內科學誌 2005 ：16 ： 18-25

The Clinical Study of Autoimmune Hepatitis

Lok-Beng Koay, and Ching-Nan Lin[1]

Division of Gastroenterology and Hepatology,
Department of Internal Medicine, and Pathology[1]
Chi Mei Medical Center, Tainan, Taiwan, R.O.C.

Abstract

Autoimmune hepatitis (AIH) is thought to be rare in Taiwan. By using the diagnostic scoring system recommended by the International Autoimmune Hepatitis Group, 27 cases of AIH patients had been diagnosed. We herein describe the study with respect to the clinical, laboratory and histological features at presentation. All patients were consecutively diagnosed over a period of 3 years. The medical records of patients were reviewed and analyzed. The clinical information, laboratory data and histological findings were assessed in great detail. Twenty-seven patients were diagnosed as AIH Type Ⅰ, with a median age of 58 years and a female-male ratio of 23:4. The most common clinical features at presentation were fatigue, jaundice and abdominal fullness. ANA positive rate was 96%, and most of the patients showed elevated values of AST, ALT, serum globulin and bilirubin. A substantial proportion of patients presented with poor liver function at entry and 37% of patients had liver cirrhosis, with relatively prolonged PT (p=0.030) and poorer outcome (p=0.041) as compared to the non-cirrhotics. As a whole there was a favorable treatment response and the overall survival rate is 88.9%. The AIH incidence is much higher than previously presumed and AIH type Ⅰ is the predominant type of the disease. Although a substantial proportion of AIH patients presented with poor hepatic function at entry, the overall survival rate is high (88.9%). (J Intern Med Taiwan 2005; 16: 18-25)

Key Words ： Autoimmune hepatitis, Antinuclear antibodies, Liver cirrhosis, Autoimmune hepatitis type Ⅰ

Correspondence and requests for reprints : Dr. Lok-Beng Koay
Address : Division of Gastroenterology and Hepatology, Department of Internal Medicine, Chi Mei Medical Center, 901 Chung Hwa Road, Yung Kang City, Tainan, Taiwan, R.O.C.

Introduction

Autoimmune hepatitis (AIH) is an uncommon disease characterized by hypergammaglobulinemia, circulating autoantibodies, female predominance, response to immunosuppressive therapy and association with an immunogenetic background[1,2]. The disease is more prevalent in western countries, with an estimated prevalence of 170 cases per million population in Northern Europe[3], whereas low prevalent rates were reported in Asian countries[4-6], and likewise very low incidence was found in Taiwan. AIH is difficult to diagnose, as there is no single diagnostic test for it. The diagnosis of AIH is based on careful exclusion of other causes of liver diseases, together with the finding of suggestive features that are typical of AIH. A diagnostic scoring system for AIH was recommended by the International Autoimmune Hepatitis Group (IAIHG) in 1993, and subsequently revised in 1999 to improve its specificity and to simplify its usage[7,1]. The scoring system had been widely accepted and well validated[8]. It provides an effective and convenient method for the clinicians to define AIH. We had used this diagnostic scoring system and had diagnosed 27 cases of AIH in a period of three years. We herein describe our study with repect to the clinical, biochemical, serological and histological findings.

Methods

The medical records of 27 patients who were diagnosed as AIH in Chi Mei Medical Center, a tertiary referral center in southern Taiwan, were collected and analyzed. All patients were consecutively seen between April 2000 and September 2002. The clinical information of the patients was then assessed in detail by using a predesigned uniform protocol for data collection. The diagnosis of AIH was made according to the revised criteria of the International Autoimmune Hepatitis Group. Both the definite and probable categories of AIH patients were included in this study.

The history of disease onset, hepatitis B and C, alcohol, prescription of chronic medication, blood transfusion and family history of autoimmune disease was obtained. The presenting symptoms and signs were obtained in great detail. The clinical outcome, treatment with steroids, survival were recorded.

The laboratory data included: a complete liver function test, serum albumin and globulin levels, immunoglobulin (Ig) G, prothrombin time, viral markers for hepatitis B and C by using the method of Microparticle Enzyme Immunoassay (Abbott laboratory, Chicago, USA), HCV-RNA by polymerase chain reaction test. Serological tests for antinuclear antibodies (ANA), antismooth muscle antibodies (SMA), antimitochondrial antibodies (AMA) and perinuclear staining anti-neutrophil cytoplasmic antibodies (pANCA) were performed using the indirect immunofluorescence technique (Medical & Biological laboratories Co, LTD, Japan). Additional tests including serum iron, TIBC, ceruloplasmin, Alpha 1-antitrypsin test and serological tests for Epstein-Barr virus and cytomegalovirus were done in a large proportion of the patients to exclude other causes of liver disease. Thyroid function tests and antibodies to antithyroglobulin were done in patients with thyroid dysfuction, and HLA-tissue typing was performed in 8 patients for study of the genetic status.

Liver biopsies were performed and the specimens were reviewed twice by experienced pathologists. The histological features relevant to AIH were assessed according to the IAIHG criteria, and presence of liver cirrhosis was documented. Abdominal sonography (using Toshiba SSA-340A ultrasound machine, Tokyo, Japan) was performed in every patient to exclude obstructive biliary disease, and detect the presence of cirrhosis, ascites, hepatomegaly and splenomegaly. Abdominal CT scan and endoscopic retrograde cholangiography was performed in selected cases to exclude liver tumors and biliary tract diseases.

All values were expressed both as means and medians. Fisher's exact test and Wilcoxon Rank Sum test were used for statistical analysis, and Kaplan-Meier curve was used for survival evaluation.

Results

Sex and age distribution

During the period of April 2000 to September 2002, 27 cases of AIH were identified in Chi Mei Medical Center. Patients with hepatitis B and C were excluded from this study. All cases were ANA positive and/or SMA positive, and were defined as AIH type Ⅰ. The diagnosis was made according to the criteria recommended by the International Autoimmune Hepatitis Group, 13 patients were defined as definite AIH and 14 patients were defined as probable AIH. There was a predominance of female patients with a ratio of 23:4. The mean age of disease presentation was 56.1 ± 15.6 years and the median age was 58 (range 17-80) years. (Table 1)

Table 1. Sex and age distribution of AIH patients

Sex	No	Mean age ± SD	Median age (range)
Total	27	56.1 ± 15.6	58 (17-80)
Male	4	56.8 ± 13.4	57.5 (40-72)
Female	23	56.0 ± 16.2	58 (17-80)

The values are expressed in years.

Clinical features

The clinical features were summarized in Table 2. 63% (17/27) of the AIH patients had an insidious onset of the disease and 37% (10/27) had an acute onset, with 37% (10/27) presented with liver cirrhosis at the time of diagnosis. The most common clinical features were fatigue (15/27), jaundice (14/27) and abdominal fullness (14/27). The presence of splenomegaly (9/27), ascites (6/27) and hepatomegaly (3/27) were confirmed by abdominal sonography. Association with other autoimmune disorders were noted in 6 patients presenting with acute arthralgia and 5 patients presenting with thyroiditis.

Table 2. Clinical features of AIH patients

	No	(%)
Acute onset	10	37
Insidious onset	17	63
Cirrhosis at presentation	17	37
Fatigue	15	55.6
Jaundice	14	51.9
Abdominal fullness	14	51.9
Anorexia	11	40.7
Splenomegaly	9	33.3
Abdominal pain	8	29.6
Ascites	6	22.2
Arthralgia	6	22.2
Edema	5	18.5
Hepatomegaly	3	11.1
Pruritus	2	7.4
Fever	2	7.4
Hepatic encephalopathy	1	3.7

Laboratory data

The laboratory data were summarized in Table 3. Elevated aminotransferases were noted in 93% of patients. The median values were AST 248 IU/L (range 33.5-3090 IU/L) and ALT 169 IU/L (range 21.5-1490 IU/L). 71% of patients presented with hyperbiliru-

Table 3. Laboratory data of AIH patients

	Mean ± SD	Median (range)	Percentage of patients with abnormal value (%)
AST (8-40 IU/L)	436 ± 626	248 (33.5-3090)	93
ALT (8-40 IU/L)	395 ± 445	169 (21.5-1490)	93
Globulin (2-3.5g/DL)	4.1 ± 0.9	4.3 (2.6-5.9)	77
Bilirubin (0.2-1.2 mg/dL)	6.9 ± 7.2	5.2 (0.5-26.4)	71
Albumin (3.8-5.3 g/dL)	3.5 ± 0.8	3.5 (2.2-4.9)	64
Prothrombin time (11.3-13.3s)	15.6 ± 4.3	15.3 (10.3-26.8)	63
Immunoglobulin G (700-1600 mg/dL)	1850 ± 701	1760 (862-3790)	61
ALP (66-220 IU/L)	313 ± 198	313 (69.0-772)	54
ANA (normal titer<1:40)	396 ± 683	160 (20-2560)	96

ALP, alkaline phosphatase; ANA, antinuclear antibodies
Note : ANA value is expressed as titer.

Table 4. Histological features of AIH patients

Histology	Percentage (%)
Interface hepatitis	100
Lymphoplasmacytic infiltration	100
Rosetting of liver cells	23
Biliary changes	0
Liver cirrhosis	23

binemia, with a median value of 5.2 mg/dL (range 0.5-26.4 mg/dL). Prolonged PT was observed in 63% of patients with a median value of 15.3 s (range 10.3-26.8 s). Decreased albumin level was noticed in 64% of patients with a median value of 3.5 g/dL (range 2.2-4.9 g/dL). Increased globulin level was noticed in 77% of patients with a median value of 4.3 g/dL (range 2.6-5.9 g/dL). ANA was performed in all patients and was found to be positive in 26 patients. The only one case with ANA negative at presentation was SMA positive. 22 patients were examined with SMA and only 10% (2/22) of patients exceeded the titer of 1:40. Likewise only 18% (2/11) of patients examined with P-ANCA were positive. The majority of patients were tested with AMA and all were negative for it.

HLA-tissue typing

HLA-tissue typing was tested in 8 patients to determine the genetic status of the AIH patients, an increased tendency of HLA-A11 association (5/8 patients) and HLA-DQ5 association (5/8 patients) was noted.

Histological features

Liver biopsy was performed in 13 patients. Histological assessment was made according to the morphological criteria proposed by IAIHG. The results (Table 4) had shown that the main histological features were interface hepatitis (100%) and lymphoplasmacytic infiltration (100%).

Comparison of cirrhotic and non-cirrhotic patients

In the present study, liver cirrhosis was noted in 37% of cases, and the non-cirrhotic patients comprised 63% of cases. The clinical profile of these 2 groups of AIH patients were compared and analyzed (Table 5). It showed that no statistically significant difference was obtained with respect to age, AST, ALT, Bilirubin, Albumin, sex and treatment response. In patients with cirrhosis at entry, a significantly prolonged prothrombin time (median, 17.3s vs 12.2s, p=0.030), elevated ANA titer (mean titer, 480 vs 80, p= 0.035) and a poorer outcome in terms of mortality (p= 0.041), were observed.

Table 5. Characteristics of AIH patients with cirrhosis and without cirrhosis

	Patients with cirrhosis	Patients without cirrhosis	p-value
Age	59.4 ± 15.2	54.1 ± 15.9	0.181
	62.5 (29.0-80.0)	51.0 (17.0-80.0)	
AST (IU/L)	668.4 ± 934.2	299.8 ± 302.9	0.628
	383.4 (41.5-3090.0)	127.0 (33.5-1030.0)	
ALT (IU/L)	443.3 ± 578.2	367.3 ± 362.2	0.945
	114.2 (30.9-1490.0)	259.0 (21.5-113.0)	
Bilirubin (mg/dL)	10.8 ± 9.0	4.7 ± 4.9	0.073
	12.8 (0.6-26.4)	2.9 (0.5-16.4)	
Albumin (g/dL)	3.1 ± 0.5	4.0 ± 0.8	0.394
	3.2 (2.2-3.9)	4.1 (2.6-4.9)	
Prothrombine time	17.3 ± 2.2	14.1 ± 5.2	0.030*
	17.3 (14.0-20.3)	12.2 (10.3-26.8)	
ANA (titer)	840.0 ± 981.2	135.3 ± 148.6	0.035*
	480.0 (40.0-2560.0)	80.0 (20.0-640.0)	
Sex (M/F)	2/8	2/15	0.613
Mortality (alive/expired)	7/3	17/0	0.041**
Remission/poor response	4/2	5/2	1.000

Values are expressed as both mean ± SD and median (range), *significant (Wilcoxon rank-sum test), **significant (Fisher's exact test).
Statistical methods, Wilcoxon rank-sum test.
Fisher's exact test for sex, mortality and treatment response.

Clinical course and treatment response

13 patients received prednisolone therapy. Complete remission was noted in 8 patients and partial remission in 1 patient. The poor response group included 3 treatment failure cases and 1 relapse case. The clinical course of the remaining patients who were not treated by prednisolone were documented. 3 patients died due to advanced decompensated liver disease, shortly after the diagnosis of AIH. Spontaneous improvement was noted in 3 patients and 4 patients had low disease activity, in whom the aminotransferases values were well below 2-fold upper normal limit. The remaining 4 cases refused to receive prednisolone treatment. The survival analysis curve had shown that the overall survival rate was 88.9%. It is noteworthy to mention that the initial abrupt decline of the survival curve was caused by 3 cases of early death, who survived less than 20 days after the diagnosis of AIH.

Discussion

Autoimmune hepatitis (AIH) is a chronic inflammatory liver disease characterized by hypergammaglobulinemia, circulating autoantibodies, female predominance, response to immunosuppressive therapy and association with an immunogenetic background[1-2]. The disease is not easy to diagnose, as there is no single diagnostic test for it, and so it is easily ignored by the clinicians. AIH was previously thought to be rare in Taiwan, and likewise lower incidence of AIH was observed in other Asian countries such as Japan, Singapore and India[4-6], as compared to western countries. The reason of lower incidence of AIH in Asian countries is not fully understood, it may be attributed to genetic and geographic factor, or an underestimation of cases.

In 1993, the International Autoimmune Hepatitis Group (IAIHG) had proposed a diagnostic scoring system for AIH, and later revised in 1999 to improve its specificity[7,1]. The proposed criteria and scoring system not only provides an objective mean to select relatively homogeneous groups of patients for research purposes, it is also an effective method to assist clinicians in confirming the diagnosis of AIH. We had used the revised criteria of IAIHG published in 1999 (the Chicago report) for the diagnosis of AIH patients, and had identified 27 cases of AIH in the period of 3 years. We had thus shown that the AIH incidence in Taiwan is much higher than previously thought.

In common with other reported series[4,9], female predominance was noticed in our AIH patients. The median age at diagnosis was 58 years, and most of the cases (19/27) were found to be over 50 years of age, which is compatible to some recently reported series[4-5,10]. The most common clinical features were fatigue, jaundice and abdominal fullness which are as usual. An insidious onset of disease had been reported in the earlier literature, and is a common finding of most authors[9,1]. On the contrary, Parker et al had reported that the majority of their cases presented with painless acute icteric jaundice[10]. In our investigation, 37% of the cases had an acute onset versus 63% of cases with chronic insidious clinical presentation.

AIH can be subdivided serologically into AIH type Ⅰ, which is characterized by ANA and/or SMA, comprising 80% of AIH cases, and AIH type 2, characterized by LKM-1 antibodies, comprising 3-4% of cases[11-12]. The autoantibodies ANA was known to be the most important and earliest defined marker of AIH, it is of diagnostic value when a titer of greater than 1:40 is detected by immunofluorescence[13]. In the present study, AIH type Ⅰ appeared to be the determinant type of AIH in our cohort, nearly all patients (26/27) showed a positive ANA titer, with a mean titer of 1 ： 160 at initial presentation. The only case with initial ANA negative but SMA positive was later found to have positive ANA titers 6 months after the diagnosis. A low positive rate of SMA tests (2/22) was noticed in our study. High prevalent rate of SMA in AIH type Ⅰ had been reported in western countries[14]. On the other hand, low prevalence of SMA were also

reported by other Asian countries such as Japan and Singapore. It is not certain whether a racial or geographical factor existed to explain the discrepancy of SMA prevalence between the West and the East.

Newer modalities of autoantibodies more specific to liver cells, such as p-ANCA, antibodies reacting with asialoglycoprotein receptor (ASGPR) and soluble liver antigen (SLA) had been introduced and examined in recent years[15]. These antibodies were described in the Brighton Report as of relevance to AIH diagnosis[7], and high titers of p-ANCA had been documented in the sera of up to 90% of patients with AIH[16]. We had examined 11 patients for p-ANCA, and disappointedly only 2 out of 11 cases were found to be positive.

There was an emphasis of genetic predisposition factor associated with AIH[17]. Increased frequency of HLA-B8 and HLA-DR3 associated with AIH were reported among white patients[18], and in Japan they had reported a high frequency of HLA-DR4 and HLA-BW54[19]. No HLA-typing data of Taiwanese patients was reported previously. In the present study, we had performed HLA-typing for A, B, C, DR and DQ loci in 8 patients, and had found that 5 out of 8 patients presented with HLA-A11 and also 5 out of 8 patients presented with HLA-DQ5. Conclusion cannot be made due to only small number of patients studied.

Hypergammaglobulinemia is one of the key factor in determining AIH. Increased serum globulin with selective elevation of IgG is regarded as a characteristic feature[1,7], and is essential for definite diagnosis of type Ⅰ AIH[20]. In our series, raised levels of IgG and serum globulin were observed in 61% and 77% of the AIH patients respectively. The relatively low percentage (61%) of IgG elevation in our series could be explained in parts by the fact that among the 7 cases of AIH patients with lower level of IgG, 3 of them had low disease activity and 1 case had elevated value later in the course of the disease. For the remaining 3 cases, no specific reason could be given for their low level of IgG value. The three leading abnormal laboratory tests in this series were abnormal AST (93%), ALT (93%) and bilirubin (71%). This is comparable to most reported series in which a "hepatitic" pattern of serum biochemical abnormalities is frequently encountered.

It is noteworthy that laboratory data implicating hepatic dysfunction, namely increased bilirubin level, decreased albumin level and prolonged prothrombin time were noted in 71%, 64% and 63% of our patients. This denoted that a substantial proportion of our AIH patients had poor hepatic function at presentation. One of the reasons that might explain this finding is the large fraction of cirrhotic patients (37%) in this series. The prevalence of liver cirrhosis was reported in other series in the range of 25-34%[10,20-21]. Low prevalent rate of 12-14% were recently reported by the Japanese authors[4,22], which may be attributed to predominance of HLA-DR4 allotype associated with Japanese AIH patients. AIH patients with HLA-DR4 allotype had previously been reported to show milder disease as compared to those who have HLA-DR3[15]. In the present study, 37% of patients had liver cirrhosis at entry. We had found that patients with liver cirrhosis had a significantly prolonged prothrombin time ($p= 0.030$) and poorer outcome in terms of mortality ($p= 0.041$). The ANA titer was also found to be significantly elevated in patients presenting with cirrhosis ($p= 0.035$) (Table 5). In patients with liver cirrhosis, the aminotransferases and bilirubin levels tended to be elevated, but did not reach the statistical significant level (Table 5). Most importantly, no significant difference was observed in terms of responsiveness to corticosteroid therapy in both the cirrhotic and non-cirrhotic groups (Table 5). This is in consistence with the work of Roberts SK et al [21]. who specified that the presence of liver cirrhosis at presentation does not affect responsiveness to corticosteroid treatment and that they warrant a similar therapeutic option as those AIH patients without cirrhosis.

We have classified the histological pattern of AIH patients according to the revised criteria recom-

mended by the IAIHG, 1999. The most frequent histological features were interface hepatitis and predominant lymphoplasmacytic infiltration.

We had only treated half of the cases with immunosuppressive therapy, the remaining cases were not treated with prednisolone due to early deaths, low disease activity and refusal of therapy. A favorable response to low dose prednisolone treatment was observed, but the time of follow up was short in this series.

The overall survival rate in the present series is 88.9%. It should be noted that the three cases of early mortality were due to severe decompensated liver cirrhosis, survived not more than 20 days after the diagnosis of AIH. If these 3 cases were not counted, a favorable survival curve was observed. All patients survived through 15 days to 824 days, although the time of follow up was too short to confer a significant conclusion in this aspect.

In conclusion, by using the diagnostic criteria of IAIHG, we had identified and diagnosed more cases of AIH, and thus had shown that the AIH incidence in Taiwan is much higher than previously presumed. AIH type Ⅰ is the predominant type of AIH, and in common with other reports, female predominance, an insidious disease onset and a "hepatitic" pattern of clinical manifestations were observed. In addition, a substantial proportion of patients presented with poor hepatic function at entry. 37% of patients had liver cirrhosis at entry and they had displayed a poorer clinical course. As a whole, there was a favorable treatment response in our AIH patients.

Reference

1.International Autoimmune Hepatitis Group report. Review of criteria for diagnosis of autoimmune hepatitis. J Hepatol 1999; 31: 929-38.

2.International Working Party. Terminology of chronic hepatitis. Am J Gastroenterol 1995; 90: 181-89.

3.Boberg KM, Adland E, Jahnsen J, et al. Incidence and prevalence of primary biliary cirrhosis, primary sclerosing cholangitis, and autoimmune hepatitis in a Norwegian population. Scand J Gastroenterol 1998; 33: 99-103.

4.Toda G, Zeniya M, Watanabe F, et al. Present status of autoimmune hepatitis in Japan — correlating the characteristics with international criteria in an area with a high rate of HCV infection. J Hepatol 1997; 26: 1207-12.

5.Lee YM, Teo EK, NG TM, et al. Autoimmune hepatitis in Singapore: a rare syndrome affecting middle-aged women. J Gastroenterol Hepatol 2001;16, 1384-89.

6.Gupta R, Agarwal SR, Jain M, et al. Autoimmune hepatitis in the Indian subcontinent: 7 years experience. J Gastroenterol Hepatol 2001;16, 1144-48.

7.Johnson PJ, Mcfarlane IG. Meeting report: International Autoimmune Hepatitis Group. Hepatology 1993; 18: 998-1005.

8.Czaja AJ, Carpenter HA. Validation of scoring system for diagnosis of autoimmune hepatitis. Dig Dis Sci 1996; 41: 305-14.

9.Sherlock S, Dooley J. Chronic autoimmune hepatitis. In: Diseases of the Liver and Biliary System. Oxford: Blackwell Science 1993: 299-306.

10.Parker DR, Kingham JGC. Type 1 Autoimmune hepatitis is primarily a disease of later life. QJ Med 1997; 90: 289-96.

11.Czaja AJ, Mann MP. The validity and importance of subtypes of autoimmune hepatitis: a point of view. Am J Gastroenterol 1995; 90: 1206-11.

12.Homberg JC, Abuaf N, Bernard O, et al. Chronic active hepatitis associated with anti-liver/kidney microsome type 1: a second type of "autoimmune" hepatitis. Hepatology 1987; 7: 1333-39.

13.Manns MP. Recent developments in autoimmune liver diseases. J Gastroenterol Hepatol 1997; 12(suppl) s256-71.

14.Whittingham S, Mackay IR, Irwin J. Autoimmune hepatitis: immunofluorescence reactions with cytoplasm of smooth muscle and glomerular cells. Lancet 1966; 1: 1333-5.

15.Mcfarlane IG. The relationship between autoimmune markers and different clinical syndromes in autoimmune hepatitis. Gut 1998; 42: 599-602.

16.Targan SR, Landers C, Vidrich A, et al. High-titer antineutrophil cytoplasmic antibodies in type-1 autoimmune hepatitis. Gastroenterology 1995; 108: 1159-66.

17.Obermayer-Straub P, Strassburg CP, Manns MP. Autoimmune hepatitis. J Hepatol 2000; 32(suppl 1): 181-97.

18.Donaldson PT, Doherty DG, Hayllar KM, et al. Susceptibility to autoimmune chronic active hepatitis: human leukocyte antigens DR4 and A1-B8-DR3 are independent risk factors. Hepatology 1991; 13: 701-6.

19.Seki T, Kiyosawa K, Inoko H, et al. Association of autoimmune hepatitis with HLA-Bw54 and DR4 in Japanese patients. Hepatology 1990; 12: 1300-4.

20.Czaja AJ. Diagnosis and therapy of autoimmune liver disease. Med Clin North Am 1996; 80: 973-94.

21.Roberts SK, Therneau TM, Czaja AJ. Prognosis of histological cirrhosis in type Ⅰ autoimmune hepatitis. Gastroenterology 1996; 110: 848-57.

22.Omagari K, Kinoshita H, Kato Y, et al. Clinical features of 89 patients with autoimmune hepatitis in Nagasaki Prefecture, Japan. J Gastroenterol 1999; 34: 221-6.

自體免疫性肝炎的臨床研究

郭樂明　林靖南[1]

台南奇美醫學中心 內科部　胃腸肝膽科 病理部[1]

摘　要

自體免疫性肝炎(Autoimmune hepatitis)(AIH)是一種少見的肝臟慢性炎症，尤其在台灣更爲罕見，之前國內文獻記載病例數目不多。AIH 的診斷不易，因爲缺乏簡單及有效的診斷方法，自從 International Autoimmune Hepatitis Group (IAIHG)於1993 年推薦 AIH 的診斷評分制度(diagnostic criteria and scoring system)，復於1999 年經過修正後，診斷 AIH 變爲更有效及平易，奇美醫院憑藉此評分制度於最近三年診斷了27 例的 AIH 病例，今提出此臨床研究報告。自2000 年到2002 年9 月，共收集27 例診斷爲 AIH 病例，並對病人之詳細病史(包括疾病表現、B 型及 C 型肝炎、酒精、藥物以及家族史)，臨床症狀，類固醇治療，預後以及存活率進行詳盡的調查及分析。憑藉IAIHG 的評分制度，15 分以上歸類爲 Definite AIH ，10-15 分歸類爲 Probable AIH ，兩組病人都列入此研究。其中又對有肝硬化及無肝硬化組進行比較分析。病人臨床數據皆用平均值及中位數表示，統計方法採用 Fisher's exact test ，Wilcoxon Rank Sum test 及 Kaplan-Meier curve 。27 位病人被診斷爲 AIH type 1 ，Definite AIH 13 人，Probable AIH 14 人，平均年齡爲58 歲，男女比例爲 4 ： 23 。63% 病人以漸進性發作表現，37% 病人爲急性發作，最常見的臨床症狀依序爲倦怠 (56%)、黃疸 (52%)、及腹脹 (52%)。96% 病人 ANA(antinuclear antibodies) positive ，檢驗結果異常依序爲 AST (93%)、ALT (93%)、serum globulin (77%)，bilirubin (71%)，病人呈現嚴重肝功能異常者 (bilirubin 增加，albumin 降低，prothrombin time 延長) 比例偏高。37% 的 AIH 病人呈現肝硬化，與非肝硬化組比較，統計顯示 prolonged prothrombin time (17.3s vs 12.2s ，P = 0.030)，poorer outcome (P = 0.041) 及 ANA titer 偏高 (480 倍 vs 80 倍 P = 0.035) 爲有意義差別。13 位病人接受肝組織切片檢查，最常見病理表現爲 Interface hepatitis (100%) 及 Lymphoplasmacytic cells infiltration (100%)。部份病人接受類固醇治療，反應良好。值得一提爲肝硬化及非肝硬化病人對類固醇治療反應並無多大差異。整體存活率爲 88.9% 。本研究顯示自體免疫性肝炎 (AIH) 在台灣的發生率比以往所認知的高很多。絕大多數病人屬於 AIH type 1 。雖然疾病呈現期肝機能損害偏於嚴重，總體而言，此類病人的預後尚佳。