Episode 6

In this week's episode of the CPSers, guest Dr. Erin Chew presents an exciting clinical unknown to Rabih!

Problem Representation
A 23-year-old woman presented with subacute Raynaud's syndrome complicated by digital ulceration, dysphagia, dry mouth/eyes, and leukopenia.

Schemas
To help reason through this case, Rabih deployed a schema for Raynaud's syndrome.

This approach uses the presence of "alarm symptoms" to help differentiate between primary (isolated) and secondary (associated with an underlying disease process) Raynaud's syndrome.

Diagnosis
High titer ANA, anti-Ro52, anti-Ro60, and anti-Scl70 suggested a diagnosis of **diffuse cutaneous systemic sclerosis**!

Teaching points

- Alarm symptoms in Raynaud's include abrupt onset, longer duration (>20 min), associated ulcerations, occurrence in men or older adults, and signs or symptoms suggestive of an autoimmune disease
- Most cases of leukopenia are caused by infection (usually sepsis), drugs, or autoimmune disease (especially lupus)
- Systemic sclerosis can present with either a "limited" or "diffuse" phenotype, with the majority of disease manifestations being related to tissue fibrosis and vascular involvement

Clinical Reasoning Pearl

Rabih reminds us to anchor ourselves in epidemiology and think about the **"base rate" of disease**, especially as we consider rarer diagnoses.

Owing to the base rate, no matter how well an obscure disease fits for a given case, making the diagnosis requires a high burden of proof, simply because the epidemiological argument for a rare diagnosis is weak.

This line of thinking is the basis diagnostic principle: *an atypical presentation of common disease is more likely than a typical presentation of a rare disease.*