

자가면역성 간염

- 최근의 국내 추세를 중심으로

Autoimmune Hepatitis ; Recent Korean Trend

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Abstract

Autoimmune hepatitis is defined as an unresolving, predominantly periportal hepatitis of unknown etiology, usually with hypergammaglobulinemia and tissue autoantibodies, which is responsive to immunosuppressive therapy. Clinical manifestations, diagnostic criteria, differential diagnosis and treatment guideline have been described. In Korea the prevalence of autoimmune hepatitis seemed to be lower than that in western countries, but clinical features were similar, except for more severe female preponderance. According to recent clinical survey, mean age is 47.8, and MF ratio is 9:1. Mostly type 1, and had cirrhosis in 22% at the time of diagnosis. 16% of patients had associated immunologic disorders. The diagnosis is definite is 47.3%, and probable in 51.3%. 72% were treated with immunosuppressive therapy with remission rate of 70%. To understand the clinical features of autoimmune hepatitis adequately, nationwide prospective clinical and epidemiological studies are needed urgently.

Keywords : Autoimmune hepatitis;
Clinical manifestation; Diagnosis;
Treatment; Korean trend

핵심용어: 자가면역성 간염 ; 진단 ; 치료 ; 한국 추勢

자가면역성 간염 (autoimmune hepatitis)
(periportal hepatitis)

최근 국내 자가면역성 간염의 추세를 중심으로
자가면역성 간염 (autoimmune hepatitis)
(periportal hepatitis)의 진단, 치료, 한국 추세를
중심으로
(1).
2.6~5.9%가
자가면역성 간염, Wilson
(2).
B, C, D, alpha-1-antitrypsin, Wilson,
자가면역성 간염, Wilson,
(1)(3).
자가면역성 간염

1.

: B, C, D, NBNC

: Wilson, alpha - 1 - antitrypsin

가

(cryptogenic)

2. 가

- ()

- AST/ALT, Bilirubin
ALP

- , IgG

- copper, ceruloplasmin
alpha - 1 - antitrypsin

- ANA, SMA, anti - LKM1

- AMA

-

- , (가)

- ,

- (interface hepatitis)

가

가

(1:4) 10 70

40~60 가

30~40%

, 30%

가

가 1 (, anti

smooth - muscle antibody) 2 (anti - LKM1)

(5). 1

, 2

. 2 1

가

(antinuclear antibody) anti - LKM1(1

) 가 . 가

(40%),

(5).

가 10

가

Wilson

2 ,

가

(bridging necrosis)

(multiacinar col-

lapse)

(interface hepatitis)

(4).

(6).

3. 가				가	
		1	2	(primary	
가 ()		ANA (?)	anti - LKM1(Cyt P450 2D6)	biliary cirrhosis)	
		ASMA (actin)		(overlap syndrome)	
가		p - ANCA	anti - LC1	가	
		anti - ASGPR	anti - ASGPR	가 (7).	
			2~14	가	
				가	
			1	가	
HLA		B1 B8 DR3 DR4	B14 DR3 C4AQO	anti - SLA/LP	
		+++	+	가	
		+++	++	가	
		45%	82%	IgG	

가

가 , rosetting

가 , A, B, C 가

가 (6). 가 (8). A, B,

6 C ,

가 가

가 가

가 , anti - LKM1, (antimito- 가 (9).

chondrial antibody, AMA) 가 , 가

가 가 HLA 가 ,

가 가 . AMA

가 (10).

4. 가

Catetory	Factor	Score	Category	Factor	Score
Gender			Concurrent immune disease		
	Female	+ 2	Other autoantibodies	Any nonhepatic disease of an	+2
Alk Phos : AST(or ALT) ratio	>3	- 2		immune nature	+2
	<1.5	+ 2	Histologic features	Anti - SLA/LP, actin, LC1, pANCA	
- globulin or IgG	>2.0	+ 3			+3
(times above upper limit of normal)	1.5~2.0	+ 2		Interface hepatitis	+1
	1.0~1.5	+ 1		Plasma cells	+1
	<1.0	0		Rosettes	-5
				Non of above	-3
				Biliary changes	-3
				Atypical feature	+1
ANA, SMA, or anti - LKM1 titers	>1: 80	+ 3	HLA	DR3 or DR4	
	1: 80	+ 2			
	1: 40	+ 1			
	<1: 40	0			
	Positive	- 4	Treatment response		+2
AMA				Remission alone	+3
				Remission with relapse	
Viral markers of active infection	Positive	- 3			
	Negative	+ 3			
Hepatotoxic drugs	Yes	- 4	Pretreatment score		
	No	+ 1	Definite diagnosis		>15
			Probable diagnosis		10~15
Alcohol	<25 g/d	+ 2	Posttreatment score		
	>60 g/d	- 2	Definite diagnosis		>17
			Probable diagnosis		12~17

ALP/AST HLA

AMA , , 가 (4). 가

가 가 (variant)

(11). 가 (5), 가 가

가 가 (overlap syndrome),

(PBC) AMA ANA

(cryptogenic hepatitis) 가 (autoimmune cholangitis;

가 가 outlier syndrome)

(12). p - ANCA, anti SLA/LP 가

5. 가		가	
Outlier	AIH + PBC	,	가 2
	AIH + PSC	,	,
	AIH + AIC	,	,
	AIC	,	,
	AIH PBC	,	가 10
	AIH PSC	,	가 5
AIH; autoimmune hepatitis, 가 PBC; primary biliary cirrhosis, PSC; primary sclerosing cholangitis, AIC; autoimmune cholangitis, 가		가 2 , 가 , 가 (1). (sequential syndrome) , (13). 가 가 , 가 가 가 (systemic lupus erythemato- sis, SLE) APS - 1(autoimmune polyglandular syndrome type 1) 가 (11). 가 Prednisone(PD) prednisone azathio- prine(AZA) (6), 65% , , 가 (6, 14). (incomplete response) 가, , , , , PD , , thiopurine methyltransferase(TMT) , AZA . (6, 14). PD (treatment failure) 가 (6, 14).	

6. 가

		Contraindication	
Prednisone(mg)	30mg · 1		PD (
	20mg · 1	TMT	5
	15mg · 2) , AZA
	10mg		(2mg/kg) (6, 14).
Azathioprine(mg)	50mg		
Prednisone(mg)	60mg · 1		
	40mg · 1	emotional lability	PD (2
	30mg · 2	brittle DM, labile HT) ,
	20mg	, acne	

*TMT; thiopurine methyltransferase

(6 98%).

가 67%

(PD 60mg/D, PD 30mg/D (15).

+ AZA 150mg/D) (1) , , , ,

(PD 10mg/M, AZA 50mg/M) , ,

(6,

14). (14).

, , , 83~92%, 10 75%

, brittle DM, , (16, 17).

.

PD 가 PD

AZA 가 AZA 100mg/D 2 (14, 18).

.

가 가

3 6

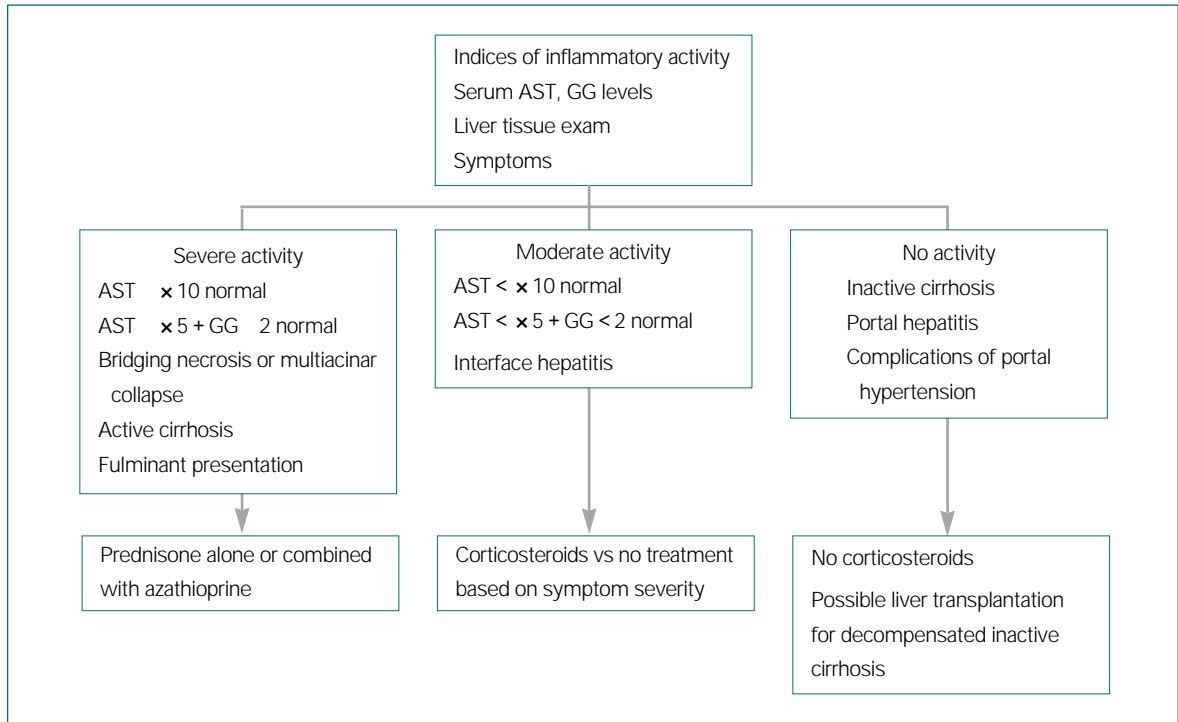
(6, 14). 가 1986 (19)

(6 49%), 20

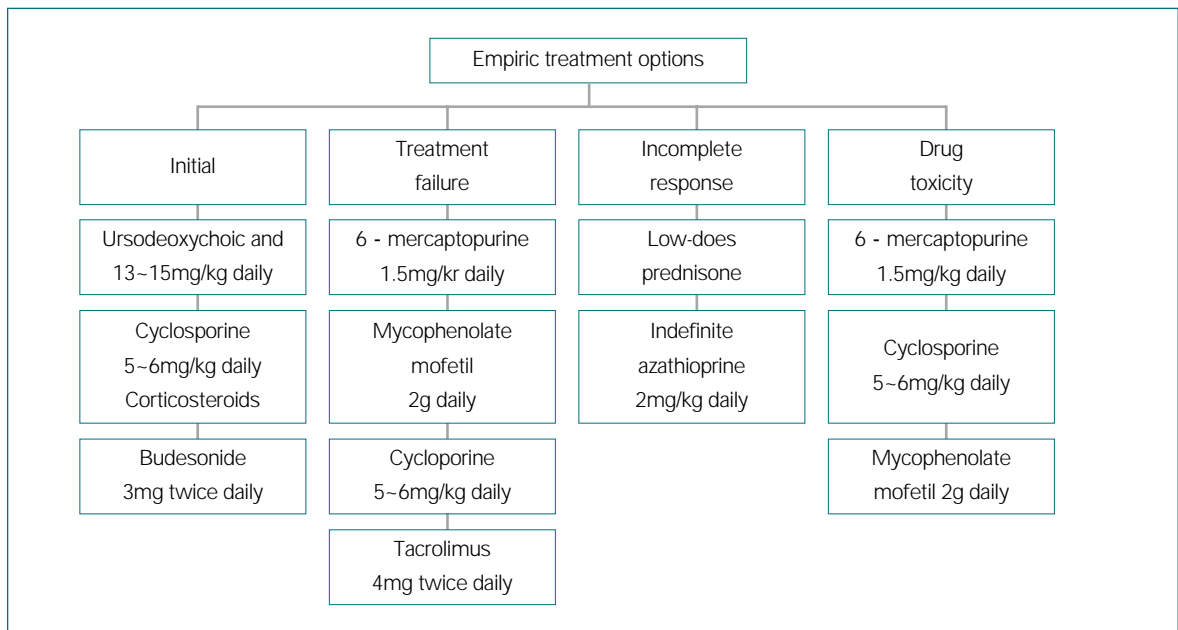
. 1 가 10% 2004 115 가 (5), 2002

. 가 3 가 2004 가 10

, 172 (5). 10



1. 가



2. 가

1999 50 50 (20)

가 6 가 가

(2), 10 0.1 ~ 1.9 ,

, 10 8.0 ~ 16.9 .

(21) . ,

가

가 가 (2, 20) 가

가 가 가

가 , 가

2002 ~ 2004

(20) 가 가 .

47.8 .

90% .

가 22% . 2

(anti LKM1) 1 1 (

) . 16%

, , , ,

, , Sjogren syndrome,

, Raynaud disease,

, IgA nephropathy,

, 51.3%가

‘probable’, 47.3%가

72% 가 46%가

, 54%가

, 70%

, , HLA

가

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