57. A Very Rare Case of Waldenström Macroglobulinemia Complicated by Cardiac Amyloidosis

Si Nguyen Van, Vuong Van Dang, Dung Hoang Nguyen, Duy Thanh Vo, Binh Duc Doan, Dung Truong My Pham, University of Medicine and Pharmacy at Ho Chi Minh city, Ho Chi Minh, Vietnam, Nhan dan Gia Dinh hospital, Ho Chi Minh, Vietnam

Body

Background: Waldenström macroglobulinemia (WM) is characterized by IgM-related multiorgan disorder. Amyloidosis is a rare complication of WM with the prevalence of 5% in all light chain amyloidosis cases and a yearly expected incidence of 0.6 cases per million. Both conditions are challenging and early treatment helps preserve organ function and prolong survival.

Case: A 68-year-old man was admitted to the hospital because of fever, fatigue, exertional dyspnea and weight loss for 3 months. A fever of unknown origin approach was initiated. Physical examination revealed macroglossia with dental impression and periorbital purpura (Figure 1A). The routine and screening test results were displayed in Table 1 with neutropenia (2.03 k/uL), hypochromic anemia and hypoalbuminemia (24.7 g/L) with A/G ratio < 1 despite normal liver function. Computed tomography scan showed right pleural effusion, mediastinal lymphadenopathy and splenomegaly. Pleural fluid is exudate with leukocytosis but tuberculosis testing was negative. Of note, electrocardiography showed low voltage on limb leads but left ventricular (LV) hypertrophy was evident on cardiac echogram with mildly abnormal LV ejection function (52%) and strain (-15.9%). Moreover, increased hs-troponin T (43.5 ng/L) and NT pro-BNP (4743 pg/mL) were detected. Systemic amyloidosis was suspected including cardiac involvement. Serum protein immunofixation electrophoresis revealed increased IgM (3630 mg/dL) with lambda light chains positive and beta 2 microglobulin was elevated (4808 ug/L). Bone marrow biopsy showed increased density (>10%) of lymphoplasmacytic colonies (Figure 1B). The diagnosis of WM was established and the resulted amyloidosis was confirmed by Congo red stain positive in both tongue and belly fat biopsies (Figure 1C). The patient was treated with Bortezomib + Dexamethasone + Rituximab. After 3 months, he showed no adverse progression of LV ejection function and strain.

Discussion: A systematic approach is essential in the diagnosis of WM since the clinical manifestation is heterogeneous. Though the rate of cardiac amyloidosis is rare in WM, this condition should be suspected from red flag signs and requires specific treatment.

Laboratory investigation	Results	
Blood count		
WBC	2.01	K/uL
NEU	0.62	K/uL
LYM	0.96	K/uL
HGB	79	g/L
HCT	25.2	%
Blood tests		
Glucose	6.74	mmol/L
Creatinine	96.5	umol/L
AST	32	IU/L
ALT	38	IU/L
Bilirubin total	10.6	mmol/L
Bilirubin direct	4.0	mmol/L
Protein	80.8	g/L
Albumin	24.7	g/L
A/G ratio	0.44	
LDH	112.5	IU/L
CRP	31.95	mg/L
ESR 1h-2h	136 - > 140	s
Serum Fe	22	IU/dL
Ferritine	264	mg/dL
TSAT	9.87	%
Calcium	2.09	mmol/L
Bacterial culture	Negative	
Pleural fluid tests		
ADA	29	IU/L
Protein	38.4	g/L
Glucose	6.32	mmol/L
LDH	115.2	IU/L
Cells	1440 leukocytes (90% monocytes)	
Bacterial culture	Negative	
TB PCR	Negative	
Cardiac biomarkers		
hs troponin T	43.5	ng/L
NT pro-BNP	4743	pg/mL
Immunoglobulin tests		
Beta 2 microglobulin	4804	IU/L
Serum fixation immunoelectropheresis		
- Kappa, Lambda light chains	IgM Lambda detected	
- Protein	72	g/L
- IgG	960	mg/dL
- IgA	69	mg/dL
- IgM	3630	mg/dL
- Kappa	Not detected	
- Lambda	Detected	

Table 1. Laboratory investigation results.