Case report

LUPUS VASCULITIS

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SUMMARY:
Combination between Leucocytoclastic vasculitis and Subacute cutaneous lupus erythematosus has been only rarely reported in the literature. Pathological cutaneous lesions are presented by purpura, erythematous macules, urticarial, nodules and necroses. We present a 58-year-old woman with painful nodular lesions on the left thigh. The disease started in June 2015 with photosensitivity and psoriasiform rashes on the back. Pathological cutaneous changes affected lateral and dorsal surface of the left thigh. It was presented by painful, indurated in base, ulcerative plaques, with unclear borders and necrotic surface. There were no deviations from complete blood count and biochemistry. Immunological investigations revealed elevated levels of MPO(pANCA) – 1.38 U/ml, antinuclear antibodies (ANA)- 134.1 U/ml, SS-A(Ro) antibodies – 233.7 U/ml CRP- 12.7mg/l. Urine analysis revealed positive protein results. Escherichia coli was isolated from microbiology testing. Histopathological result from ulcerative lesions from the left thigh of the patient revealed necrotizing leukocytoclastic vasculitis. The diagnosis lupus vasculitis was made. Systemic therapy with chloroquine phosphate, methylprednisolone, methotrexate was administered. Topical treatment included proteolytic enzyme in dressings and vacuum therapy with good effect.

Keywords: Lupus vasculitis, antinuclear antibodies (ANA), SS-A(Ro) antibodies, vacuum therapy, chloroquine phosphate, methylprednisolone, methotrexate.

INTRODUCTION:
Leukocytoclastic vasculitis associated with subacute cutaneous lupus erythematosus (SCLE) has been described for the first time by Sontheimer. Purpura, erythema, urticaria, nodules and necrosis affect the skin of the trunk, palmoplantar areas and lower limbs. The histopathological changes are presented by leukocytoclastic vasculitis of the small blood vessels in the deep dermis. The prognosis of the disease is good [1].
necrosis and fragmented neutrophilic nuclei (nuclear “dust”) - histological pattern of necrotizing leukocytoclastic vasculitis (fig. 2a, b, c).

Fig. 1. a, b. Ulcerative plaques with undermined borders and necrotic surface on the dorsal (a) and dorsal-medial surface (b) of the left thigh.

Fig. 2. a, b, c. Histological pattern of necrotizing leukocytoclastic vasculitis - fibrinoid necrosis and fragmented neutrophilic nuclei (nuclear “dust”) with inflammation of the small-sized vessels in deep dermis. (a) H&Ex100, (b) H&Ex25, (c) H&Ex40

Chest X-ray was without deviations. Ultrasound of the kidneys established hydronephrosis IV grade. After consultation with cardiologist and echocardiography 10mm pericardial effusion and concentric left ventricular hypertrophy were found. Based on the data from the medical history, clinical picture results from the laboratory investigations and the consultations, the diagnosis lupus vasculitis was made. Systemic treatment with chloroquine phosphate tabl. 250mg 3 times daily, methylprednisolone in tapering doses from 60 mg to 20 mg daily and methotrexate 15 mg. weekly p.o. was administrated for one month. Maintenance therapy with chloroquine phosphate 250mg daily and Methotrexate 15 mg. weekly was conducted. Two-months vacuum therapy course was performed, and the result was complete epithelization of the lesions with atrophic scars (fig. 3a, b).
DISCUSSION:
SCLE was first described by Sontheimer et al. in 1979 [2]. Gilliam and Sontheimer created a classification that divided Lupus erythematosus into specific and non-specific forms. Specific forms include chronic, subacute and acute cutaneous lupus erythematosus. The non-specific forms comprise leukocytoclastic vasculitis, livedo race- 
omos, thrombophlebitis, occlusive vasculopathy, Ray- 
naud's syndrome, periangual telangiectasia, diffuse non- 
cicatrical alopecia, erythema multiforme, calcinosis cutis and papular mucinosis [3].

The combination of SCLE with histological vascular changes is rarely observed [1]. The most common non-specific findings affect mainly small-sized blood vessels in the dermis. The histopathological picture corresponds to leukocytoclastic vasculitis and is presented by perivascular neutrophilic infiltration, edema and fibrinoid necrosis in the walls of the vessels. The same changes have been described in the patient observed by us. According to the immunological studies elevated ANA values have been reported in the majority of patients with SCLE. There are also increased anti-Ro antibodies and elevated levels of anti-La antibodies [5, 6]. The immunological results of our patient support the diagnosis SCLE. Purpura, erythema, urti- caria, nodules and necrosis on the trunk, palmpoplantar ar- eas and lower extremities have been described in patients with SCLE in combination with vasculitis. This clinical picture was observed in the case described by us.

Vascular lesions may precede SCLE, but more common- 
vously vasculitis develops in patients with prolonged SCLE [7]. According to a histopathological study by Cribier et al. that the degree of vascular necrosis and the depth of vascular changes were not severe in patients with systemic complications such as glomerulonephritis and gastrointe- 

ginal involvement [8].

The systemic antimalarial drugs such as hydro- 
xychloroquine, chloroquine and mepakine are the most commonly used. Hydroxychloroquine is best tolerated, and 
therefore it is administered as first-line medication in the treatment of SCLE. Its therapeutic effect could be seen 6 weeks after the first dose [9].

Systemic corticosteroids and immunosuppressive agents are also used in patients suffering from SCLE. Aza-
thioprine and cyclophosphamide have been reported as ef- 
effective in the treatment of SCLE in small groups of patients. Thalidomide has been used in severe course of the disease. Improvement is fast, although there are frequent recidives. [10,11]. Vacuum therapy was reported as an effective method for treatment of refractory for systemic therapy ulcers [12].

CONCLUSION:
We presented a clinical case of 58-year-old woman diagnosed with lupus vasculitis, treated with chloroquine phosphate, methotrexate and vacuum therapy, with good result. There was complete epithelization of the ulcerative lesions with atrophic scars without new vascular changes. The patient is monitored regularly for systemic manifestations of lupus erythematosus and is under observation by cardiologist, rheumatologist and ophthalmologist. There are no side effects from the systemic treatment so far.

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