

SUBCUTANEOUS PANNICULITIS- LIKE T-CELL LYMPHOMA: REPORT OF TWO CASES

by Jyoti Ramnath Kini, Ancy Susan John, Hema Kini, Flora Dorothy Lobo, Pramod Kumar, Krishna Prasad

comment:

Ass. Prof. Małgorzata Sokołowska-Wojdyło

Department of Dermatology, Venereology and Allergology,

Medical University of Gdansk,

Gdansk, Poland

E-mail: mwojd@gumed.edu.pl

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Subcutaneous panniculitis-like T-cell lymphoma (SPTCL), originally described as lymphoma of the cytotoxic T lymphocyte, is characterized by a tendency to infiltrate the subcutaneous tissue. The rapid clinical course and aggressive multidrug chemotherapy was the treatment of choice by many years – but it has been changed since 2008. SPTCL term is used only in relation to disease with TCR $\alpha\beta$ phenotype, while TCR $\gamma\delta$ + panniculitis-like T-cell lymphomas have become classified by WHO and EORTC as primary cutaneous $\gamma\delta$ T-cell lymphoma - PCGD-TCL. The course and prognosis of those two entities differs significantly. SPTCL occurs in children and adults. The major symptoms include single or multiple nodules or deep-seated infiltrates localized mainly in the skin and subcutaneous tissue of the lower limbs, arms and trunk. The ulcerations are rare (6% of cases – versus 45% in PCGD-TCL) [7]. The general symptoms as fever, fatigue and weight loss and laboratory abnormalities (eg. cytopenia and increased liver enzymes), occur frequently, but full-blown hemophagocytic syndrome (HPS) occurs in only 15% patients [1]. The involvement of other organs beyond the skin is rare – contrary to PCGD-TCL, where extranodal involvement, as well as mucosal lesions occurs very often [2]. SPTCL has a very good prognosis if it is not accompanied by HPS. Recent reports indicate that the proportion of surviving 5 years is 91% if the disease is not accompanied by HPS and 46% when it is associated with HPS [1]. That is why, the 1st sentence of the article “ Subcutaneous panniculitis-like T-cell lymphoma (SPTCL) originally described by Gonzalez et al is a primary cutaneous lymphoma (PCL) characterized by an aggressive clinical course ... [3]“ is not true anymore. It is very important to remember that the publications written before the last classification had collected the SPTCL and PCGD-TCL into one group– that is why chemotherapy was the first choice before 2008 in many clinical centers. The recommended treatment in SPTCL without HPS nowadays involves the use of systemic corticosteroids and other immunosuppressive agents, while in the case of isolated lesions the radiotherapy can be the first choice. The chemotherapy,

especially multidrug one, as well as bone marrow transplantation, should be introduced only in case of patients with progression of the disease, resistance to recommended treatment and/ or SPTCL with accompanying HPS. It is well to remember that in addition to HPS, elevated lactate dehydrogenase enzyme level and low white blood cell count are known poor prognosis factors [2]. But even in those cases, as well as in cases with relapses after chemotherapy – the immunosuppressive therapy can be considered - ex. with cyclosporine A – with long lasting follow up [2,4]. Especially that the risk of infection is still lower in case of systemic steroids and / or cyclosporine A than in cases of multidrug chemotherapy. But anyway – the described cases can be interesting because they can remind the difficulties in differential diagnosis of the disease. Diagnosis of SPTCL is based on pathological examination of skin and subcutaneous tissue, immunohistochemical staining patterns, molecular analysis, and clinical characteristics. The differentiation of Lupus erythematosus profundus (LEP) and SPTCL seems to be the biggest diagnostic challenge. When the vasculitis, mucin deposition, reactive germinal centers, B-cells clusters, considerable number of admixed plasma cells and polyclonal TCR-gene rearrangements are typical for LEP, the atypical cellular infiltrates of the subcutaneous fat (both lobules and septae), mimicking panniculitis are also typical for SPTCL [5]. Lymphocytes exhibit slight atypical features, including hyperchromatic, angulated nuclei, and indistinct cell borders in SPTCL. Scattered mitoses, apoptotic cells, karyorrhectic debris, focal areas of fat necrosis, and rimming of individual fat cells by neoplastic cells are also common in SPTCL [6]. Immunophenotyping can be very helpful. The neoplastic cells in SPTCL are cytotoxic T cells CD3+ CD4-. TCR $\alpha\beta$ cells are CD8+ and usually CD30- CD56- whereas PCGD-TCL is usually CD8-CD30+CD56+. In terms of differential diagnosis, benign panniculitis usually has aggregates of CD20- B-cells mixed with CD3- cells that are both CD4- and CD8-. LEP is commonly CD4+ without CD8+ T cells [6].

Although this is still controversial, some authors suggest that patients with LEP are at risk for the development of abnormal, clonal T-cell proliferations and/or overt SPTCL. In cases of atypical lymphocytic lobular panniculitis that fail to meet diagnostic criteria for subcutaneous panniculitis-like T-cell lymphoma, patients should be clinically followed indefinitely, as future subcutaneous lymphoma cannot be excluded [7].

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