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CUTANEOUS T-CELL LYMPHOMATOID PAPULOSIS INVOLVED FACE AND NECK

S.S DURUSOY[1], M. SOKUCU [2] and M. YILMAZ[1]
1. Sanko University School of Medicine Department of Hematology, Gaziantep/TURKEY
2. Sanko University School of Medicine Department of Pathology, Gaziantep/TURKEY

SANKO UNIVERSITY HOSPITAL

INTRODUCTION

Primary cutaneous CD30+ lymphoproliferative disorders (LPDs) are the second most common group of CTCL, accounting for ~25% of all CTCLs. This group includes primary cutaneous anaplastic large lymphoma (C-ALCL) and lymphomatoid papulosis (LyP), which form a spectrum of disease(1). LyP is a non-aggressive skin disorder characterized by papulonodular injuries, sometimes necrotic, often scattered, relapsing, which frequently regress spontaneously. LyP represents about 12% of cutaneous lymphomas. The etiology of LyP is unknown. Based on its histopathology, in 2018, the World Health Organization (WHO) classified LyP into six types with similar prognosis (A,B,C,D,E and DUSP22). Cutaneous relapses are common, but extra cutaneous dissemination occurs in only 10% to 15% of patients. We present a case of primary cutaneous CD30+ T-cell lymphoma that involved in the head and neck region. To demonstrate that Brentuximab and CHP combination therapy works in CD30+ cutaneous T cell lymphoma(2).

CASE

A 82-year-old female patient presented with lesions involving face and neck. The patient did not have any additional disease or any medical treatment. Diffuse infiltrating papulonodular and papulonecrotic lesions in the head and neck region were observed. A 82-year-old female patient presented with lesions involving face and neck. The patient did not have any additional disease or any medical treatment.

The biopsy taken from the lesioned area in front of the ear was reported as T-cell lymphoma. (CD45+, CD30+, CD5+, CD4+, MUM-1+, CD20-, CD38-, CD15-, CD56-, CD8-, ALK-1-,CD10-, PAX5-, CD3-). The diagnosis of 'primary cutaneous CD30+ lymphoproliferative disease lymphomatoid papulosis type was made. The patient treated with 4 courses of systemic B+CHP (brentuximab 1.8 mg/kg, cyclophosphamide 750 mg/m2, doxorubicin 25 mg/m2, prednisolone 100 mg, 5 days). Response evaluation with PET-CT was planned for the patient after 4 cycles. No side effects were observed in the patient during 4 cycles of treatment.

CONCLUSIONS

Primary cutaneous T-cell lymphomas are a heterogeneous group and may show an aggressive course. Forty-eight evaluable patients received an infusion of Brentixumab 1.8 mg/kg every 21 days (22 women and 26 men; median age, 59.5 years) had an overall response rate of 73% and complete response rate of 35%. All patients with lymphomatoid papulosis (n = 9) and primary cutaneous anaplastic T-cell lymphomas (n = 2) responded (3).

A total of 12 patients with LyP received brentuximab vedotin Intravenous 1.8 mg/kg infused over 30 minutes every 21 days. All patients were 18 years or older with a diagnosis of LyP and were also required to have scarring, more than 10 lesions, or active lesions on the face, hands, or feet. All 12 patients (8 men and 4 women; median age, 46 years) responded to brentuximab vedotin, and 7 exhibited a complete response. Time to response was 3 weeks in all patients. The median duration of response was 20 weeks (range, 6-103 weeks) (4).

Although no approved treatment at the present time, to date, there are four main and widely practiced therapeutic approaches in lymphomatoid papulosis: phototherapy, topical steroid use, low dose methotrexate, and an alternative therapeutic route including antibiotics, antihistamines, or oral steroids (5). All these therapeutic approaches can be classified as first-line treatment after standard patient observation. Brentixumab vedotin combination with CHP ameliorated lesions of lymphomatoid papulosis in our patient. The complete response of patient is continuing for 1 year. Accordingly, Brentixumab Vedotin with CHP should be considered as an alternative treatment option of these patients.



Diffuse infiltrating papulonodular and papulonecrotic lesions in the head and neck



The lesions regressed almost completely, leaving a hyper pigmented plaque in place

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CONTACT INFORMATION

myilmaz@sanko.edu.tr