

Title: New Frontier: A Complex Case of a 65 year-old Filipino Male with Sezary Syndrome

Learning Objective: This case report aims to encourage dermatologists to be vigilant in doing re-biopsies in questionable cases, order for complete surface markers including CD 3,7, and 30, and for dermatologists to help take the lead in the management of Sezary Syndrome.

INTRODUCTION: Sezary Syndrome is a rare cutaneous T-cell lymphoma that is very challenging to manage because of a general poor response to treatment. Herein is a report of a Sezary Syndrome patient who received CHOP and Methotrexate therapies who later on became unresponsive and suffered adverse events. Brentuximab vedotin, an anti-CD30 antibody drug conjugate, was given as an alternative treatment due to its mechanism of action and safety profile.

CASE REPORT: This is a case of a 65 year-old Filipino male living in Saudi Arabia who presented initially with scaly plaques. He was diagnosed with Psoriasis Vulgaris through biopsy and received several systemic treatments such as risankizumab but was only partially responsive with noted progression to persistent erythroderma, hence, a rebiopsy of the patient was done which pathology reported as atypical lymphocytes with CD3+, increased CD4 to CD7ratio, decreased CD7, and CD 30+. Bone marrow aspirate and peripheral blood smear showed 20-30% lymphocytic infiltration with cerebriform appearance which correlated with the histologic findings. This prompted the diagnosis of Sezary Syndrome. PET scan, Ultrasound and CT Scans of the chest and abdomen were taken which showed several lymph nodes enlargement. He was then referred to oncology where he received CHOP for 6 sessions and achieved remission. Methotrexate 20 mg weekly and folic acid 1 mg cap OD were maintained. The patient tolerated the treatment but then he relocated to the Philippines where he was evaluated by a team of dermatologist, oncologist, and nephrologist. Topical halobetasol and tacrolimus 0.1% were started in addition to his current treatment which was continued for several months until there was note of methotrexate toxicity such as GI bleeding and renal toxicity. The patient was CD30+ on histology, hence the dermatologist opted to shift to Brentuximab vedotin 150 mg IV infusion every 3 weeks. Currently, there is partial response seen as of the 4th cycle with normal creatinine levels and decreasing erythroderma however pruritus is still prominent. The plan is to complete 8 cycles and reassess if there is a need for 16 cycles. Side effects of brentuximab noted were body malaise, leukocytosis, and mild respiratory infection.

DISCUSSION:

This case highlights the importance of repeated skin punch biopsies to catch cutaneous lymphoma in a questionable clinical profile. Another learning point is the importance of collaboration between a dermatologist and other disciplines such as oncology and pathology, especially in the decision-making process. Finally, the use of Brentuximab vedotin in a Sezary Syndrome is quite novel in our local setting and it was decided due to the CD30+ on skin histology, making it important to request for complete surface markers for cutaneous lymphoma such as CD30 since this is not currently practiced.

