Late diagnosis of autoimmune hepatitis resulting in severe hepatic failure: Case report

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Introduction

Autoimmune hepatitis (AIH) is a disease that affects the liver diffusely and still doesn't have a defined etiology. Its symptoms are numerous and they don't distinguish by gender or age (1). For the diagnosis it is necessary to analyze the clinical condition of the patient, laboratory tests (biochemical and serological), and to rule out other liver diseases as viral and drug-induced hepatitis, among others (2).

On this, serological tests are essential to conclude the case, once it's according to whether or not they react, that the illness can be classified into subtypes 1 or 2 (3). Usually, the prognosis is good, as almost 80% of the patients present fully remission. The treatment consists in immunosuppressants (corticosteroids and azathioprine). Despite this, relapses can occur in nearly 50% of the cases (2).

The aim of this article is to report an AIH case in a 49 year old female patient, with the purpose of emphasizing the clinical and laboratory aspects, which are important for diagnosis, treatment and prognosis. This is necessary to present disease that, despite its low incidence rates, occurs in a chronic and, occasionally, fulminating manner.

Case description

Female patient, 49 year old, born in Coelho Neto – Brazil, living in Rio Verde – Brazil for 7 years. Previously healthy, she didn't take drugs chronically and denied previous surgeries or blood transfusion. Her main complaint was postprandial fullness for a year, being submitted to several upper digestive endoscopies that verified gastritis, being treated with Proton-pump Inhibitors, without improvement. She also reported weakness and pruritus for over a month, acholic feces for two months and choluria for a year; and denied nausea, vomiting and hematemesis.

She presented icteric, asthenic and with anasarca; had an abdomen flat and flaccid; her liver was palpable 10 cm from the right costal border and the spleen 4 cm from the left one.

The patient informed that she had been submitted to several abdominal computed tomography scan, nuclear magnetic resonance, abdominal ultrasound, all of them showing the increased spleen and liver, without finding a probable cause.

On this, new laboratory tests, including serology for some viruses and antibodies, were requested. The results are shown in Table 1.

Table 1. Tests results

Tests	Results	References
Alanine aminotransferase (ALT)	96 U/L	<41 U/L
Aspartate aminotransferase (AST)	242 U/L	<40 U/L
Prothrombin time (PT)	25.00s 23% activity	70-120% activity
Activated partial thromboplastin time	60s	26.7-37.6s
Alkalinephosphatase	446 U/L	Women: <105 U/L
Gamma-glutamyltransferase (γGT)	111 U/L	Women: <38 U/L
Bilirubin	Total bilirubin (TB): 13.48mg/dL Direct bilirubin (DB): 7.93mg/dL Indirect bilirubin (IB): 5.55mg/dL	TB: until 1.00mg/dL DB: until 0.20mg/dL IB: until 0.80 mg/dL
Calcium	6.7mg/dL	8.3-11mg/dL
Creatinine	0.8mg/dL	Women: 0.4-1.1 mg/dL
Urea	14mg/dL	15-39mg/dL
Serumiron	134µg/dL	Women: 145µg/dL
Serumferritin	192µg/dL	Women 20-200µg/dL
Serummagnesium	1.67mg/dL	1.5-2.5mg/dL
Anti-HIV1/2 antibody	Non-reactive	Non-reactive
Anti HBc IgG/IgM	Negative	Negative
HBSag	Non-reactive	Non-reactive
Anti HCV	Non-reactive	Non-reactive
Anti HAV IgG/IgM	Non-reactive	Non-reactive
Anti-smooth muscle antibody (ASMA)	1:80	Negative
Anti-liver/kidney microsomal antibody type 1 (anti-LKM-1)	Negative	Negative
Antinuclear fator (ANF)	Nuclear: reactor (1:160)	Non reactive
Anti-liver cytosol type 1 antibody (anti-LC1)	Negative	Negative
Anti-soluble liver antigen antibody	Negative	Negative
Anti-neutrophil cytoplasmic antibody	pANCA: non-reactive cANCA: non-reactive	pANCA: non-reactive cANCA: non-reactive
Proteinelectrophoresis	Albumin:1.94g/dL Total protein: 7.80g/dL γglobulin: 3.83g/dL	Albumin: 3.5-4.85/dL Total protein: 6.6 – 8.3g/dL γ globulin: 0.74 – 1.75 g/dL

Therefore, the AIH type 1 was detected. It wasn't possible to perform the histological examination from a diagnostic laparoscopy, as the patient didn't have conditions to perform the procedure, even after administrating fresh frozen plasma four times a day and replacement of vitamin K for three days.

A seven-week immunosuppressive treatment regimen was installed, and weakly weaning of corticosteroids was performed until maintenance treatment. The schedule can be seen in Table 2 (2). In addition, spironolactone 50 mg, furosemide 40mg and Lanzoprazol 30 mg were prescribed.

Table 2. Treatment scheme

Time in weeks	Prednisone	Azathioprine
1st	50mg/day	50-150mg/day
2nd	40mg/day	
3rd	30mg/day	
4th	20mg/day	
5th	15mg/day	
6th	12.5mg/day	
7th	10mg/day	
8th	Maintenance	Maintenance

Discussion

It is believed that the AIH is the result of an interaction between individual genetic factors and environmental ones, affecting the liver chronically, creating areas of necrosis. There is a predominance in the female sex, agreeing with the reported case, and also observed in a survey with 268 patients attended at the Hospital das Clínicas of Medical School of University of São Paulo – Brazil, showing a proportion of 5.7:1 (4).

According to the patient's condition added to the laboratory results showing significant increases of aminotransferases, γGT, alkaline phosphatase and bilirubin, and the positive results for some antibodies, it is understood that the patient had a severe hepatic failure progressing with hepatic cirrhosis and splenomegaly, both inserted in a diagnosis of AIH. In view of the observations made, the Simplified Scoring for AIH observed in Table 3 was applied. It is worth noticing that it was not possible to perform the liver biopsy for histological analysis (3).

Table 3. Simplified Scoring for Autoimmune Hepatitis

Category	Variable	Score
ANF	1:40	+1
ASMA	≥1:80	+2
Anti LKM-1	≥1:40	+2
Anti-soluble liver antigen antibody	Positive	+2
Immunoglobulin G	>Upper normal limit	+1
	>1.1 times upper normal limit	+2
Liverhistology	Compatible with AIH	+1
	Typical AIH	+2
Absence of viral hepatitis	Yes	+2
Definitive diagnosis		*≥7*
Probable diagnosis		*6*

In view of the antibodies found, it was possible to differentiate between the types of AIH and initiate the treatment. The presence of ANF and ASMA, and the absence of the anti LKM-1, corroborate for the diagnosis of AIH type 1 (3).

After two months of treatment, the patient's improved her hepatic failure condition, reducing the levels of transaminases, bilirubin, alkaline phosphatase and γGT, and improving the levels of total proteins and albumin.

In conclusion, the fact that there was a patient, who sought diagnosis to alleviate her symptoms for a year, characterizes the failure in making early and correct diagnosis. This delay culminated in the patient's progression to a hepatic insufficiency. With this case report, we aimed at exemplifing pathologies that need further investigation and a more arduous study to make an early diagnosis, thus reducing the suffering of patients who "pilgrim" for months in several medical services searching a cure for their symptoms.

Conflicts of interest: None declared.

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