FEVER, RASH AND CHOLESTATIC HEPATITIS: AND WHEN THE DIAGNOSIS IS UNUSUAL?

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BACKGROUND
Common symptoms of infectious diseases like fever, rash and hepatomegaly may also be clinical features of uncommon diseases, posing a challenge to the clinician.

17 year-old girl
Léri–Weill syndrome (limb dyschondrosteosis)
Oral and vaginal ulcers
BEHÇET DISEASE
SULFASALAZINE 6 WEEKS

D1 Anorexia and asthenia
D2 HIGH FEVER (39–40°C)
D3 Vomiting
Facial rash
Right hypochondrium pain
D9 JAUNDICE AND CHOLURIA

D12 At hospital admittance
CHOLESTHATIC HEPATITIS
THROMBOCYTOPENIA (84X10^9/L)
EOSINOPHILIA (1,36X10^9/L)
LIVER + SPLEEN ENLARGEMENT
DIFFUSE PRURITIC RASH
FACIAL EDEMA

A few days after completing corticotherapy...
DISEASE RELAPSE
Fever + Rash
Eosinophilia
Cholestatic hepatitis

Corticotherapy 9 months
弥漫性血管炎综合征

10 months later...
Persistent elevation of indirect bilirubin
Suspension of prednisolone with clinical remission

Gilbert syndrome
Homzygous for TA7 mutation in the UGTA1 promoter

Eosinophilia >0,7x10^9/L

LEARNING POINTS
• DRESS syndrome is a rare but potentially fatal disease (10%), especially when liver involvement occurs.
• When important peripheral eosinophilia, multiorgan involvement and a history of recent drug exposure is present, DRESS syndrome should be suspected.
• Diagnosis can be challenging and a high suspicion index is needed. Infectious causes and autoimmune diseases must be excluded and a slow taper of corticotherapy is advisable. When the response is unfavorable, concomitant diseases should be ruled out.