



# PReS Latin America Basic Pediatric Rheumatology Course

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Case presentation by:  
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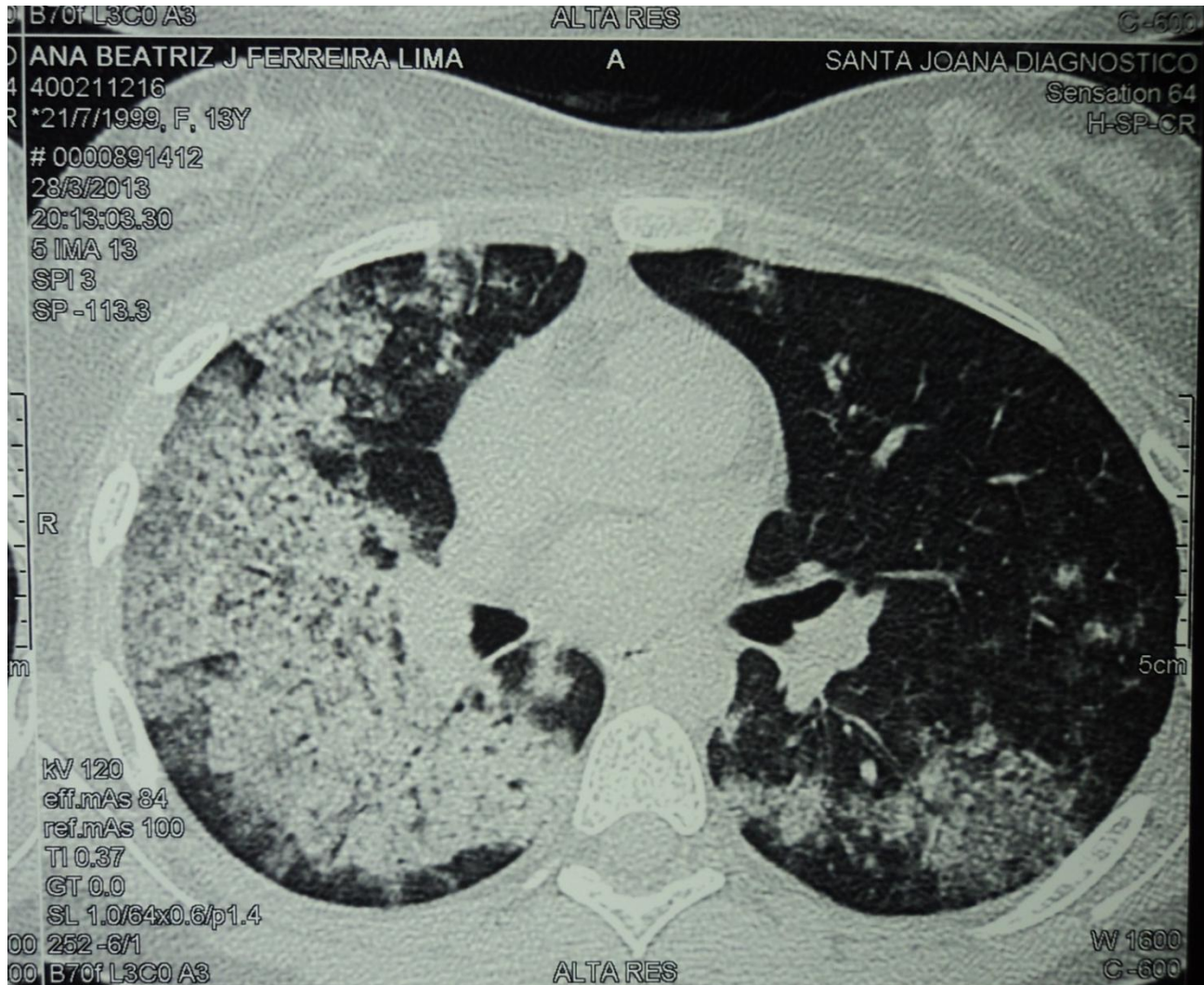
- Ana, 14 years old, presents arthritis in wrists and ankles for 3 days
- After the second day of onset, she develops palpable purpura on lower limbs and abdominal pain
- Fever  $\ominus$  Diarrhea/melena  $\ominus$  No other systemic symptoms

EXAMS	16/03/2013
Hb (g/dl)	13,4
WBC (mm3)	<b>16.210</b>
Platelets (mm3)	272.000
CRP (< 0,5 mg/dl)	<b>5.7</b>
ESR (mm/h)	<b>66</b>
ASO (UI/ml)	25
Urinalysis	<b>Blood + 4 RBC/hpf</b>

- Henoch-Schonlein Purpura (HSP)
  - Palpable purpura
  - Abdominal pain
  - Arthritis
  - Kidney involvement
- Management:
  - Naproxen
  - Abdominal US and chest X-ray
- Continues with abdominal pain and reports onset of melena, which made us start her on prednisone 2 mg/kg/day
- After 2 days with prednisone, she presented with hemoptysis

EXAMS	18/03/2013	25/03/2013
Hb (g/dl)	13,4	<b>9,1</b>
WBC (mm3)	<b>16.210</b>	<b>22.400</b>
Platelets (mm3)	272.000	382.000
CRP (< 0,5 mg/dl)	<b>5,7</b>	<b>12</b>
ESR (mm/h)	<b>66</b>	<b>51</b>
Urinalysis	<b>Blood + 4 RBC/hpf</b>	<b>Proteins ++ 35 RBC/hpf</b>

ANA: negative  
 C3/C4: normal  
**PR3-ANCA: 88 U/ml (< 2U/ml)**  
 MPO-ANCA: 0.1 U/ml (< 7U/ml)  
 Anti-GBM: negative



Chest HRCT show diffuse shadowing consistent with acute pulmonary hemorrhage.

- Pulmonary-renal syndrome: pulmonary hemorrhage and glomerulonephritis
- Methylprednisolone pulse (5 days) and cyclophosphamide iv (CYC)
- Pack red cell transfusion ( Hb= 5.7 g/dl)
- Kidney biopsy (light microscopy): 22 glomeruli, 19 presenting with segmental sclerosis and cellular crescents. Tubular atrophy and tubulointerstitial nephritis
- Direct immunofluorescence: pauci-immune pattern
- She had complete resolution of pulmonary hemorrhage and melena, however even with the use of CYC she persisted with hematuria, proteinuria and increase in Cr level

- Due to unfavorable outcome, patient was administered rituximab (375mg/m<sup>2</sup>) once a week for 4 weeks with subsequent improvement in renal function
- Currently she is taking AZA and losartan

**Rituximab**



	APR/13	MAY/13	JUN/13	AGO/13	SET/13	NOV/13	FEB/14	APR/14	JAN/15
Cr (mg/dl)	0,4	0,4	1,1	1,6	1,3	1,2	1,0	0,9	0,7
Prot. (mg)	880	1097		1900		1000	900	421	167

# QUESTIONS

- What is really the diagnosis of this patient??
- Should we request in all patient with the preliminary diagnosis of HSP an ANCA test??
- What would you have done differently in terms of treatment?