

华氏巨球蛋白血症32例临床特征及预后分析

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Title: Clinical characteristics and prognosis analysis of 32 patients with Waldenström macroglobulinemia

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摘要: 目的: 评价华氏巨球蛋白血症患者的临床病理特征、治疗及预后情况。方法: 回顾性分析本院 2001年1月1日至 2018年5月30日收治的32例华氏巨球蛋白血症患者临床资料, 总结其临床病理特点、治疗方案及转归, 并探讨影响预后的因素。结果: 32例患者中, 男25例, 女7例, 中位年龄为64岁, 低、中及高危组患者分别占3.1%、43.8%及 53.1%。90.6%以贫血症状为首发表现。中位血红蛋白水平为75.5(26~146) g/L, 中位IgM水平为52.4(13~110) g/L, 其中κ型23例, λ型9例。11例患者接受过含新药的化疗。全组中位生存期为42个月。单因素分析显示, 年龄、B2微球蛋白、白蛋白水平及是否接受新药治疗是影响总生存时间(OS)的预后因素。COX多因素回归分析显示, B2微球蛋白是影响OS的独立预后因素(P=0.05)。结论: 华氏巨球蛋白血症好发于老年男性, 临床表现无特异性, B2微球蛋白>3 mg/L提示患者预后不良, 新药治疗可能有助于改善患者预后。

Abstract: Objective: To evaluate the clinicopathological characteristics, therapy and prognosis of Waldenström macroglobulinemia(WM).Methods: The clinical data of 32 patients with WM diagnosed in our center between January 1, 2001 and May 30, 2018 were retrospectively analyzed.The clinical and pathological features, treatment and outcome were summarized and the high risk factors affecting the prognosis of patients were investigated.Results: 25 patients were males and 7 patients were females.The median age was 64 years.The patients in low-, intermediate-and high-risk group accounted for 3.1%, 43.8% and 53.1%, respectively.90.6% of the initial symptoms were anemia.The median hemoglobin was 75.5 (26~146) g/L, and the median IgM was 52.4(13~110) g/L.23 patients were κ light chain and 9 patients were λ light chain.11 patients received chemotherapy including novel drugs.The median overall survival was 42 months.In univariate analysis, age, B2-microglobulin(B2-MG), albumin and novel drugs were prognostic factors.Multivariate analysis showed B2-MG was a significant prognostic factor for overall survival(P=0.05).Conclusion: WM patients were commonly seen in elder patients with no special clinical symptoms.B2-MG>3 mg/L indicated poor prognosis.Novel drugs may improve the outcome.

参考文献/REFERENCES

- [1]Gertz MA.Waldenstrom macroglobulinemia: 2015 update on diagnosis, risk stratification, and management [J].Am J Hematol, 2015, 90(4): 346-354.
- [2]Buske C, Leblond V, Dimopoulos M, et al.Waldenstrom's macroglobulinaemia: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up [J].Ann Oncol, 2013, 24 Suppl 6: vi155-159.
- [3]Hematology Oncology Committee of China Anti-Cancer Association, Leukemia & Lymphoma Group Society of Hematology at Chinese Medical Association, Union for China Lymphoma Investigators.The consensus of the diagnosis and treatment of lymphoplasmacytic lymphoma/Waldenström macroglobulinemia in China (2016

- version) [J]. *Chin J Hematol*, 2016, 37(9): 729-734. [中国抗癌协会血液肿瘤专业委员会, 中华医学会学会业分会白血病淋巴瘤学组, 中国抗淋巴瘤联盟. 淋巴浆细胞淋巴瘤/华氏巨球蛋白血症诊断与治疗中国专家共识(2016年版) [J]. *中华血液学杂志*, 2016, 37(9): 729-734.]
- [4] Morel P, Duhamel A, Gobbi P, et al. International prognostic scoring system for Waldenstrom macroglobulinemia [J]. *Blood*, 2009, 113(18): 4163-4170.
- [5] Buske C, Leblond V. How to manage Waldenstrom's macroglobulinemia [J]. *Leukemia*, 2013, 27(4): 762-772.
- [6] Braggio E, Philipsborn C, Novak A, et al. Molecular pathogenesis of Waldenstrom's macroglobulinemia [J]. *Haematologica*, 2012, 97(9): 1281-1290.
- [7] Simon L, Baron M, Leblond V. How we manage patients with Waldenstrom macroglobulinaemia [J]. *Br J Haematol*, 2018, 181(6): 737-751.
- [8] Castillo JJ, Ghobrial IM, Treon SP. Biology, prognosis, and therapy of Waldenstrom macroglobulinemia [J]. *Cancer Treat Res*, 2015, 165: 177-195.
- [9] Sahin I, Leblebjian H, Treon SP, et al. Waldenstrom macroglobulinemia: From biology to treatment [J]. *Expert Rev Hematol*, 2014, 7(1): 157-168.
- [10] Chin CK, Leslie C, Grove CS, et al. The diagnostic, prognostic, and therapeutic utility of molecular testing in a patient with Waldenstrom's macroglobulinemia [J]. *Int J Mol Sci*, 2017, 18(10): 1-6.
- [11] Treon SP, Xu L, Yang G, et al. MYD88 L265P somatic mutation in Waldenstrom's macroglobulinemia [J]. *N Engl J Med*, 2012, 367(9): 826-833.
- [12] Cao XX, Meng Q, Cai H, et al. Evaluation of clinical characteristics, MYD88 mutation, CXCR4 mutation and prognosis in Waldenstrom's macroglobulinemia: A single center retrospective study of 93 patients [J]. *Chin J Hematol*, 2017, 38(6): 494-498. [曹欣欣, 孟琦, 蔡浩, 等. 华氏巨球蛋白血症患者的临床特征、MYD88L265P、CXCR4WHIM突变和预后: 单中心93例回顾性分析 [J]. *中华血液学杂志*, 2017, 38(6): 494-498.]
- [13] Gertz MA. Waldenstrom macroglobulinemia treatment algorithm 2018 [J]. *Blood Cancer J*, 2018, 8(40): 1-7.
- [14] Dimopoulos MA, Kastritis E, Owen RG, et al. Treatment recommendations for patients with Waldenstrom macroglobulinemia (WM) and related disorders: IWWM-7 consensus [J]. *Blood*, 2014, 124(9): 1404-1411.
- [15] Paludo J, Abeykoon JP, Kumar S, et al. Dexamethasone, rituximab and cyclophosphamide for relapsed and/or refractory and treatment-naive patients with Waldenstrom macroglobulinemia [J]. *Br J Haematol*, 2017, 179(1): 98-105.
- [16] Buske C, Hoster E, Dreyling M, et al. The addition of rituximab to front-line therapy with CHOP (R-CHOP) results in a higher response rate and longer time to treatment failure in patients with lymphoplasmacytic lymphoma: Results of a randomized trial of the German low-grade lymphoma study group (GLSG) [J]. *Leukemia*, 2009, 23(1): 153-161.
- [17] Souchet L, Levy V, Ouzegdouh M, et al. Efficacy and long-term toxicity of the rituximab-fludarabine-cyclophosphamide combination therapy in Waldenstrom's macroglobulinemia [J]. *Am J Hematol*, 2016, 91(8): 782-786.
- [18] Kapoor P, Ansell SM, Fonseca R, et al. Diagnosis and management of Waldenstrom macroglobulinemia: Mayo stratification of macroglobulinemia and risk-adapted therapy (mSMART) Guidelines 2016 [J]. *JAMA Oncol*, 2017, 3(9): 1257-1265.
- [19] Lee HS, Kim K, Yoon DH, et al. Clinical factors associated with response or survival after chemotherapy in patients with Waldenstrom macroglobulinemia in Korea [J]. *Biomed Res Int*, 2014, 2014: 1-7.

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