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Clinical and histological correlation in the diagnosis of lipodermatosclerosis

Povezanost kliničke slike i histološkog nalaza u postavljanju dijagnoze lipodermatoskleroze

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Summary

Introduction: Lipodermatosclerosis is a chronic fibrosing panniculitis associated with venous insufficiency. Clinical manifestation refers to the induration and hyperpigmentation of the skin on one or both legs. The characteristic feature of chronic lipodermatosclerosis is a pseudomembranous type of fat necrosis, known by most pathologists. Case report: A female patient was admitted for a diagnostic procedure and treatment of sideropenic anemia. The patient complained about purple subcutaneous painful nodules on the skin in the lower limbs. The pathohistological finding of skin biopsy indicated lipodermatosclerosis. Our patient didn't accept any suggested therapy. Conclusion: When the diagnosis of lipodermatosclerosis is suspected, it is essential to do a biopsy of the affected skin and a pathohistological analysis, to support the diagnosis, while it can be confused with other panniculitides.

Key words: lipodermatosclerosis, fat necrosis, panniculitis, venous insufficiency

Sažetak

Uvod: Lipodermatoskleroza je kronični fibrozirajući panikulitis povezan s venskom insuficijencijom. Kliničke manifestacije odnose se na zadebljanje i hiperpigmentaciju kože jedne ili obje noge. Karakteristična značajka kronične lipodermatoskleroze je pseudomembranozni tip nekroze masti, poznat većini patologa. Prikaz slučaja: Bolesnica je primljena radi obrade i liječenja sideropenične anemije. Kao glavne tegobe navodila je ljubičaste bolne potkožne nodule prisutne na potkoljenicama. Patohistološki nalaz bioptata kože upućivao je na lipodermatosklerozu. Bolesnica nije pristala na preporučenu terapiju. Zaključak: Pri kliničkoj sumnji na dijagnozu lipodermatoskleroze, bitno je učiniti biopsiju kože i patohistološku analizu bioptata, kako bi potvrdili dijagnozu, te isključili druge moguće uzroke.

Ključne riječi: lipodermatoskleroza, masna nekroza, panikulitis, venska insuficijencija

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Introduction

Lipodermatosclerosis is a chronic fibrosing panniculitis associated with venous insufficiency.[1] The induration and hyperpigmentation of the skin on one or both legs are clinical manifestations. International Consensus Committee on chronic venous disease proposed a classification, in which the clinical part of the classification grades patients in six classes, from those with no visible evidence of venous disease (class I), followed by those with skin changes including lipodermatosclerosis (LDS) (class IV), to those with active venous ulceration (class VI). Manifestations in the acute stage of LDS are extremely painful, red to purple, poorly demarcated, indurated plaques, mostly in the part of the leg above the medial malleolus. The chronic phase of LDS is characterized by sharp demarcation of the induration, and may be present with

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pain, which tends to be more dull and aching.[2] One study found pain in the lower extremities the most common symptom of LDS.[3]

Due to many possible differential diagnoses and common misdiagnoses, it is suggested to exclude the diagnosis of LDS in the case of indurated, well demarcated cellulitis.

When the diagnosis of LDS is suspected, it is essential to do a biopsy of the affected skin and pathohistological analysis, to support the diagnosis, because it can be confused with other panniculitides.[4] The scleroderma-like hardening of the skin in LDS is characterized by fibrous scar tissue of the reticular dermis, which is a build-up of collagen bundles and degraded elastic fibers.[5] A characteristic feature of chronic LDS is a pseudomembranous type of fat necrosis, known by most pathologists. The histopathologic features of early and evolving LDS are less well documented.[6]

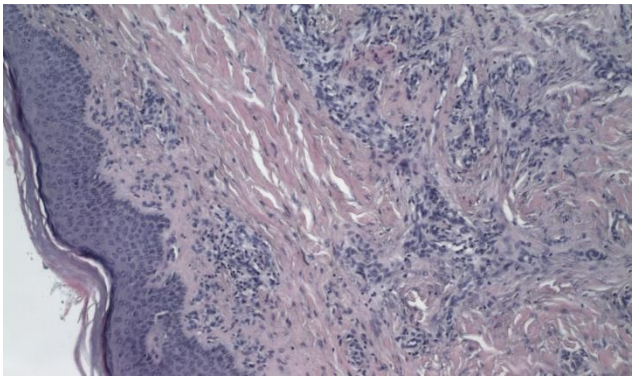
Case report

A 44-year-old female was admitted to the Department of Internal Medicine in General Hospital "Dr. Josip Benčević", for a diagnostic procedure and treatment of sideropenic anemia. The patient was transferred from the Department of Psychiatry where she was treated for schizophrenia. A year earlier she had been diagnosed with anemia, but she had not been taking her recommended therapy.

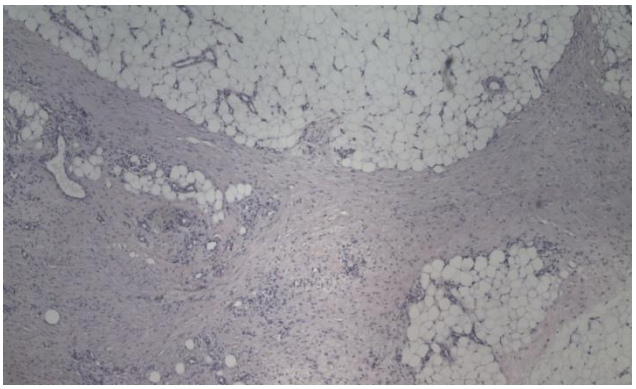
The patient complained about her legs feeling heavy, particularly in the evening, and she couldn't distinguish which part was more painful – muscles, bones or joints. Firstly, she noticed purple subcutaneous painful nodules of the skin in the lower limbs.

In the clinical status, the skin on both legs was solid with brown rash and subcutaneous nodules in the proximal parts of her lower legs. On the medial side of the left leg the skin was light red, pulsations of the peripheral arteries were palpable. We considered differential diagnoses such as dermatomyositis and eosinophilic fasciitis. Therefore, additional laboratory testing was performed: creatine kinase, immunological diagnostics and consulting with a dermatologist, based on which we excluded those diagnoses. Our conclusion was that the etiology of anemia is an inadequate diet, since the patient did not eat meat. Endoscopic treatment of the gastrointestinal tract was not done due to the patient's request. We planned to do a magnetic resonance of the legs, but the patient could not withstand the test.

The dermatologist found it to be prurigo, and therefore recommended skin biopsy. A skin biopsy of both legs was taken, but pathohistological finding came after the patient's dismissal. The pathologists suggested that we should think about a differential diagnosis as lipodermatosclerosis, the late phase of septal panniculitis or erythema nodosum. Panniculitis and erythema nodosum were excluded by previous diagnostics. The pathohistological finding described proliferation of capillaries coated with endothelium in underlying papillary dermis, interrupted by swelling and the proliferation of fibroblasts in the surrounding dermis (Picture 1). In the deeper part of the reticular dermis edema was visible, while most prominent changes in fat tissue were fibrous interlobular septas filled with mononuclear inflammatory infiltrate (Picture 2). There were no signs of membranocystic changes nor hemosideric deposits.



Picture 1: On the surface of the skin sample is a regular epidermis with a thin layer of ortho-keratinotic type, and dermis proliferation of the capillaries and fibroblasts. (HEx100)
Slika 1. Površina primjerka kože prikazuje redovni epidermis s tankim slojem ortokeratinotinskog tipa te proliferacije kapilara i fibroblasta u dermisu



Picture 2: In the subcutaneous fatty tissue, the fibrosis of the interlobular septas is observed, within which is an infrequent mononuclear inflammatory infiltrate, adipocytes of different sizes in some lobules.
Slika 2. U subkutanom masnom tkivu primjećuje se fibroza interlobularnih septa u kojima se nalazi rijedak mononuklearni upalni infiltrat

Discussion

In searching for available literature, we found that LDS diagnosis is uncommon and still requires clinical and histological correlation, as demonstrated in this case. The disadvantage of our case view was the lack of photos, as the patient did not give permission. Another disadvantage was that we did not have the opportunity to try the therapy because the patient did not come in for a check up.

The constellation of findings including septal fibrosis, necrosis of lipid membranes, prominent vascular changes of stasis, and erythrocytic extravasation can be used to define LDS as histopathological diagnosis. Iron deposition in the subcutaneous tissue is a useful finding for this chronic condition.[4] LDS is most commonly diagnosed in middle aged women and it also may occur as an acute form referring to the pain present as a symptom, often misdiagnosed as cellulitis or erythema nodosum. Bruce et al,[3] in the retrospective study of 97 patients presented to the Mayo Clinic with a diagnosis of LDS, described that LDS is connected with high body mass index. In this case, the difference is that our patient was underweight (body weight 45 kg, height 162 cm, body mass index 17 kg/m^2) and did not have venous insufficiency of the legs.

Demitsu et al[7] reported three cases of LDS. The first case demonstrated the histologic features of lipogranuloma with mild septal fibrosis. The second case showed pronounced fibrotic changes in the deep dermis as well as lobular panniculitis with membranocystic fat necrosis, and in the third case both lobular panniculitis with fat necrosis and fibrosis were prominent. The authors suggested oral administration of tocopherol nicotine combination with topical steroid as a first choice of standard therapy for LDS.[7]

Walsh et al [8] prospectively made a study of 25 cases and clinically described that the lesions were erythematous, indurated plaques or nodules. In the subcutaneous tissue characteristic histologic findings were seen, primarily involving the lobules. Changes in the fat tissue included: micro and macro pseudocystic formation, necrosis of adipocytes, nodules of lipoid material associated with granulomatous inflammation and macrophages filled with fatty material. There was no neutrophil or lymphocyte inflammation in these lesions.[8]

Conclusion

According to our opinion, this patient's illness was found during the transition phase from subacute to the chronic one. In fact, the subcutaneous nodes were the symptoms of the acute phase. We concluded, as well as the previously mentioned authors, that a clinical and histological correlation was required to establish a correct diagnosis.

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