

With the colaboration

Of

& Dr IMChierra Laso



# **CAUSE OF ADMISSION**: 73years old woman with general syndrome.

**PERSONAL BACKGROUND:** 

- No known drug allergies.
- No toxic habits.
- Hypertension since the last 13 years with a good ambulatory control. No DM. Dyslipidemia.
- Deafness .
- Cronic hepatopathy of no filiated cause since the last 15 years (probable hepatitis autoinmune type I (diagnosis based on antinuclear antibodies and smooth-muscle antibodies, no biopsy)).
- Gallstones.
- Previous surgical interventions: Cataracts, left knee prosthesis and curettage.
- No habitual treatment.

### **CURRENT DISEASE**

- Constitutional symdrome since 2 months beside an abdominal pain that it begins at dorsal region and it radiates forward in belt, without relation with intake and meals and it doesn't changes after deposition.
- Weight loss of 9 Kgs and an apparent asthenia.
- No fever and no distermic sensation.
- Hiporexia, no nauseas, no vomiting. No digestive rythm alteration.
- No other concomitant syntomathology.

### **Physical Examination**

- BP 125/75. HR 80 lpm.BR 15 rpm. SatO2 96%. Asthenic habit.
- No adenopathies, no goiter, normal JVP.
- <u>Pulmonary auscultation</u>: breath sounds without superinduced pathologic sounds.
- <u>Cardiac auscultation:</u> CsSsRs without extratones.
- <u>Abdomen:</u> soft, no painful. Hepatomegaly of 4 cms below the right costal margin.
- <u>Lower limbs:</u> No edemas, pedal pulses present. Homans negative.

### **COMPLEMENTARY TESTS**

- Blood count: normal. ESR 120 mm/h.
- Coagulation: No alterations.
- Biochemistry: Renal function and ionogram within normal limits. AST 39 mU/mL, ALT 48mU/mL, PA 317 mU/mL, CRP 3.4 mg/dL, BR 1 mg/L, the rest of parameters were normal.
- <u>EKG:</u> Sinus Rhythm 75 lpm. No alterations of the repolarization.
- <u>Chest X-Ray</u>: No significant alterations.
- Abdominal X-Ray: Non specific alterations. Nonspecific gas pattern.



#### **ANALYSIS OF THE CASE**

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CLL

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informations,

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#### $\bigcirc$ 76 years old

#### -BACKGROUND:

- Deafness.
- Cronic liver disease (probable autoinmune
- hepatitis with ANA + & ASMA +).
  - HTA. DL. Gallstones.

#### **PRESENT ILLNESS:**

- Constitutional syndrome since 2 months
- + abdominal pain.
- Loss of weigh 9kgs. Hiporexia +asthenia.

#### **PHYSICAL EXAMINATION:**

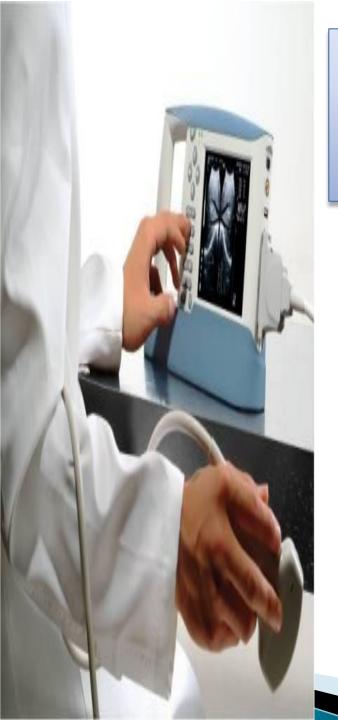
To highlight only a non painful hepatomegaly.

#### LABORATORY FINDINGS:

- -APR: ESR 120. RCP 3.4.
- Minimun alterations liver function.



#### WHAT WOULD YOU DO NEXT??? WHICH PROCEDURES WOULD YOU ASK FOR??



### ABDOMINAL ULTRASOUNDS

• Hepatomegaly with big spaceoccupying lesiones. Gallstones.







#### - HEPATIC POLILOBULATED MASS OF 12 CMS OF DIAMETER.

- Left subserosic leiomioma.



#### DIFERENTIAL DIAGNOSIS HEPATIC MASS

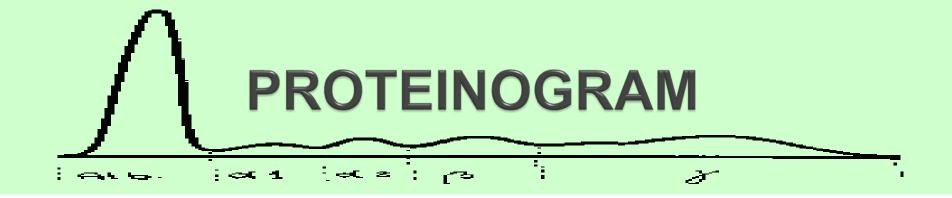
<b>BENIG LESION</b>	MA LIGN LESION	INFECTIONS	MISCELLANY
<ul> <li>-Adenoma,</li> <li>Hemangioma</li> <li>⇒Different</li> <li>radiologic behavoir</li> <li>in ecography &amp; CT.</li> <li>-Hepatic</li> <li>regenerations</li> <li>nodules (against the size, no biopsy, no data in favor on cirrhosis).</li> </ul>	-Metastasis. -Hepatocellular carcinoma. - Lymphoma. -Others - The size, radiology and the agresivity of the mass.	<ul> <li>-Piogenic,</li> <li>Candidiasic/</li> <li>Amebian abscess)</li> <li>=&gt; No fever, no</li> <li>leucocitosis ni</li> <li>neutrophilia,</li> <li>radiology (no</li> <li>acustic</li> <li>reinforcement).</li> <li>-Actinomicosis,</li> <li>Hidatidosis abscess.</li> <li>Tuberculosis.</li> </ul>	<ul> <li>-Hepatic artery aneurism (PAN, traumatism).</li> <li>- Granulomatosis illnesses.</li> <li>- Peliosis.</li> <li>- Hematoma.</li> </ul>



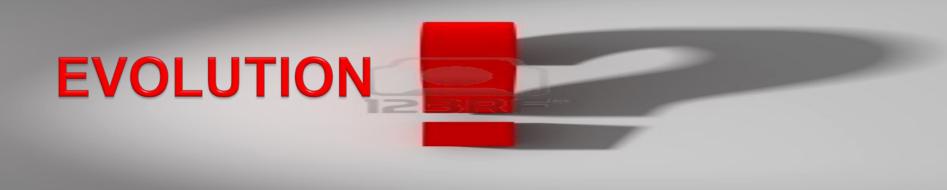
- **COMPLETE BLOOD ANALYSIS:** 
  - Ferric serius: normal. Tyroides hormons: normal.
  - Tumoral markers:
    - CEA, alfa-fetoprotein: negatives.
    - Beta-2 microglobulin: 2.6 mg/L (N  $\leq$  2.2mg/L).
    - Beta-HCG: 20 mUI/mL (N < 5mUI/mL).

#### AUTOINMUNITY

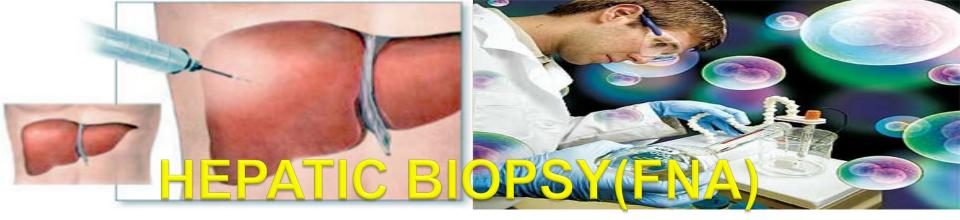
- RF, ANA, antiDNA, ANCA, antiLKM, antimitocondrial antibodies: NEGATIVES.
- Antirreticulin antibodies that interferes with smoth muscle antibodies.



- Gammaglobulin 1.3 g/L. Band of monoclonal aspect.
- Inmunofijation shows one monoclonal protein band IgM kappa.
- Orine inmunofijation: negative.



- No fever, good control of pain with habitual endovenosus analgesic.
- After reevaluating the case, the patient has been previously followed-up by Digestive Unit and she had absolutely denied to do an hepatic biopsy.
- Analitically she only had a minimun alteration of liver function without any descompesation until the present admission. They had never put her any treatment at all.



• The anapathologic study of the sample show...

#### HEPATOCELLULAR CARCINOMA

The patient has been derived to Oncology Unit where she has been receiving Target Mollecular Therapy (Soratinib).

### **DIAGNOSTIC IMPRESSION**

#### - HEPATOCELLULAR CARCINOMA IN PATIENT WITH CRONIC AUTOINMUNE HEPATITIS TYPE I.

## **AUTOINMUNE HEPATITIS**

Classification of and autoantibodies in autoimmune hepatitis

Туре	Autoantibodies
1 (classic)	Antinuclear
	Anti-smooth muscle
	Anti-actin
	Anti-soluble liver/ liver pancreas antigen (Anti- SLA/LP)
	Atypical pANCA
	Antimitochondrial
2	Anti-LKM-1
	Anti-liver cytosol -1 (Anti-LC1)
	Anti-soluble liver/liver pancreas antigen (Anti- SLA/LP)

Chronic liver disease characterized by: autoinmunes features and hyperglobulinemia with circulating antibodies.

- Heterogeneity.
- Overlap syndrome (Primary biliary cirrhosis & Primary Sclerosing cholangitis).
- In general a "steroid-responsive" condition, with great benefits of treatment and an aproppiate manage can improve quality of life.
   Progress to cirrhosis & HCC.



### TREATMENT

- Individualized decision upon severity of symptons, degree of serum transaminases (x10) and gammaglolin elevation (not a necessarily correlation with histology injury).
- **GOLD STANDARD: Histologic study.**
- □ Uncertain benefit of treatment in inactive cirrhosis.
- **Choices of treatment:** 
  - **Prednisolone** 60mg/d (lower rate) or 30mg/d + **Azatioprime** 50mg daily.
  - **Ciclosporine** 4mg/kg/d (reasonable alternative children).
  - **Budesonide** 3mg/8-12 hours (not really extensively studied).

# MESSAGES TO REMEMBER

- The carcinoma hepatocellular is one long-term complication in autoinmune hepatitis, specially if they are not treated.
- The need of biopsy for an appropriate manage.
- Our clinical practice is limited by patients's decission, althoutgh sometimes they aren't correct (ethical problem).

