

Aggressive Progression of Mantle Cell Lymphoma: A Case Report

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Abstract Mantle cell lymphoma is a relatively rare disease, mostly affecting the elderly. It behaves aggressively although it has features of an indolent lymphoma. Here, we presented a case of an 85-year old female patient applying to emergency service with difficulty in breathing due to huge lumps around her neck. She had mantle cell lymphoma which has aggressively progressed despite chemotherapy. She was at an advanced stage of the disease and taken to the intensive care unit. Despite best supportive care, steroid therapy and respiratory support, she passed away within 24 hours.

Keywords: mantle cell lymphoma, stem cell transplantation, MIPI

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1. Introduction

Mantle cell lymphoma (MCL) is a relatively rare disease accounting for approximately 3% to 6% of all non-Hodgkin lymphoma (NHL) cases [1]. It's mostly a disease of the elderly who usually present with widespread involvement and show poor response to therapy. Although it has features of an indolent NHL, clinically it behaves as an aggressive NHL and even with the most aggressive therapies, a short remission duration is usually observed and the median overall survival is about 4-5 years [2]. A chromosomal translocation t(11:14) is the molecular hallmark of the disease, resulting in the overexpression of cyclin D1 which is detected by immunohistochemistry in 98% of cases. The absence of SOX-11 or a low Ki-67 may correlate with a more indolent form. The mantle cell lymphoma international prognostic index (MIPI) is a prognostic model including ECOG performance status, age, leukocyte count, and lactic dehydrogenase. A modification of the MIPI also adds the Ki-67 proliferative index if available [3]. For selected indolent, low MIPI MCL patients, initial observation may be appropriate therapy. For patients younger than 65 years with intermediate or high risk MIPI MCL, aggressive therapy with stem cell transplantation after R-CHOP treatment or R-Hyper-CVAD should be considered as first line treatment. For MCL patients older than 65 years with intermediate or high risk MIPI, conventional combination chemotherapy with R-CHOP should be considered as first line treatment and stem cell transplantation should be considered for patients with good performance status. At the time of relapse, allo-transplant following R-FC should be considered for younger patients, where autologous stem

cell transplantation following R-FC or R-Bendamustin should be considered at older patients. At further relapse, molecular approaches such as bortezomib, lenalidomide should be considered [4].



Figure 1. Aggressive head and neck involvement of mantle cell lymphoma

Case Presentation: Our patient was a 85 year-old woman who was brought to emergency service by her relatives because of her poor condition, besides huge lumps all over her body, some causing difficulty in breathing. According to followup notes received from her son, she had been diagnosed as mantle cell lymphoma 9 months ago and received 6 courses of R-CHOP (rituximab 375 mg/m², cyclophosphamide 750 mg/m², vincristin 1.4 mg/m², prednisolone 100 mg/day for 5 days) therapy as first line treatment at another centre (Figure 1). After the end of the therapy, her doctors evaluated her as having stable disease with partial remission and asked her to come to followup visits monthly. The last course of chemotherapy had been given 3 months ago. At physical examination, there were remarkable lumps on her face and huge lymph nodes surrounding her neck causing difficulty in breathing, the liver could not be palpated but the spleen could be palpated at 6 cm below the left costal margin, there were huge lymph nodes in servical, supraclavicular, axillary, pectoral, inguinal and even popliteal regions, some reaching a diameter of 10 cm. Lung sounds were diminished and she had prolonged expirium. She had a very poor performance status, and she was given best supportive care. In order to downsize the lumps around her neck and relieve respiratory distress, she was given prednisolone 100 mg/day. She couldn't be further evaluated radiologically due to her poor performance status. Twelve hours after she arrived, she was moved to the intensive care unit, entubated due to asphyxiation and passed away the next day.

2. Discussion

MCL usually has features of an indolent NHL, but clinically it behaves as an aggressive NHL. Most patients with mantle cell lymphoma present at an advanced stage, with generalized lymphadenopathy, and frequent involvement of extranodal sites and sometimes even

peripheral blood. The clinical progression is sometimes evolved to a relatively aggressive manner with a poor response to conventional therapeutic regimens, where frequent relapses may occur and a median overall survival of 4-5 years may be observed. This biological behavior has led to the recommendation for early treatment with intensive therapeutic regimens that may even include hematopoietic stem cell transplantation [5]. According to Williams [6], eligible patients with newly diagnosed MCL requiring treatment should initially be considered for a rituximab + a high-dose cytarabine-containing regimen, followed by autologous stem cell transplantation consolidation with high dose chemotherapy after first remission. Therefore, we recommend hematologic stem cell transplantation at eligible patients with mantle cell lymphoma at first remission as it may evolve into an aggressive disease.

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