PedsCases Podcast Scripts

This is a text version of a podcast from Pedscases.com on "**Systemic Lupus Erythematosus**." These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedcases.com/podcasts.

Systemic Lupus Erythematosus

Developed by Dr. Tara McGrath and Dr. Dax Rumsey for PedsCases.com.
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Introduction:

Hi, my name is Tara McGrath and I am a pediatrics resident at the University of Alberta in Edmonton, AB, Canada. This podcast was developed in conjunction with Dr. Dax Rumsey, Pediatric Rheumatologist at the University of Alberta and Stollery Children's Hospital in Edmonton, AB, Canada.

Today we will be discussing Systemic Lupus Erythematosus, "SLE" or simply "lupus".
Objectives for the SLE PedsCase:

1. Recognize the signs and symptoms suggestive of SLE
2. List the 11 classification criteria for SLE
3. Elicit a focused history for patients suspected to have SLE
4. Demonstrate a pertinent physical exam for patients suspects to have SLE
5. Discuss the investigations and management of patients with SLE

Definition:

SLE is a systemic multi-system autoimmune disease characterized by the presence of autoantibodies and multi-organ system involvement. The clinical presentation is very diverse, ranging from a mild disease characterized by rash and arthritis to a severe life-threatening disease involving one or multiple organs. SLE is characterized by flares and remissions.
SLE is more common in adults than in pediatrics. Childhood-onset SLE (onset <16 years of age) makes up approximately 15-20% of all SLE cases. It's important to know that pediatric lupus is often more severe than adult lupus: it has a more aggressive presentation and course, including more frequent renal involvement. In terms of etiology, it is suspected that lupus is due to a combination of genetic susceptibility and environmental factors, including exposure to sunlight, infections, drugs and chemicals.

The Case:

You are working with a general paediatrician in a community clinic. You encounter a 13 year-old girl of Asian descent. She describes a two month history of general malaise and decreased energy. Her mother states that her daughter has felt "feverish" on and off for the last 2 months. She used to play soccer regularly, but she has stopped participating not only because she feels "too run down" but she has also been having pain and stiffness in her left knee. Last week when she was out in the sun with her classmates she developed a rash on her cheeks.

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You think the patient might have lupus…What would your approach be to this patient??

Classification Criteria:

Because of the diverse presentations of SLE, the American College of Rheumatology devised a classification system for lupus to allow comparison between different populations and to enable research. They are not diagnostic criteria. However, the classification criteria provide an organized approach to SLE manifestations. To be classified as having SLE for research purposes, one needs to meet at least 4 of 11 ACR criteria.

11 criteria may seem like a lot, but you can use a helpful pneumonic to remember them! The pneumonic is **MD SOAP BRAIN**

M- Malar rash – this rash is a fixed erythematous rash over the malar eminences – commonly called a 'butterfly rash'. It can be flat or raised, and tends to spare the naso-labial folds
D- Discoid rash – this is a raised, scaling and scarring rash. It is extremely rare in paediatric patients
S- Serositis – literally means 'inflammation of a serous membrane'; most commonly involving the heart or lungs in SLE, i.e. pleuritis or pericarditis
O- Oral/Nasal ulcers – this ulceration is usually painless, and is often on the palate or nasal septum
A- Arthritis – this is non-erosive arthritis involving two or more peripheral joints, characterized by tenderness, decreased range of motion, and effusion.
P- Photosensitivity– rash from sun exposure (notably, sun exposure can trigger a flare of SLE and therefore sun protection is key in patients with SLE).
B – Blood abnormalities – this includes hemolytic anemia (with reticulocytosis), leukopenia, lymphopenia and thrombocytopenia.
R – Renal dysfunction – there are 6 classes of SLE nephritis and a broad range of presentation and severity, from asymptomatic to severe nephritic or nephrotic syndrome.
A – ANA positive – This is anti-nuclear antibody. Essentially all patients with SLE are ANA positive.
I – Immunologic – These are other specific antibodies, including anti-ds-DNA, anti-Smith, anticardiolipin antibodies and lupus anticoagulant.
N – Neurologic – Lupus can present with a wide array of central and/or peripheral signs and symptoms. This can be anything from headaches and difficulty concentrating to seizures and psychosis.

SLE History:

So now that you have an organized way to remember some of the main features of SLE, lets review these by outlining what we would ask our patient from the case.

HPI: General description from the patient on the course of her symptoms

"Rheumatologic Review of Systems": SLE can literally involve every body system. So, an organized approach, from head to toe, is necessary.

General: Any fever? Weight loss? Malaise?

CNS: Any headaches? Recent difficulties with concentration, school performance, depression, memory loss, hallucinations? Seizure activity?
Head and Neck: Very dry eyes and/or dry mouth (SICCA symptoms)? Nose bleeds or sores in your nose? Sores in your mouth? Any hair thinning or falling out?
Cardiorespiratory: Any chest pain (worse with deep breaths)? Shortness of breath or exercise intolerance?
GI: Any stomach pain or diarrhea (serositis in SLE can include abdominal lining)?
GU: Any blood or coke-coloured urine? Any foamy urine? Any swelling in your feet or around your eyes (nephrotic syndrome)? Any headache or visions changes consistent with hypertension?
MSK: Any joint pain, swelling or tenderness (associated morning stiffness?)? Any muscle weakness or pain?
Derm: Any colour changes from white-blue-red in your hands or feet consistent with Raynauds phenomenon? Any rashes (specifically after being out in the sun)?

As in all cases, a thorough Pediatric History should include:

- Medications (some medications can trigger Drug-induced lupus)
- Allergies
- Immunizations
- Developmental History
- Past Medical History
- Family History (often other autoimmune diseases in the family)
- Social History (including HEAADS in adolescent history)

So far your history is consistent with many features of SLE, so you move onto your physical exam…

SLE Physical Exam:

- Vitals (?tachypnea, ?hypertension?)
- Alopecia?
- Facial rash or edema?
- Oral or nasal ulcers? (Make sure you look on the palate and in the nose because the patient will often not even realize that they have them.)
- Palpate for lymphadenopathy
- Thorough respiratory and cardiovascular exam
- Thorough GI exam
- Complete detailed MSK and neurological exam assessing for arthritis and weakness and other neurological features
- Exam for peripheral edema
- Thorough skin exam for rashes (malar, discoid, vasculitic, periungal erythema)

After you complete your exam you begin to think about further investigations. Which tests would you like to send this patient for?

<u>SLE Investigations:</u>
Start with your more 'basic' workup. It would be best to consult with a Pediatric Rheumatologist before ordering more extensive investigations.

- CBCd, blood film, extended lytes, Cr, Urea, ESR, CRP, urinalysis including microscopy, urine protein: creatinine ratio

- AST, ALT, albumin, INR, PTT (can have liver dysfunction in the form of acute autoimmune hepatitis and coagulation abnormalities), CK (myositis)
- TSH, ANA, C3, C4, Immunoglobulin panel, Rheumatoid Factor, DAT (Coombs test)
- Consider infection (either DDx or trigger of a flare of SLE): Depending on the patients features -parvovirus, respiratory panel, CMV, EBV, blood and urine culture (consider)
- More extensive work-up: ENA Panel (4 Extractable Nuclear Antigen Antibodies), Anti-dsDNA, Anti-phospholipid antibodies
- May consider: glucose, anti-TTG, D-dimer, fibrinogen, cholesterol studies, ferritin, haptoglobin, LDH
- As symptoms indicate: CXR, ECG, echo – other investigations and consultations as necessary
- Opthalmologic Eye exam is essential

Treatment approach:
Non-pharmacologic – sun protection is key to preventing rashes and avoiding flares

Pharmacologic treatment (these would all be initiated in conjunction with advice from pediatric rheumatology)
- Hydroxychloroquine (Plaquenil)
- NSAIDS (If no renal involvement)
- Steroids (from low dose oral to high dose IV steroids)
- Disease Modifying Anti-Rheumatic Drugs (DMARDS), biologic medications or cytotoxic drugs

Key Take Home Points:
1. SLE is a multi-system autoimmune disease
2. SLE is more common in adults but certainly can be seen in children
3. The 'classic' pediatric SLE patient would be a non-Caucasian, adolescent female
4. Renal disease is more common in pediatric SLE than in adults with SLE
5. Remember to consider SLE on your differential and use the pneumonic to help you recall the criteria.
6. Consult Pediatric Rheumatology if you suspect SLE

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