

PReS Latin America Basic Pediatric Rheumatology Course

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Aguas de São Pedro, Sao Paulo - Brazil



Case presentation by:
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- GBS, 13 years old, presents arthritis in knees and elbows for the last 4-5 weeks
- She was referred to a pediatric rheumatologist by an orthopedist
- Fever ⊖ Skin rash ⊖ Diarrhea/abdominal pain ⊖
- She had abdominal pain 6 months ago. All tests were normal and she was discharged from the pediatric gastroenterology unit
- Parents and brother are healthy

- Physical exam: arthritis in knees and elbows
- Lab tests:

	06/01/2015
Hb (g/dl)	11.5
WBC (mm ³)	18.500
Platelets (mm ³)	293.000
ESR (mm/h)	46
CRP (mg/l)	44 (<5 mg/l)
RF	negative

- Management:
 - Naproxen
 - X-ray chest, ECHOCARDIO and abdominal US
 - New tests and return in 1 week

Return after 1 week



- Mild improvement in pain
- Unchanged physical exam
- 1 feverish peak/week
- Imaging exams were normal

	06/01/2015	16/01/2015
Hb (g/dl)	11.5	11.3
WBC (mm ³)	18.500	17.400
Platelets (mm ³)	293.000	291.000
ESR (mm/h)	46	66
CRP (mg/l)	44	42
RF	negative	negative
AST/ALT (U/l)		17/12
Ferritin (ng/ml)		236 (<180)

ANA: negative
Anti-dsDNA: negative
C3/C4: normal
Urinalysis: normal

Return after 2 weeks



- No improvement in pain
- Unchanged physical exam
- 2 feverish peak/week
- We added MTX

Follow up



- No improvement with naproxen and MTX
- Unchanged physical exam
- Daily fever
- Colonoscopy was requested by Gastro-Ped

- Colono: only one polyp, remaining was normal. Performed excision of polyp + biopsy



- Biopsy:
 - Rectal inflammatory pseudo-polyp
 - Mild ileitis + minimum chronic colitis
- Pediatric gastroenterology started her on sulfasalazine + prednisone 2mg/kg
- First week, Patient was afebrile with resolution of the joint disease
- Second week, the patient returns fever and onset of skin rash (independent of fever, not fixed)



Sulfassalazine



	06/01/15	16/01/15	23/02/15	23/03/15
Hb (g/dl)	11.5	11.3	10.2	11
WBC (mm ³)	18.500	17.400	18.600	22.000
Platelets (mm ³)	293.000	291.000	401.000	125.000
ESR (mm/h)	46	66	97	48
CRP (mg/l)	44	42	95	77
RF	negative	negative		
AST/ALT (U/l)		17/12	20/14	62/97
Ferritin (ng/ml)		236 (<180)	430	10.032

- Bone marrow aspirate: several histiocytes in activity, some in hemophagocytosis
- MAS probably secondary systemic JIA ?
- She was treated with pulse methyprednisolone and cyclosporine
- Excellent response since the end of march.
- Nowadays, she is with prednisone 15mg/day and CSA 150mg/day

QUESTIONS

1. Does Gabriela really has the diagnosis of systemic JIA??
2. What is the relationship between systemic JIA and sulfasalazine??
3. And the possibility of IBD??
4. And if we had done bone marrow aspirate early, what we would find?

KEY MESSAGES

- The final diagnosis we will just know with the follow up
- Emergency MAS: early diagnosis and aggressive treatment