Distinct characteristics of Chinese patients with Waldenström macroglobulinemia: a report with 90 cases from a single representative center

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Abstract

Background

Waldenström macroglobulinemia (WM) is an uncommon lymphoid malignancy. The studies of WM were mostly conducted in western countries. Little is known about the WM in East Asian populations. We present the results of the systematical analysis of the clinical characteristics, immunophenotypic profile, cytogenetic aberration and prognosis of WM in a series of 90 cases from China.

Methods

Ninety patients were diagnosed with WM between June 1994 and May 2011 by reviewing the clinical information and histologic sections according to the consensus panel definition of WM at the second International Workshop for WM. Immunophenotypic profiles studied by flow cytometry in 61 patients were underwent re-analysis, and 43 patients had more than 10% cells with aberration immunophenotype, which were used here. Thirty-five patients with available bone marrow sample were examined the cytogenetic aberrations by fluorescence in situ hybridization (FISH). The prognostic factors were analyzed and compared with the western reports.

Results

The median age was 62 years with male-to-female ratio of 3.74. The most common symptoms at diagnosis were fatigue (77.8%), bleeding (20%) and edema (14.4%),

while only 6 patients (6.7%) were asymptomatic. The immunophenotypic profile of 43 cases assessed by flow cytometry was as follows:
The median percentage of lymphoplasmacytic cells was 36% (ranging from 11% to 90%). All cases tested were positive for CD19 and CD20. The positive rates for other markers were: CD22 (65.9%), CD25 (65.7%), CD23 (50.0%), FMC7 (48.6%), sIgM (18.9%), CD38(18.6%), CD11c (12.5%), CD5 (7.1%). No cases were positive for CD10 and CD103. Of the 37 patients successfully studied by FISH, the overall positivity was 41.7%, with 21.4% positivity for del(6q), 14.8% for del(17p), 7.7% for trisomy 12,7.1% for del(13q14), and 3.1% for IgH translocation. After the median follow-up of 41 months, the estimated median OS was 135 months with a 5-year OS rate of 61.8%. Parameters associated with adverse prognosis were age ($>$ 62 years), thrombocytopenia, leucopenia, cytopenias (≥2), serum creatinine (elevated) and the International prognostic scoring system for WM(high risk group) in the univariate analysis, while in the multivariate analysis, only age ($>$62 years), cytopenias (≥2) and serum creatinine (elevated) were the independent prognostic factors. The immunophenotye and cytogenetic aberrations had no prognostic value for WM. The international prognostic scoring system for WM was not the best method to predicate survival of Chinese WM patients. Using age$>$62 years and ≥2 cytopenias, three significantly different prognostic groups could been distinguished as low risk group with no factor(30%), medium risk group with one of the factors (45%) and high risk group with the two factors(25%), with 5-year's OS 71.6%, 48.6% and 17.0%, respectively ($p<0.001$).

Conclusions

Compared to the Western countries, WM in China has distinct characteristics, such as younger age, advanced disease, lower expression of sIgM and CD38, and lower incidence of del(6q). And we describe a new simple prognostic model for overall survival for newly diagnosed WM patients.