Original article

Clinical manifestations and outcomes of systemic light chain amyloidosis: Chiang Mai University Hospital experience

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Abstract:

Objectives: *The study's primary objective aimed to investigate clinical manifestations including symptoms, signs and laboratory findings among patients with systemic light chain (AL) amyloidosis. The secondary objective was to study clinical outcomes among patients with AL amyloidosis.* ***Material and Methods:*** *This study employed a retrospective design. We included all patients who had received a diagnosis of systemic AL amyloidosis and were treated in Chiang Mai University Hospital from January 2002 to October 2018. Data were obtained through the electronic database and clinical records including demographic data, clinical presentation, laboratory characteristics, treatment and outcomes.* ***Results:*** *A total of 28 patients were identified. The median age at diagnosis was 63 years old (range 39 to 85 years old) and 71.4% were male. The most common initial clinical presentation was dyspnea on exertion (42.8%). The two most frequent organs involved among our patients were the heart (60.7%) and kidney (53.5%). Most patients underwent chemotherapy (85.7%), mainly a melphalan-based regimen (87.5%). Of the treated patients, 25% had a hematologic response (very good partial response 12.5%, partial response 12.5%). The mortality rate was 82.1% with a median follow-up time of 11 months. Univariate and multivariable Cox regression analysis revealed that presence of Bence Jones protein in the urine was a poor prognostic factor for survival.* ***Conclusion:*** *The most common organ involved in systemic AL amyloidosis was the heart. Overall-survival of patients with AL amyloidosis was poor and most cases had a poor response to treatment.*

Keywords : ● *AL amyloidosis* ● *Amyloidosis* ● *Immunoglobulin light chain* ● *Multiple myeloma*

J Hematol Transfus Med. 2020;30:51-60.

Received 12 November 2019 Corrected 13 November 2019 Accepted 27 December 2019

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นิพนธ์ต้นฉบับ

ลักษณะทางคลินิกและผลการรักษาโรคอะมัยลอยโดสิสชนิดไลท์เชน: ประสบการณ์ของโรงพยาบาลมหาวิทยาลัยเชียงใหม่

จอมชัย ลือชูวงศ์ เอกรัฐ รัฐฤทธิ์ธำรง ปกป้อง พิริยคุณธร ธนาวัฒก์ รัตนธรรมเมธี ศศินี ฮันตระกูล
ชาตรี ชัยอดิศักดิ์โสภา อดิศักดิ์ ตันติวรวิทย์ และ ลลิตา นรเศรษฐ์ธาดา
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บทคัดย่อ

วัตถุประสงค์ *วัตถุประสงค์หลักเพื่อศึกษาลักษณะทางคลินิกได้แก่ อาการ อาการแสดง และผลการตรวจทางห้องปฏิบัติการในผู้ป่วยอะมัยลอยโดสิสชนิดไลท์เชน (เอแอลอะมัยลอยโดสิส) วัตถุประสงค์รองเพื่อศึกษาผลการรักษาของผู้ป่วยเอแอลอะมัยลอยโดสิส* ***วัสดุและวิธีการ*** *การศึกษาแบบย้อนหลัง โดยรวบรวมผู้ป่วยที่ได้รับการวินิจฉัยเป็นโรคเอแอลอะมัยลอยโดสิส และได้รับการรักษาในโรงพยาบาลมหาราชนครเชียงใหม่ตั้งแต่มกราคม พ.ศ. 2545 ถึงตุลาคม พ.ศ. 2561 โดยเก็บข้อมูลจากเวชระเบียนผู้ป่วยและข้อมูลระบบอิเล็กทรอนิกส์ ได้แก่ ข้อมูลพื้นฐาน อาการที่มาพบแพทย์ การตรวจทางห้องปฏิบัติการ การรักษาและผลการรักษา* ***ผลการวิจัย*** *ได้รวบรวมข้อมูลจากผู้ป่วยทั้งหมด 28 ราย ค่ามัธยฐานของอายุเท่ากับ 63 ปี (พิสัย 39-85 ปี) และร้อยละ 71.4 เป็นเพศชาย อาการที่มาพบแพทย์ที่พบบ่อยที่สุดคือ อาการเหนื่อยเมื่อออกแรง (ร้อยละ 42.8) 2 อวัยวะที่พบโรคมากที่สุดคือ หัวใจ (ร้อยละ 60.7) และไต (ร้อยละ 53.5) ผู้ป่วยส่วนใหญ่ (ร้อยละ 85.7) ได้รับยาเคมีบำบัดโดยเฉพาะสูตรยาที่มี melphalan (ร้อยละ 87.5) ในผู้ป่วยที่ได้รับการรักษาพบว่ามีการตอบสนองของระบบโลหิตร้อยละ 25 (very good partial response ร้อยละ 12.5 และ partial response ร้อยละ 12.5 อัตราการเสียชีวิตเท่ากับร้อยละ 82.1 เมื่อติดตามผู้ป่วยที่ค่ามัธยฐาน 11 เดือน การวิเคราะห์แบบ univariate และ multivariable Cox regression analysis พบว่าการมี Bence Jones protein ในปัสสาวะเป็นปัจจัยที่มีผลต่อการเสียชีวิต* ***สรุป*** *อวัยวะที่พบโรคเอแอลอะมัยลอยโดสิสมากที่สุดคือหัวใจ มีอัตราการรอดชีวิตที่ต่ำและการตอบสนองต่อการรักษาของผู้ป่วยส่วนใหญ่ยังไม่ดีนัก*

คำสำคัญ : ● *เอแอลอะมัยลอยโดสิส* ● *อะมัยลอยโดสิส* ● *อิมมูโนโกลบูลินชนิดไลท์เชน* ● *มะเร็งเม็ดเลือดชนิดมัยอิโลมา*
วารสารโลหิตวิทยาและเวชศาสตร์บริการโลหิต. 2563;30:51-60.

Introduction

Amyloidosis is a rare disease characterized by the deposition of insoluble fibril proteins in various organs resulting in abnormal organ function.[1-3] Recently, 36 amyloidogenic precursors have been described among humans.[1] Of these, monoclonal immunoglobin light chains are the most common, leading to a condition known as light-chain (AL) amyloidosis.[3] Approximately 20% of cases of AL amyloidosis are secondary to multiple myeloma (MM).[4] However, primary systemic AL amyloidosis or localized AL amyloidosis can be found separate to MM.[1] The incidence rate of AL amyloidosis in Olmsted County, Minnesota, from 1990 through 2015 was 1.2 per 100,000 person-years with a median age at diagnosis of 76 years. Fewer than 5% of cases occurred among patients aged less than 50 years.[5]

Clinical manifestations of patients with systemic AL amyloidosis depend on organ involvement, commonly the heart, kidneys, liver, peripheral nerve and soft tissue.[6] A large cohort study in the USA found that the majority of patients presented nephrotic syndrome (28%) followed by congestive heart failure (17%).[6] Apart from these amyloid-related systemic syndromes, diagnosing systemic AL amyloidosis requires the presence of an amyloid substance in a tissue biopsy with evidence of monoclonal light-chain deposition in an amyloid substance.[7] Abdominal fat pad aspiration is a minimally invasive procedure with a high specificity (100%) and acceptable sensitivity (80-93%) useful to diagnose amyloidosis.[8] Laboratory tests for monoclonal light chains such as serum free light chain (FLC), serum and urine immunofixation electrophoresis (IFE) are recommended.[2,3]

The treatment of AL amyloidosis is mainly adapted from that for MM to reduce amyloidogenic light chain production.[2,3] The hematologic response is associated with good survival outcomes[9], while organ response may occur among some patients.[10] In addition, cardiac involvement, as determined by an increase in serum cardiac markers (cardiac troponins and N-terminal pro-brain natriuretic peptides, NT-proBNP), is associated with poor survival outcomes among patients with AL amyloidosis.[11] Melphalan combined with dexamethasone (MDex) is considered a mainstay therapy, especially in transplant ineligible patients[2] with a hematologic response of 68% and organ response of 39%.[10] Novel agent-based induction therapies, such as bortezomib, thalidomide and lenalidomide have been shown to improve response rate and overall survival rate (OS) when compared with historical data associated with the MDex regimen.[2,3,12] Autologous stem cell transplantation (ASCT) is considered among fit patients without advanced cardiac dysfunction.[2,3]

Limited data is available regarding systemic AL amyloidosis in Thailand[13,14] leading to the two main objectives in this study. The primary objective aimed to study clinical manifestations, including symptoms, signs and laboratory findings among patients with AL amyloidosis. The secondary objective was to study clinical outcomes among patients with AL amyloidosis in Chiang Mai University Hospital, Thailand.

Materials and Methods

This study employed a retrospective designed and was conducted in Chiang Mai University Hospital, Chiang Mai, Thailand between January 1st, 2002 and October 31st, 2018. The study was approved by the Institutional Ethics Research Committee of the Faculty of Medicine, Chiang Mai University in accordance with the declaration of Helsinki. Patients aged at least 18 years who had received a diagnosis of systemic AL amyloidosis were enrolled in the study. The diagnosis of AL amyloidosis required: (1) the presence of amyloid protein determined by positive Congo red staining under polarized light in tissue biopsy and (2) either immunohistochemical characterization of the amyloid deposits or presence of a monoclonal protein (M-protein) in serum or urine or a monoclonal staining of plasma cells in bone marrow.[10] M-protein was detected by serum (SPEP), urine protein electrophoresis (UPEP), the urine Bence Jones protein test (UBJ), serum IFE or serum FLC. The latest 2 methods were available in Chiang Mai University Hospital after

year 2009. Patients with non-AL amyloidosis and localized amyloidosis were excluded.

Clinical data, including demographic data (age, sex), signs, symptoms, organ involvement according to 10th International Symposium on Amyloid and Amyloidosis[15], the association with MM, laboratory investigations (complete blood count, urinalysis, urine protein, M-protein analysis), pathological findings, location of tissue biopsy and treatment were collected from the electronic database. Hematologic response was defined as the following: stringent complete response (sCR); complete response (CR); very good partial response (VGPR); partial response (PR); stable and progressive disease.[16] Organ response according to consensus criteria[15]was assessed after completion of treatment. Survival, cause of death and factors associated with survival were analyzed.

Statistical analysis

Data regarding clinical manifestations, laboratory tests, and treatment are presented using descriptive statistics. Dichotomous variables are presented as percentages or proportions. Continuous variables are presented as mean ± standard deviation (SD) or median (range) as appropriate. Overall survival was calculated using the Kaplan-Meier method. Factors associated with OS were determined by univariate and multivariate analysis using the Cox proportional hazard model. SPSS, Version 23.0 was used for data analysis.

Results

Clinical presentations

Twenty-eight patients were included in the study. Of these, 20 patients (71.4%) were male and 8 patients (28.6%) were diagnosed before year 2009. Baseline characteristics of the patients are shown in Table 1. Median age was 63 years (range 39-85) with 3 patients (10.7%) being diagnosed at an age less than 50 years (39, 40, and 49 years). The most common presentation was dyspnea (12 patients, 42.8%), followed by edema (10 patients, 35.7%). Other presentations included thickening of the skin, eyelid mass, abdominal mass, lymphadenopathy, macroglossia and numbness (1 patient each, 3.5%).

The heart was the organ most frequently involved (17 patients, 60.7%) with AL amyloidosis in this study, followed by the kidney (15 patients, 53.5%). Ten patients (35.7%) had combined cardiac and kidney involvement. The remainder of the patients had liver (3 patients, 10.7%), dermatologic (3 patients, 10.7%) and peripheral nerve involvement (1 patient, 3.6%). The most common symptoms of patients with cardiac involvement were dyspnea on exertion (66%), followed by orthopnea (26%) and paroxysmal nocturnal dyspnea (8%). Mean pro-BNP obtained from 6 patients was 3,741 ± 6,704 pg/mL and mean cardiac troponin-T (obtained from 3 patients) was 28.2 ± 23.2 pg/mL (normal < 14 pg/mL).

All patients with kidney involvement had generalized edema, proteinuria and hypoalbuminemia (nephrotic syndrome, 100%) with a mean 24-hour urine protein of 5.1 ± 2.8 gram/day and spot urine protein and creatinine ratio of 3.6 ± 3.4 gram/day. All 3 patients with liver involvement had hepatomegaly (100%) and 1 patient (33.3%) had an abnormal liver function test.

Four patients (14.3%) had a concurrent diagnosis with MM, whereas two patients (10.7%) subsequently had a progression to full blown MM in 16 and 52 months.

Laboratory investigations

Tissue diagnoses of amyloidosis were evidenced by abdominal pad fat biopsy among 16 patients (57.1%), kidney biopsy among 4 patients (14.3%), and skin biopsy among 3 patients (10.7%). A single patient (3.6%), four in total, had a diagnosis of AL amyloidosis from one of the following biopsies: bone marrow (BM), liver, lymph node, and periorbital mass.

Complete blood count results showed a mean hemoglobin level of 11.6 ± 2.3 g/dL, a mean white blood cell count of 8.018 ± 3.268 $x10^9$/L and a mean platelet count of 236.750 ± 68.730 $x10^9$/L. To determine the M-protein, a positive UBJ was detected among 18 patients (64.3%), and a positive SPEP or UPEP among 3 patients each (10.7%). Serum IFE detected monoclonal immunoglobulin

Table 1 Clinical characteristics and treatment outcomes of patients with systemic AL amyloidosis

Clinical Characteristic and treatment outcome (n= 28)		
Age, median (range)	Age (year)	63 (39-85)
Sex, n (%)	Male	20 (71.4)
	Female	8 (28.5)
First Clinical Presentation, n (%)	Dyspnea	12 (42.8)
	Edema	10 (35.7)
	Abdominal Mass	1 (3.5)
	Orbital Mass	1 (3.5)
	Macroglossia	1 (3.5)
	Lymphadenopathy	1 (3.5)
	Loss of Sensation	1 (3.5)
	Skin Thickening	1 (3.5)
Organ Involvement, n (%)	Heart	17 (60.7)
	Kidneys	15 (53.5)
	Heart with Kidneys	10 (67.8)
	Liver	3 (7.1)
	Skin	3 (7.1)
	Soft Tissue	1 (3.5)
	Peripheral Neuropathy	1 (3.5)
Clinical Presentations according to Organ Involvement		
Kidney, n (%)	Proteinuria	15 (53.5)
	Hypoalbuminemia	15 (53.5)
	Generalized Edema	15 (53.5)
Heart, n (%)	Dyspnea on Exertion	10 (35.7)
	Orthopnea	4 (14.2)
	Paroxysmal Nocturnal Dyspnea	1 (3.5)
Liver, n (%)	Hepatomegaly	3 (10.7)
	Abnormal Liver Function Test	1 (3.5)
Soft Tissue, n (%)	Skin thickness	1 (3.5)
	Macroglossia	1 (3.5)
	Orbital mass	1 (3.5)
	Lymphadenopathy	1 (3.5)
Complete Blood Count, mean ± SD	Hemoglobin (g/dL)	11.5 (±2.3)
	White Blood Cells (cell/cu.mm)	8,018 (±3,268)
	Platelet (cell/cu.mm)	236,750 (±68,730)
Urine Protein, Mean ± SD	24 Hours Urine Protein	5.08 (±2.8)
	Spot Urine Protein Creatinine Ratio	3.61 (±3.4)

Table 1 Clinical characteristics and treatment outcomes of patients with systemic AL amyloidosis (continue)

Detection of Monoclonal Protein, n (%)	Urine Bence Jones Protein	18 (64.3)
	Serum Protein Electrophoresis	3 (10.7)
	Urine Protein Electrophoresis	3 (10.7)
	Serum Immunofixation Electrophoresis	5/7 (71.4)
	Serum Free Light Chain	16/20 (80)
Serum free light chain, mean ± SD	Difference in Free Light Chains (mg/L)	672.9 (±1474)
	Serum Free Light Chain Ratio (Kappa/Lambda)	35.51 (±119)
Type of monoclonal protein, n (%)	Lambda Light Chain	7 (38.9)
(Identified in 18 patients)	Kappa Light Chain	5 (27.8)
	IgG Lambda	2 (11.1)
	IgA Kappa	1 (5.6)
	IgA Lambda	1 (5.6)
	IgM Lambda	1 (5.6)
	IgE	1 (5.6)
Site of Tissue Diagnosis	Abdominal Fat Pad	15 (53.5)
	Kidney	5 (17.8)
	Skin	3 (10.7)
	Liver	2 (7.1)
	Bone marrow	1 (3.5)
	Lymph node	1 (3.5)
	Periorbital Mass	1 (3.5)
Treatment Options, n (%)	Chemotherapy	24 (85.7)
	Autologous Stem Cell Transplantation	0 (0)
	No treatment	4 (14.3)
Chemotherapy Regimens, n (%)	Melphalan-Dexamethasone	11 (45.8)
	Melphalan-Prednisolone	10 (41.6)
	Bortezomib-Dexamethasone	2 (8.3)
	Bortezomib-Cyclophosphamide-Dexamethasone	1 (4.1)
Response Rate, n (%)		
Hematologic Response	Complete Response	0 (0)
	Very Good Partial Response	3 (12.5)
	Partial Response	3 (12.5)
	No Response	18 (75.0)
Organ response	Kidney	2 (7.1)
Death, n (%)		23 (82.1)
Ongoing Follow Up, n (%)		5 (17.9)

was found among 5 of 7 patients (71.4%). Of the tested patients, an abnormal FLC ratio was found in 16 of 20 patients (80%). Direct immunofluorescence in amyloid substances was performed among 3 patients (10.7%) and revealed lambda light chain restriction in all cases including one patient who had normal SPEP, UPEP and serum FLC results. Lambda light chain was the most common subtype of AL amyloidosis (38.9%) followed by kappa light chain (27.8%). Median levels of BM plasma cells were 4.5% (range 1%-43%) and 21.5% (range 10%-55%) in primary and secondary systemic AL amyloidosis, respectively.

Treatment and outcomes

Twenty-four patients (85.7%) underwent chemotherapy, whereas 4 patients (14.3%) did not receive specific treatment. The most frequently prescribed chemotherapy was a melphalan-based regimen, including melphalan-dexamethasone (MDex) (11 patients, 45.8%) and melphalan-prednisolone (MP) (10 patients, 41.6%). Other patients received bortezomib-dexamethasone (BD) (2 patients, 8.3%) and bortezomib-cyclophosphamide-dexamethasone (BCD) (1 patient, 4.1%). No patient underwent ASCT.

Hematologic response was achieved among 6 patients (25%) with VGPR among 3 patients (12.5%) (1 BCD, 1 BD, 1 MDex) and PR also among 3 patients (12.5%) (1 BD, 1 MDex, 1 MP). Organ response was observed only in 2 of 24 (8.3%) patients. Both cases involved kidney response among patients receiving MP.

With a median follow-up period of 11 months (range 2-97 months), 23 patients (82.1%) died. The documented causes of death among 3 patients were pneumonia (2 patients) and renal failure (1 patient). Median OS was 42 weeks (range 8-257 weeks) and estimated 5-year OS was 18% (Figure 1). Univariate and multivariable analysis showed the presence of UBJ was the only factor significantly associated with survival ($p < 0.05$). (Table 2)

Discussion

Amyloidosis is a rare disease. The reported annual incidences in England and the USA are only 0.3 and 1-1.4 per 100,000 patients, respectively.[17,18] In the current study, only 28 patients with systemic AL amyloidosis were identified during a 16-year period in Chiang Mai University Hospital. The majority of patients were male (71.4%) and elderly as only 10.7% were aged

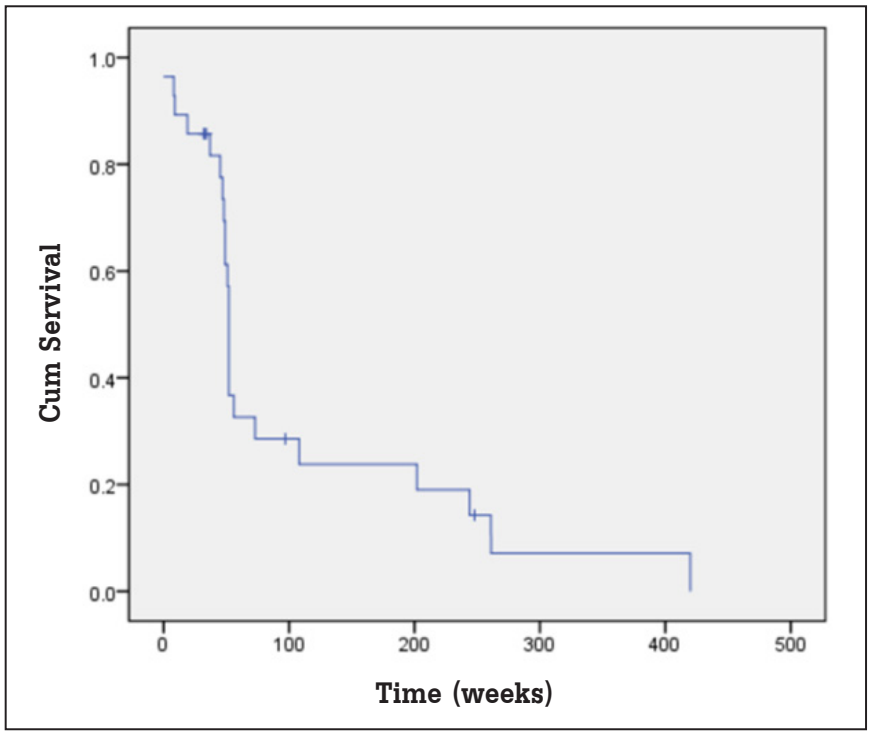

Figure 1 Kaplan–Meier survival curves showing overall survival among patients with systemic AL amyloidosis

Table 2 Association of factors with death among patients with systemic AL amyloidosis

Factors	95%CI	P-value
Sex	0.52-2.12	0.533
Age	0.72-2.53	0.386
Clinical Presentation	0.42-3.40	0.58
Proteinuria	0.43-1.35	0.52
Hypoalbuminemia	0.55-1.76	0.502
Edema	0.55-1.76	0.502
AKI	0.27-2.25	0.154
Dyspnea on Exertion	0.28-1.95	0.391
PND	0.27-1.52	0.63
Orthopnea	0.49-2.25	0.31
Cardiomegaly	0.90-1.90	0.09
Peripheral Neuropathy	0.46-2.14	0.63
Hepatomegaly	0.75-2.25	0.39
Urine Bence Jones protein	1.52-7.81	0.005
SPEP Positive	0.72-3.31	0.69
UPEP Positive	0.69-2.98	0.602
Serum Immunofixation Positive	0.95-1.93	0.082
Hematologic Response	0.96-2.21	0.085
Association with MM	0.63-1.32	0.536

AKI: acute kidney injury; PND: paroxysmal nocturnal dyspnea; SPEP: serum protein electrophoresis; UPEP: urine protein electrophoresis; MM: multiple myeloma

less than 50 years. Studies conducted in Minnesota[5] and England[17] also revealed a lower proportion of AL amyloidosis among younger patients, aged less than 50 years, of 2.8% and 6.9%, respectively and a similar male predominance (54% in the Minnesota study[5]). The related studies from Thailand comprised case reports of systemic AL amyloidosis with cardiac[13] and respiratory tract involvement.[14] The current study aimed to provide more information about clinical and laboratory features of systemic AL amyloidosis in Thailand.

The most common presentations of AL amyloidosis in the current study were dyspnea and edema, comprising 40% and 35% of patients, respectively. This indicated that the heart (60.7%) and kidneys (53.5%) were the organs most involved. These findings were comparable with the Minnesota study reporting that the majority of patients presented congestive heart failure (29%-54%) and nephrotic syndrome (26%).[5] However, a nationwide survey of 741 patients with systemic AL amyloidosis from Japan showed the kidneys (73.1%) were the most affected organs, followed by the heart (34%).[19] Other clinical findings including hepatomegaly, skin involvement, lymphadenopathy and peripheral neuropathy were relatively uncommon. In our study, no patients were documented as having GI tract or autonomic nervous system involvement although these were identified at an incidence of 22.1% and 17.6%, respectively, in the Japanese study.[19]

Tissue biopsy demonstrating a positive staining with Congo red is important to diagnose systemic amyloidosis. A combination of abdominal fat pad biopsy and BM biopsy improved the diagnostic yield from 49-76 to 90%. In addition, a corresponding figure for immunohistochemical staining for typing of the amyloid was also demonstrated.[20] In this study, an abdominal fat pad biopsy led to diagnosing amyloidosis in more than

one half of the patients and then supported the role of fat pad biopsy as an initially less invasive investigation for making the diagnosing.

The next step for diagnosing systemic AL amyloidosis is to demonstrate the presence of light chain-related amyloid substances using mass spectrometry-based proteomic analysis or immunoelectronmicroscopy.[7] However, these methods are still not widely available. As a result, indirect evidence by determining monoclonal protein in serum or urine is still useful to diagnose AL amyloidosis in real-life practice. This study showed that serum FLC, a newer diagnostic method, detected monoclonal proteins in up to 80% of cases, exhibiting a higher sensitivity than other methods (SPEP 10.7%, UPEP 10.7%, UBJ 64.3%, serum IFE 71.4%). This finding supported the role of serum FLC to diagnose AL amyloidosis.[21]

Of 85.7% of the patients receiving chemotherapy, a hematologic response was only achieved in 25% of cases, including VGPR and PR. The low response rate in the current study might be explained by the pattern of treatment. Almost 90% of these patients received non-novel agent-based regimens, including MDex and MP. More importantly, no patient underwent ASCT. These results were explained in that novel agents and ASCT were not approved and could not be reimbursed for treatment of primary AL amyloidosis in Thailand during the study period. The 5-year OS rate of 18% and mortality rate of 82.1% with a median OS of 42 weeks in the current study were comparable with the data published by the authors of the Minnesota study before 1990, reporting a median OS of 2.3 years and mortality rate of 86%.[22] On the contrary, ASCT has been shown to result in a better response rate (CR rate of 51%) and long term results with a median OS of 4.75 years.[23] The 20-year OS of patients undergoing ASCT was 28.6%. The rate of OS was pronounced among patients in whom CR was achieved.[24] More recently, an induction therapy and a conditioning regimen containing a novel-agent such as bortezomib has indicated the potential for better responses, with a reported hematologic response up to 100%.[25]

Factors associated with survival among patients with AL amyloidosis according to the related study were levels of serum cardiac troponins and NT pro-BNP.[11] However the prognostic value of these cardiac markers was not analyzed in this study due to the limited numbers of tested patients. In the current study, the presence of UBJ was the only adverse prognostic factor for survival. The laboratory test for UBJ is widely available to detect urine light chain disease. However, the sensitivity of the UBJ test was not as high as serum FLC or IFE.[21] The presence of UBJ may reflect a high disease burden which affects the survival rate of patients with AL amyloidosis.

Some limitations were found in this study. First of all, this study employed a retrospective design, which might have led to selection bias and incomplete data collection especially data regarding novel prognostic markers as well as comprehensive history and physical examination. Secondly, the small sample size might have affected the analysis of prognostic factors for survival rate as well as the generalizability of the clinical and laboratory findings associated with AL amyloidosis in Thailand. A larger prospective multi-center cohort study is warranted in Thailand to determine the epidemiology, clinical manifestations and outcomes of systemic AL amyloidosis.

Summary

The most common organ involvement of systemic AL amyloidosis in Chiang-Mai University Hospital was that of the heart followed by the kidneys. Overall survival among our patients with AL Amyloidosis was poor and most cases had a poor response to treatment. Only the presence of Bence Jones proteinin urine seemed to be associated with survival outcome in this study.

Acknowledgment

This study was supported by Department of Internal Medicine, Faculty of Medicine, Chiang Mai University, Thailand.

Statement of conflicts of interest: The authors report no conflict of interest.

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