CLINICAL CASE – SCLEREDEMA ADULTORUM

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Abstract

Scleredema of Buschke is a rare dermal disease of unknown etiology which is characterized by a dense diffuse swelling of the derm and subcutaneous adipose tissue. It characterized by induration of the skin which includes a non-pitting hardening of the skin around the neck, shoulders, and trunk sometimes the face. Three basic forms are distinguished: classical, idiopathic, diabetic or metabolic.

The histopathological features of scleredema are characterized by sharply thickened derma, edema and structural impairment of collagenous fascicles metachromasia appears in the foci of the damage as a result of accumulation of glycosaminoglycans. Differential diagnostics was carried out at the other diseases similar to scleredema such as systemic scleroderma, eosinophilic fasciitis, porphyria cutanea tarda hereditaria, porphyria cutanea scleroderma-like form, Werner's syndrome, scleromyxedema, amyloidosis. Several treatments are suggested in the literature.

The present case record describes scleredema adultorum Buschke in a 52 years old male with stiffness and density of the skin of trunk. He had suffered from diabetes mellitus type-1 for more than 30 years. The patient was treated with antibiotics, corticosteroids, polyenzyme agent, hepatoprotector, cytoflavin, polyvitamins. There was improvement with slightly decrease infiltration in foci lesions, skin color in the area of rash brightened.

Skin diseases are common in diabetes mellitus. This clinical case will enable to expand knowledge of general practitioners about the rare dermal disease – Buschke's scleredema giving the possibility to reveal such patients at early stages of the disease in proper time.

Keywords: Scleredema adultorum Buschke, clinical case, infiltration.

1. Introduction

Scleredema of the adults (scleredema adultorum; Greek-Sclåros solid, dense+oidåma tumor; the synonym: swollen scleroderma, Buschke's scleredema (BS)) – a rare dermal disease of unknown etiology, which is characterized by a dense diffuse swelling of the derm and subcutaneous adipose tissue [1, 2]. The question as to the first description of this disease is being discussed till now since some authors consider that this disease was described for the first time in 1876 by Pitford [1, 3], the others – by the German dermatologist Abraham Buschke in 1902 who by the way carried out a differential diagnosis of scleredema with local scleroderma and systemic sclerosis [4]. A relationship with diabetes mellitus was established only in 1970 [5]. According to the data of some authors the term "adult scleredema" is poor because it occurs almost in 1/3 of the patients at the age of 10 years old [6].

The cause of the disease is not known in detail. There are different theories: infectious (the disease arises following the acute infectious diseases such as grippe or some acute respiratory infection, quinsy, pharyngitis, measles, scarlet fever, after which the development of temporary lymphostasis arises as a result of hyperergic streptococcus inflammation), neurogenic (dysfunctions of the peripheral nervous system or hypophysis), endocrine (diabetes mellitus, diturbances on the side of the endocrine gland are of particular importance in the development of the resistant cases), blood circulation disorders, hereditary, genger (women are being ailing two times more often than men) [1, 3, 7–11]. Diabetes mellitus is considered to be a pathogenetic factor. The form that is associated with diabetes is more prevalent in men while other forms are seen more commonly in women. Irreversible glycosylation of collagen and resistance to degradation by collagenase may lead to an accumulation of collagen. Alternatively excess stimulation by insulin, microvascular damage and hypoxia may increase the synthesis of collagen and mucin. Streptococcal hypersensitivity (type I),

injury of lymphatics, and paraproteinemia (type II) may also play the important role [5]. Various hypotheses have been put forward regarding the pathogenesis of scleredema which include obstruction to lymphatic channels by inflammation, streptococcal sensitivity, immune sensitization phenomenon, increased collagen synthesis and mucin deposition due to microvascular damage and resultant hypoxia, and resistance to collagen degradation by collagenases [3]. Very rare cases have been associated with malignant neoplasms or autoimmune disorders. There is only one report describing the association between primary Sjögren's syndrome and scleredema adultorum of Buschke which was established according to the American-European Consensus Criteria. A case of scleredema of Buschke associated with rheumatoid arthritis and Sjögren's syndrome is described too. The onset of the skin changes and rheumatoid arthritis was almost simultaneous and the sicca syndrome developed 4 years later [12].

Three basic forms of BS are distinguished: classical (acute onset following underwent infection, benign evolution, complete recovery in a few months affects primarily middle-aged women but also children), idiopathic (progressive beginning, absence of the relation with infection, lingering of many years clinical course, this type is more frequently associated with a monoclonal gammopathy), diabetic or metabolic (developing in patients more often in men with diabetes mellitus, the onset is subtle and the involvement persistent) [2, 5, 8, 10].

The onset of the disease is mostly sudden. Prodromal phenomena precede the development of induration stage on the skin. Edema of the skin which is of a symmetrical character and induration develop gradually. Posterior and lateral surfaces of the neck and face becoming amimic whereat contraction of the forehead is complicated by opening of the mouth are the usual places of localization. Later on foci acquire diffuse, more often symmetrical character, with dermal lesion of the shoulders, forearms, wrists, upper part of the trunk, but the skin of the abdomen and lower extremities is involved rarely. Erythematic edema is observed in the area of foci lesion at the initial stages. Then the skin becomes thickened, smooth, sharply solid (paraffin density) and acquires waxy color, brilliance, fossa when pressed is not left (due to the deposit of collagen and mucopolysaccharides in derma). The limits of the lesion as a rule are indistinct, foci are extended [3, 4] slowly. Difficulties may be observed in movements at joints. A significant increase of ESR and antistreptolysin-O titre is marked. Mucous tunics are not damaged. Macroglossia may occur frequently. Sensitivity is preserved. Red and white dermographism is noted [4, 13]. Derma is sharply thickened, epidermis is not changed at histological investigation. Edema and structural impairment of collagenous fascicles at the expense of extreme accumulation of the main substance of the connective tissue are marked. Metachromasia appears in the foci of the damage as a result of accumulation of glycosaminoglycans [13, 14].

2. Description of the clinical case

Here we present the case of Scleredema adultorum Buschke.

Patient of 52 years old (**Fig. 1**) a countryman of one of the villages of the Chernivtsi region appealed for medical aid. The patient complained of the skin density of the trunk, stiffness in these areas. According to the data of the past history it was determined that patient suffered from the disease for about 1 year. For the first time induration of the skin was observed by the patient's wife in the back region that occupied the area of about 5×5 cm in diameter. The patient couldn't indicate the reason of the disease or relation with any event. In the past history of the patient there are frequent chills, arterial hypertension and diabetes mellitus type I which the patient has been suffering from for 15. When new foci of rash gradually appeared the patient appealed for medical care to dermatologist of the regional dermatovenerologic dispensary. Objectively: the general state of the patient was satisfactory. Pathological process has current character and is localized in the back area where there are two evident foci of the lesion: one in the upper part of the back on the right side 8–10 cm in diameter, the second one – in the lower part of the back on the right side of 10–15 cm in diameter where the skin and subcutaneous adipose tissue are dense, not forming fold, and leave fossa when palpated. The borders of the foci lesions are not distinct. The skin within foci lesions is of the brownishred colors with violet hue, skin picture is smoothed out.

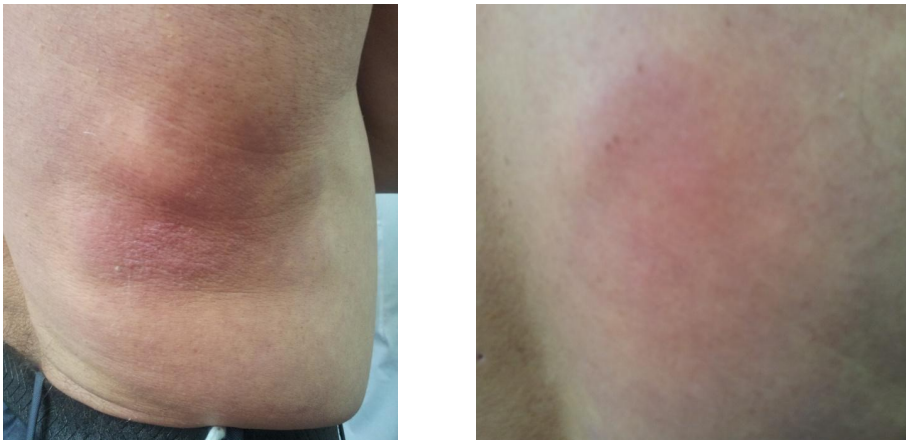

Fig. 1. Patient P., 52 years. Diagnosis – Scleredema adultorum Buschke

3. Results

The patient was administered to undergo instrumental and laboratory examination. General bloodtest: erythrocytes – 3,0×1012/l, Hb – 125g/l, leukocytes – 4,1×109/l, thrombocytes – 240×109/l, eosinofils – 3 %, stab neutrophils – 5 %, segmentonuclear – 46 %, lymphocytes – 42 %, monocytes – 4,0 %, ESR – 15 mm/hour. ESP (end systolic pressure), blood test on hepatits B and C, HIV – infection are negative. Biochemical blood examination: general bilirubin – 13,0 mcmol/l, conjugated bilirubin – 3,4 mcmol/l, unconjugated bilirubin – 9,6 mcmol/l, cholesterol – 4,5 mmol/l, total protein – 75 g/l, albumins – 46 g/l, glucose – 5,5 mmol/l, allotoaminotransferase – 11 un/l, transminase – 16 un/l. Rheumatologic complex (C-reactive protein), antistreptolysin, antihyaluronidase, rheumatoid factor, sialic acids – are without anomaly. General analysis of urine is within norm. US – signs of chronic cholecysto-pancreatitis and diffuse goiter Ist. were revealed during ultrasonic scanning. The patient suffers from arterial hypertension. Pathologic changes on the side of the skeletal-joint system were not detected. The patient was consulted by closely related specialists in order to exclude systemic disease. The results of histological examination were the following: swelling and decomposition of collagenous fibers with hollow spaces among them; availability of the cells of fibroblastic layer with a prevalence of active fibroblasts, lymphocytes and single macrophages; pan vasculitis with polymorphocellular infiltration of perivascular zones were in all fields of vision. On the basis of the data of past history, clinical picture, data of laboratory, histological methods of examinations and consultations and closely-related specialists the patient was made a diagnosis of Buschke's scleredema. The patient was instituted therapy that included: penicillin (500000 units 4 times a day i. m (intramuscularly), systemic glycocarticoid drug (prednizolone with initial dose of 60 mg.); polyenzyme agent (vobenzim) according to a scheme, asparkam, hepatoprotector (glutargin), cytoflavin (that includes xanthinol nikotinat), polyvitamins for taking inside in generally accepted doses. The patient was administered a compound combined ointment locally consisted of heparin, prednizolone, dimexid in equal parts. As a whole the course of treatment constituted 30 days. A month later a repeated course of therapy was administered.

4. Discussion

At the end of complex therapy patient was noticed to have a positive clinical dynamics, subjective sensation of the patient improved, infiltration in foci lesions slightly decreased, skin color in the area of rash brightened. The patient was discharged in the state of improvement and medico-preventive recommendations were given to him. At present patient is registered in the dispensary under our observation.

Differential diagnosis can be made considering the typical clinical features and the histological peculiarity. Differential diagnosis of BS must be carried out first of all at systemic scleroderma that is characterized by the presence of Reino phenomenon, face and distal parts of the extremities are involved into pathologic process. Acrodactylia, microglossia, the presence of telangiectasias and hyperpigmentation, atrophy of the skin and damages of inner organs are observed too. Pathohistological investi-

gation is carried out to make final diagnosis [2, 13]. Eosinophilic fasciitis has certain common signs with Buschke's scleredema, particularly under the conditions of an increase of inflammatory markers however there are differences that consist in presence of the typical data (thickening of the subcutaneous fascia, its lymphocytic and plasmic infiltration) that enable to exclude the given diagnosis completely [4, 13]. Differential diagnosis of BS with porphyria cutanea tarda hereditaria that is characterized with pigmentation arising on the open dermal areas – face, neck, upper part of the breast and rear of the wrists should be also carried out. Foci have a diffuse character, the color of the skin fluctuates from sallow-grey to reddish-bluish and bronzed tint. At the same time vesicles, hypertrichosis may be present on the skin. Porphyria cutanea tarda hereditaria is definitely accompanied with the damage of the liver that is confirmed by clinical and laboratory data. Difficulties in differentiation usually arise in case of porphyria cutanea form similar to scleroderma, however correctly obtained case history (bullous lesions in history), sclerotic lesions usually of the face and neck that are completed with atrophy, absence of the evident skin density, as well as absence of the symptoms of hepatic lesion permit to exclude the given diagnosis [1, 10, 13]. As to Werner's syndrome it is genetically determined disease that makes its debut mostly during the period of puberty and then steadily progresses. Joining of alopecia, thin beaked nose, constricted oral opening and changes from the dermal side – getting thin, skin atrophy of the face and distal portions of the extremities with foci of density like scleroderma, dyschromia and from now on chromic erosive-ulcerous rashes that are badly epithelized [1, 4, 13]. Patients with scleromyxedema also have firm papules (often in a linear array) in addition to dermal induration as well as a proliferation of fibroblasts histologically. The other types of cutaneous mucinoses are usually distinguished on the basis of clinical findings. Additional entities in the sclerodermoid differential diagnosis also should be excluded. Occasionally because of associated erythema patients with diabetes-associated scleredema are sometimes misdiagnosed as having cellulitis (usually by non-dermatologists) [5]. Differential diagnosis of BS must be carried out with amyloidosis. Cutaneous manifestation in amyloidosis depends upon the site of amyloid deposition. Superficial dermal deposition of amyloid produces shiny waxy translucent papules. Flexural areas are sites of predilection, including the eyelids, retroauricular region, neck, axillae, inframammary area, umbilicus, inguinal and anogenital regions. Lesions may also be found on the central face, lips, tongue and buccal mucosa. Other rare cutaneous alterations seen in amyloidosis are: hyperpigmentation, infiltrate similar to scleroderma, alopecia areata or universal, nail dystrophies, cutis laxa and lesions similar to cutis verticis girata in the scalp. Therefore, in case of negative findings in the fat aspirate from a patient with a persistently high clinical suspicion of amyloidosis or progressive disease that has no other explanation for, fat aspiration should be repeated and the aspirate should be examined by a experienced cytopathologist. Diagnosis was confirmed in this case by biopsy of skin lesions using congo red staining [15]. Complications include limited range of motion, restrictive lung disease, dysarthria, dysphagia, skin infections, and poor wound healing [5].

There are different approaches to the management of these patients. Therapy of BS is conducted at in-patient department (dermatological or therapeutic). Corticosteroids are prescribed to the patients in the form of intracutaneous injections round the lesion, injections of antibiotics, lidase, cyclosporine, colkhicyn, low doses of methotrexate, intravenous injections of gamma-globulin, repeated courses of hyperbaric oxidation, warm bath, massage [3, 4, 11]. Patients are also recommended ultraviolet A-1 (UVA-1) therapy that often provokes an increased sensitivity and on the background of accompanied endocrine pathology is less effective [8, 14]. It was reported two cases of scleredema diabeticorum treated successfully with UVA-1 as well as physiotherapy and topical corticosteroids; this approach led to improvement in skin changes and mobility [16]. In case of the patient with primary Sjögren's syndrome was treated with hydroxychloroquine [12]. In this communication physicians have presented a 54 year old man with scleredema successfully treated by PUVA and methotrexate [17]. Therapy is unnecessary for scleredema associated with streptococcal infections because it is self-limited. Regression of scleredema associated with diabetes or monoclonal gammopathy is uncommon and no specific treatment is available. Unfortunately, control of the hyperglycemia does not have any influence on the skin. Besides, it is necessary to examine patient carefully in order to reveal chronic foci of focal infection. Aggressive therapies however should be limited to individuals with disabling disease or systemic manifestations [5].

Prognosis irrespective of the cause of the disease development is in most cases favorable. Usually spontaneous recovering occurs during some months. The process can be rarely delayed for some years [1, 2, 4, 7].

5. Conclusions

This clinical case will enable to expand knowledge of general practitioners about the rare dermal disease – Buschke's scleredema, giving the possibility to reveal such patients on early stages of the disease in proper time. Physicians should suspect scleredema in any patient with diffuse skin thickening when the hands and feet are spared, particularly if diabetes or a preceding febrile episode were present. The observation of patients with BS by closely-related specialists in our case – endocrinologist is important.

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