

Case presentation

An 86-year-old lady with a personal history of a mamma carcinoma (2008) presented with waxing and waning nodules, which heal leaving an atrophic scar. There were no B-symptoms, lymphadenopathy, hepatosplenomegaly or changes in medication. She displayed multiple indurated subcutaneous nodules, as well as lipoatrophy on both arms, back and sternum. Biopsy showed lobular infiltrating CD8+T-cells in the subcutis, with atypia and karyorrhexis. The rimming of individual lipocytes gave the diagnosis of subcutaneous panniculitis-like T-cell lymphoma away. Further staging consisted of lab tests (lymphopenia and low-grade inflammation), bone marrow biopsy (normal) and PET-CT (hypermetabolism in the right breast, histologically compatible with lymphoma localization). After multidisciplinary discussion, treatment with methotrexate was initiated.



Discussion

SPTCL is a rare primary cutaneous lymphoma that mimics panniculitis and originates from cytotoxic T-cells. Patients present with subcutaneous plaques/nodules in different stages. Ulceration is infrequent.¹ B-symptoms and laboratory abnormalities are reported in approximately 50%.^{1,2} Most cases show an indolent disease course. However, 15-20% of patients develop a hemophagocytic syndrome.^{1,2} Vigilance for cytopenias, elevated liver enzymes and hepatosplenomegaly is warranted¹, but the golden standard for diagnosis of this syndrome is bone marrow cytology.² To evaluate disease extent a PET-CT is typically performed. Histologically, SPTCL is characterized by atypical T-cells (alpha/beta subset) in the subcutis with sparing of interlobular regions, epidermis or dermis. There is rimming of individual lipocytes, as well as a variable degree of karyorrhexis and fat necrosis.^{1,2} Cytotoxic T-cell markers are expressed.³ Immunohistochemical staining shows a high Ki67 index, in contrast to lupus panniculitis which forms a major differential diagnosis.¹ Other differential diagnoses consist of nasal type extranodal natural killer/T-cell lymphoma (EBV+), morphea or SPTCL (gamma/delta subset). Treatment guidelines are lacking. According to literature, patients are treated with systemic corticosteroids, retinoids or immunosuppressants. Currently, chemotherapy (doxorubicin based) and autologous stem cell transplantation are for progressive disease or second-line treatment. Radiotherapy can be used for palliative purposes and localized disease.^{1,2}



Figures

Figure 1-2 (left), on presentation: infiltrated erythematous subcutaneous plaques on both arms

Figures 3-7 (right), after 4 months methotrexate: non-inflammatory depressed areas of lipoatrophy on arms, sternum and back

Conclusion

SPTCL is a rare form of cytotoxic T-cell lymphoma which usually follows an indolent course. Histology assists in diagnosis and shows lobular T-cell infiltrate preferentially in the subcutis, more specifically surrounding the lipocytes. There is no consensus about first-line treatment being chemotherapy, corticosteroids or immunosuppressants.

References

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