasts between them. Alcian blue reaction is positive (Figure 4 A, 4 B).

Based on the medical history, clinical findings and results of pathomorphological studies of the patient's skin biopsy sample, the disease was diagnosed as Arndt-Gottron scleromyxedema. Topical therapy with corticosteroids (Hydrocortisone 17-butyrate (Locoid) 0.1% fatty cream) was assigned. The patient was referred for consultation to related specialists – rheumatologist, endocrinologist, hematologist, neurologist and cardiologist – to clarify any possible complications characteristic of this disease.

Conclusion

The presented clinical case demonstrates the complexity of the diagnostics of a rare disease and emphasizes the need to consolidate the clinical experience of leading dermatovenerologists and modern pathomorphological studies of skin biopsy sample.



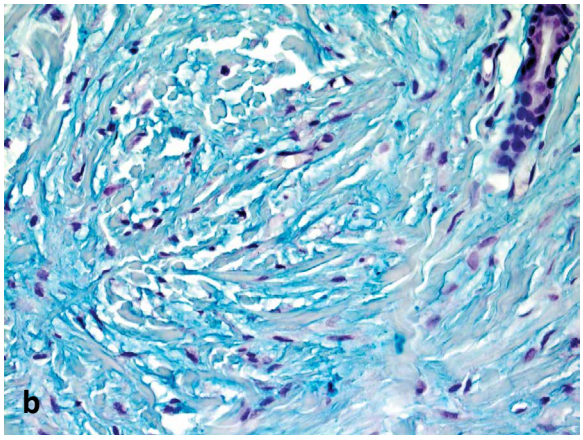


Figure 4. Patient M., histological studies of biopsy material of skin: A) middle hyperplasia and hyperkeratosis of the epidermis, massive mucin precipitations in the upper layers of the dermis between degeneratively altered bundles of connective tissue (Alcian blue and hematoxilin staining, 200 x magnification); B) a part of the reticular layer of the dermis containing mucin accumulations between thickened and partially destroyed collagen fibers, fibroblasts with a altered form of nuclei (Alcian blue and hematoxylin staining, 400 x magnification)

Conflicts of interest: none declared.
Financial disclosure: none.
Patient informed consent obtained.

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