УДК 616.5-004.3-036.1

L.O. Julei,

L.V. Mudriak

Higher State Educational Establishment of Ukraine "Bukovinian State Medical University", Chernivtsi;
Chernivtsi Regional Dermatovenerologic Dispensary

A RARE CLINICAL CASE-BUSCHKE'S SCLEREDEMA

Key words: *Buschke's scleredema, clinical case, infiltration.*

Abstract. *A clinical case of rare disease of the skin-Buschker's scleredema was described in the article. Differential diagnostics with other diseases similar to scleredema was arried out.*

Scleredema of the adults (scleredema adultorum; Greek-Scleros solid, dense+oidema 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 [3, 4]. 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 [3,10], 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 [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 [9].

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) [3, 6, 7, 8, 10, 11, 12].

Three basic forms of BS are distinquished: classical (acute onset following underwent infection, benign evolution, complete recovery), idiopathic (progressive beginning, absence of the relation with infection, lingering of many years clinical course), diabetic or metabolic (developing in patients more often in men with diabetes mellitus) [4, 7, 11].

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 colour, 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 [5, 10] 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 [1, 5]. Derma is sharply thickened, epidermis is not changed at histological investigation. Edema and structural impairment of collagenic 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 [3, 4].

Differential diagnosis of BS must be carried out, first of all, with systemic scleroderma, tipical for which, is 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 envestigation is carried out to make final diagnosis [1, 4]. Eosinophilic fasciitis has certain common signs with Buschke's scleredema, particularly under the conditions of an increase of inflammatory markers, however, there are differencies, which lie in the presence of the typical data (thickening of the subcutaneous fascia, its lymphocytic and plasmic infiltration), that enable to

exclude the given diagnosis completely [1, 5]. Differential diagnosis of BS with porphyria cutanea tarda hereditaria, which is characterized by 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 available on the skin. Porphyria cutanea tarda hereditaria is definitely accompanied by 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, case history correctly obtained (bullous lesions in history), sclerotic lesions, usually, of the face and neck, which are completed by atrophy, absence of the evident skin density, as well as absence of the symptoms of hepatic lesion permit to exclude the given diagnosis [1, 3, 11] As to Werner's syndrome it is genetically determinated disease, which 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 atrophie 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, 3. 5].

Therapy of BS in conducted at in-patient department (dermatological or therapeutic). Corticosteroids are prescribed to the patients into or 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 [5, 10, 12]. Patients are also recommended ultraviolet A-1 (uva-1) therapy which often provokes an increased sensitivity and against a background of accompanied endocrine pathology is less effective [2, 7]. Besides, it is necessary to examine a patient carefully in order to reveal chronic foci of focal infection. Prognosis, irrespective of the cause of the disease development, is in most cases favourable. Usually spontaneous recovering occurs during some months. The process can be rarely delayed for some years [3, 4, 5, 6].

Here we present the case of Buschke's scleredema.

Patient of 52 years [photo 1, 2], a countryman of one of the villages of the Chernivtsi region appealed for medical aid. The patient complained of the skin

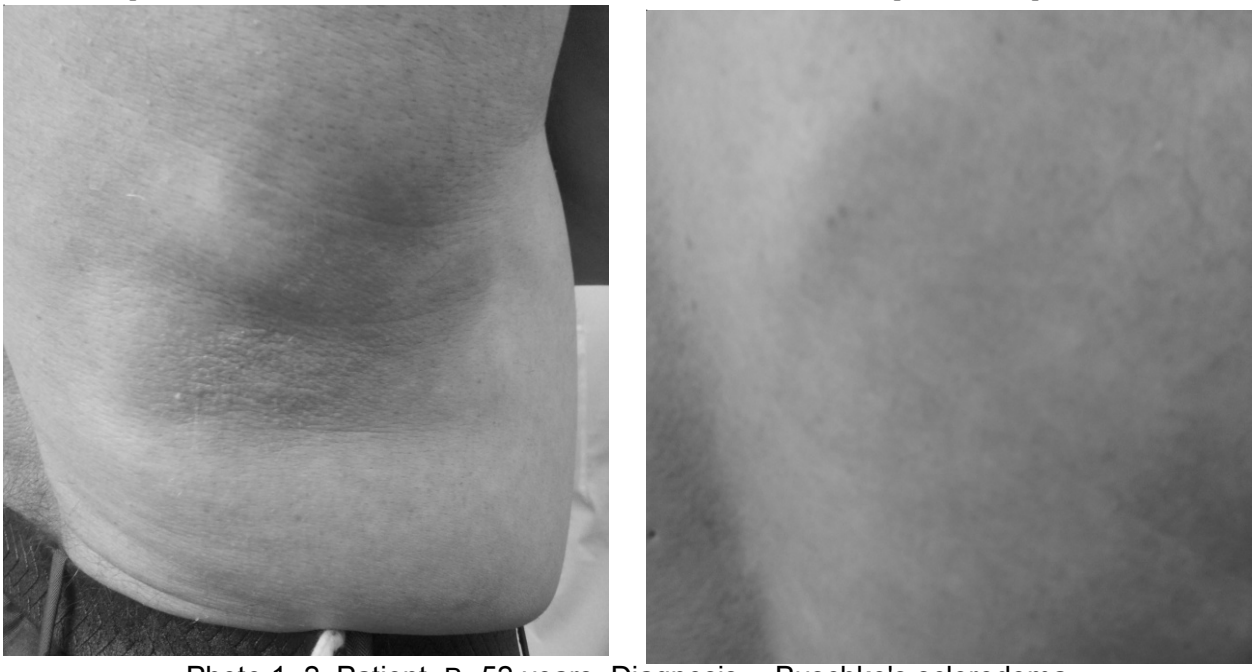

Photo 1, 2. Patient P., 52 years. Diagnosis - Buschke's scleredema

density of the trunk, stiffness in these areas. According to the data of the past history was determined patient to suffer 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 5x5 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 from which the patient has been suffering 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 cha-

racter 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 8-10cm in diameter, the second one - in the lower part of the back on the right side of 10-15cm in diameter, where the skin and subcutaneous adipose tissue are densed, 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 brownish-red colours with violet hue, skin picture is smoothed out.

The patient was administered to undergo instrumental and laboratory examination. General blood test: erythrocytes - 3,0x1012/l, Hb - 125g/l, leukocytes - 4,1x109/l, thrombocytes - 240x109/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 I st. 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, which included: penicillin (500 000 units 4 times a day i. m (intramuscularly), systemic glycocarticoid drug (prednizolone with initial dose of 60mg.); polyenzyme agent (vobenzim) according to a scheme, asparkam, hepatoprotector (glutargin), cytoflavin (which includes xanthinol nikotinat), polyvitamins for taking inside in generally accepted doses. The patient was administered a compound combined ointment locally, which 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. At the end of the complex therapy the patient was noticed to have a positive clinical dynamics, subjective sensation of the patient improved, infiltration in foci lesions slightly decreased, skin colour in the area of rash brightened. The patient was discharged in the state of improvement and medico-preventive recommendatious were given to him.

At present the patient is registered in the dispensary under our observation.

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. The moment of observation of the patients with BS by closely-related specialists, in our case - endocrinologist, is important.

Література. 1. Беренбейн Б.А. Дифференциальная диагностика кожных болезней / Под ред.. Б.А. Беренбейн, А.А. Студницин - 2-изд. перераб. и доп. - М.: Медицина, 1989. - 672 с. 2. Волнухин В.А. УФА-1 терапия локализованной склеродермии и других заболеваний, сопровождающихся склерозом кожи / В.А. Волнухин, В.А. Самсонов // Вестн. дерматол. и венерол. - 2013. - № 5. - С. 50-68. 3. Каламкарян А.А. Клиническая дерматология: редкие и атипичные дерматозы / А.А. Каламкарян, В.В. Мордовцев, Л.Я. Трофимова. - Ереван "Айастан", 1989 - 569 с. 4. Склередема взрослых Бушке / В.Ф. Оркин, А. И. Завьялов, Н.В. Богомолова, В.М. Марченко [и др.] // Росс. журн. кож. и вен. бол. - 2005. - №6. - С. 14-16. 5. Туряница С.Р. Случай диагностики склередемы Бушке - редкого склеродермоподобного заболевания / С.Р. Туряница, И.Ю. Головач, К.И. Гранчак // Укр. ревматол. журн. - 2011. - №1(43). - С. 96-99. 6. Шуваева Ф.К. Сочетание диффузно-узлового токсического зоба и склередермы Бушке / Ф.К. Шуваева, Е.И. Ворожцова, А.Ф. Вербовой // Проблемы эндокрин. - 2011. - №6. - С. 27-29. 7. Dumke A.K. Scleredema adultorum Buschke and exulcerated necrobiosis lipoidica in diabetes mellitus type 1 / A.K. Dumke. J. Tittelbach, P. Elsner // Dtsch. Med. Wochensch. - 2014. - Vol.139, №43. - P. 2184-2187. 8. Extensive scleredema adultorum with loss of eccrine glands / I.C. Lin, H.Y. Chiu, J.Y. Chan, S.J. Lin / / J. Am. Acad. Dermatol. - 2014. - Vol. 71, №3. - P. 99-101. 9. Juvenile scleredema of Buschke / J.D. Rani, S.G. Patil, S.T. Murthy, A.V. Koshy [et al.] // J. Contemp. Dent. pract. - 2012. - Vol.13, №1. - P.111-114. 10. Scleredema adultorum of Buschke over an unusual site associated with pregnancy / C.S. Vinod, H. Ambika, N. Reddy, De. J. Kumar // Indian. Dermatol. Online J.- 2014. - Vol. 5, №4. - P. 466-468. 11. Scleredema of Buschke associated with diabetes mellitus type 2: case report and review of the literature / S.L. Dinato, G.L. Costa, M.C. Dinato, A. Sementilli [et al.] // Arq. Bras. Endocrinol. Metabol. - 2010. - Vol. 54, №9. - P. 852-855. 12. Successful treatment of poststreptococcal scleredema adultorum Buschke with intravenous immunoglobulins / M.C. Aichelburg, R. Loewe, N. Schicher, P.G. Sator [et al.] // Arch. Dermatol. - 2012. - Vol.148, №10. - P. 1126-1128.

РЕДКИЙ КЛИНИЧЕСКИЙ СЛУЧАЙ - СКЛЕРЕДЕМА БУШКЕ

Л.О. Гулей, Л.В. Мудряк

Резюме. В статье описан клинический случай редкого заболевания кожы - склередемы Бушке. Проведено дифференциальную диагностику с другими склероподобными заболеваниями.

Ключовые слова: склередема Бушке, клинический

случай, инфильтрация.

РІДКІСНИЙ КЛІНІЧНИЙ ВИПАДОК - СКЛЕРЕДЕМА БУШКЕ

Л.О. Гулей, Л.В. Мудряк

Резюме. У статті описано клінічний випадок рідкісного захворювання шкіри - склередеми Бушке. Проведено диференційну діагностику з іншими склероподібними захворюваннями.

Ключові слова: склередема Бушке, клінічний випадок, інфільтрація.

Вищий державний навчальний заклад України "Буковинський державний медичний університет", м. Чернівці; Чернівецький обласний шкірно-венерологічний диспансер

Clin. and experim. pathol.- 2015.- Vol.14, №2 (52).-P.296-299.

Надійшла до редакції 20.05.2015
Рецензент – проф. О.І. Денисенко