

Sezary syndrome initially presenting as psoriasis vulgaris, partial response after 6 cycles of gemcitabine and radiotherapy: A rare case report

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ABSTRACT

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Sezary syndrome (SS) is a variant of *Cutaneous T-Cell Lymphoma*, a rare type of lymphoma in adults. The incidence of SS is estimated epidemiologically at around 2-3% of all primary cutaneous lymphomas. SS has not been reported in Mohammad Hoesin General Hospital Palembang. Here, we report one such case. A 45-year-old female presented with generalized pruritus for 2 years. She had a history of psoriasis, previously treated with secukinumab, methotrexate, and methylprednisolone for 1 year. Physical examination showed a painful lymphadenopathy and hyperpigmentation macules on the skin. Skin biopsy revealed lymphoproliferative lesions. PET-Scan showed multiple lymphadenopathies. Immunohistochemistry (IHC) examination revealed CD3, CD4 positive diffuse, with K167 positive in 20% of tumor cells, supporting a T-cell lymphoproliferative lesion. Examination of peripheral blood images found large atypical lymphocytes (3/100 cells) and indented nuclei with basophilia, suggestive of lymphoproliferative disease suspicious for SS. Leukemia phenotyping showed large atypical lymphocytes with indented nuclei suggestive of Sezary cells, a CD4+: CD8+ ratio: 25: 1, and CD4+/CD7 ratio of 70.41% of total T lymphocytes, consistent with SS markers. SS can mimic several mild dermatoses such as psoriasis vulgaris, or initially present as psoriasis vulgaris and later progress to erythrodermic skin lesions due to plaque extension. The triad of SS consists of pruritic erythroderma, lymphadenopathy, and Sezary cells in the blood circulation. This patient fulfilled all three criteria of SS, with severe skin manifestations and multiple systemic symptoms, lymphadenopathy, and large atypical lymphocytes in the peripheral blood smear (3/100 cells), CD4+: CD8+ ratio: 25: 1 and CD4+/CD7 ratio of 70.41% consistent with SS markers. The therapy administered was gemcitabine chemotherapy 1000 mg/mm² (days 1, 8, 15) and radiotherapy. The patient achieved a partial response after 6 cycles of gemcitabine and radiotherapy.

ABSTRAK

Sindrom Sezary (SS) adalah varian dari limfoma sel T pada kulit, termasuk jenis limfoma yang jarang terjadi pada orang dewasa. Insiden SS secara epidemiologis diperkirakan sekitar 2-3% dari semua limfoma primer kulit. Belum ada laporan SS di RS Mohammad Hoesin Palembang. Berikut kami laporkan 1 kasus SS. Seorang wanita berusia 45 tahun datang dengan gatal seluruh tubuh selama 2 tahun. Riwayat psoriasis, sebelumnya diobati dengan secukinumab, metotreksat, dan metilprednisolon selama 1 tahun. Pemeriksaan fisik menunjukkan limfadenopati yang nyeri dan makula hiperpigmentasi pada kulit. Pada pemeriksaan biopsi kulit, diperoleh kesan lesi limfoproliferatif. PET-Scan menunjukkan limfadenopati multipel. Pemeriksaan IHC, CD3, CD4 positif difus dengan K167 positif pada 20% sel tumor, mendukung lesi limfoproliferatif sel T. Pemeriksaan darah tepi menunjukkan limfosit atipikal besar (3/100 sel) dan inti berlekuk dengan basofilia, menunjukkan penyakit limfoproliferatif yang diduga SS. Pemeriksaan fenotipe leukemia menunjukkan sel limfosit atipikal besar dengan inti berlekuk yang menunjukkan sel Sezary, rasio CD4+:CD8+: 25:1

Keywords:

Sezary syndrome;
Sezary cell;
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gemcitabine;
radiotherapy

dan rasio CD4+/CD7 terhadap total limfosit T sebesar 70,41%, sesuai marker SS. SS dapat menyerupai beberapa penyakit kulit ringan seperti psoriasis vulgaris atau awalnya bermanifestasi sebagai psoriasis vulgaris dan berkembang menjadi lesi kulit eritroderma yang luas. Trias SS adalah eritroderma pruritus, pembesaran kelenjar getah bening, dan sel Sezary dalam sirkulasi darah. Pasien ini memenuhi ketiga kriteria SS dengan manifestasi kulit berat dan banyak gejala sistemik lainnya, limfadenopati, dan gambaran darah tepi menunjukkan limfosit atipikal besar (3/100 sel), rasio CD4+:CD8+: 25:1, dan rasio CD4+/CD7 terhadap total limfosit T sebesar 70,41%, sesuai marker SS. Terapi yang diberikan adalah kemoterapi gemcitabine 1000 mg/mm² (hari ke-1, hari ke-8, hari ke-15) dan radioterapi. Pasien mencapai respons parsial setelah 6 siklus gemcitabine dan radioterapi.