Investigations show leukopenia ($2.22x10^9$/L), normocytic anemia (hemoglobin 9.5 g/dL), an elevated ESR (57 mm/hr) and C-reactive protein (36 mg/L), hypoalbuminemia (30 g/L), mild proteinuria (0.08 g/mmol), hematuria (29/hpf), pyuria (20/hpf), slightly elevated creatinine (88 µmol/L, giving an estimated GFR of 56 ml/min/1.73 m^2). Eventually, urine culture returns positive for E. coli. She also has a positive Direct Coombs' test, antinuclear antibodies, anti-RNP, anti-β_2-glycoprotein IgG, anti-cardiolipin IgM. Her complement 3 (80 mg/dl) is slightly low, but she has a normal complement 4 level and negative anti-double stranded DNA. Ultrasound shows a dysplastic left kidney. A renal biopsy of the right kidney reveals an unexpected result. Come and see!

Learning Objectives

- Describe the lesser-known cutaneous manifestations of lupus in children
- Describe renal involvement in lupus beyond the known classification of lupus nephritis
- Discuss the treatment of lupus nephritis

Case 2: Two girls with lupus and abdominal pain

1. A 16-year-old girl presents with fever lasting one-month, extreme lethargy, loss of weight, alopecia and a facial rash. The diagnosis of systemic lupus erythematosus (SLE) is made with additional features of pancytopenia, positive direct Coombs' test, positive ANA and anti-double stranded DNA, hypocomplementemia. She is started on oral prednisolone 30 mg twice daily and hydroxychloroquine. One day after initiation of therapy, she returns to the Emergency Department with severe abdominal pain and vomiting.
2. An 18-year-old girl has received treatment for SLE since the age of 13 years old. She presented with cutaneous vasculitis and arthritis with cytopenia. Her current treatment included prednisolone 5 mg daily, mycophenolate mofetil 1 g twice daily and hydroxychloroquine. She has had persistent neutropenia for the last 2 years without infection and low complement 3 (75 mg/dl) without clinical features of lupus. She has had one month of abdominal bloating and abdominal pain following an acute food poisoning with another family member. While on a trip to Italy, the abdominal pain and nausea worsened and she was admitted to the Emergency Department. What could this be and what must be done?

Learning Objectives

- Describe the treatment approach to abdominal pain in a patient with lupus
- Describe the clinical features and outcomes of acute pancreatitis in childhood lupus
- Explain the clinical features and outcomes of 'lupus gut' in childhood lupus
- Discuss the best practice for management of acute pancreatitis in lupus and 'lupus gut'

13 MANAGEMENT OF PAEDIATRIC LUPUS

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Case 1: Newborn with cutaneous lesions

A 23-day-old newborn referred by his primary care pediatrician for skin lesions on the face, scalp, and upper trunk, in the form of erythematous annular plaques with hyperkeratosis, somewhat scaly. No other clinical manifestations were found. There was no history of fever and the baby was feeding well. No recent vaccination.

The mother had one spontaneous abortion in the past. The recent pregnancy was uneventful, and the baby was born from a spontaneous vaginal delivery at week 37. The birth weight was 2.6 kg and the Apgar score was normal.

The laboratory findings showed normal white blood cell count, no evidence of inflammation and hepatic and renal function. ANA 1/640; anti-Ro and anti-La antibodies were positive.

In relation to this clinical picture, indicate what your approach would be at this moment.

Learning Objectives

- Describe the clinical features of neonatal lupus
- Describe the diagnostic and therapeutic approach of neonatal lupus
- Discuss the prenatal study of pregnant women with lupus

Case 2: Child with skin lesions related to cold exposure

A 5-year-old boy presented with no significant medical history or known drug allergies. He was born at 39 weeks after an uncomplicated pregnancy to unrelated parents, birth weight 3.1 kg. There was no evidence of TORCH or other infections and he has been correctly vaccinated to date. His parents report that every winter, since he was 1 year old, he presents painful violaceous lesions on his hands and feet, but there are no lesions during summer. On examination, he presented chilblain lesions on his hands and feet.

Laboratory test findings were unremarkable, including liver and renal function and acute phase reactants. Rheumatic factor, ANA, anti-dsDNA antibodies, antiphospholipid serology were negative. Normal complement C3/C4. There was no evidence for hypergammaglobulinemia, cold agglutinins, viral or bacterial infection.

The father tells us that he suffers similar episodes and we see scars with tissue loss in his earlobes.

What additional examinations would you consider next?

Learning Objectives

- Discus approach to the main causes of monogenic lupus
- Describe suggestions for genetic studies on suspicion of monogenic lupus

Case 3. Diffuse alveolar hemorrhage in an adolescent girl with lupus

A 16-year-old female presents with asthenia, anorexia and low-grade fever in the last 2 weeks. She is otherwise asymptomatic with no other medical history of interest.

Laboratory test findings show lymphocytes 800/mm^3; platelets 113.000/mm^3, Hb 10.5 g/dl, creatinine 1.04 mg/dl, hypoalbuminemia 26 g/l, low C3 and C4 fractions (160 mg/l; < 29 mg/l); ESR 34 mm, CRP 4 mg/l. ANA 1/640, anti-dsDNA Ab >600 UI/ml. Urine: hematuria >100/hpf, pyuria 10/hpf, UPCR 4.74 mg/mg. Lupus anticoagulant negative, IgG/IgM β_2-GPI negative, IgG/IgM anti-cardiolipin negative.

Kidney biopsy shows class IV glomerulonephritis -ISN classification-. She received pulsed methylprednisolone plus oral prednisone and mycophenolate mofetil 2 g/day.

One week later, she suddenly deteriorated with acute severe respiratory distress, severe hypoxemia, unstable shock, and hemoptysis. Chest X-ray showed bilateral lung infiltrate suggestive of diffuse alveolar hemorrhage. She was transferred to paediatric intensive care and mechanical ventilation, including high-frequency ventilation, was required. Flexible bronchoscopy confirmed diffuse alveolar hemorrhage. She was treated with pulses of methylprednisolone, intravenous cyclophosphamide, and plasmapheresis.

Learning Objectives
- Explain pulmonary manifestations in lupus
- Discuss therapeutic approach to diffuse alveolar hemorrhage
- Describe prognosis of this life-threatening complication of SLE

14 MANAGEMENT OF CARDIOVASCULAR INVOLVEMENT IN SLE

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Case 1: A 35-year-old woman of Hispanic ancestry

A 35-year-old woman of Hispanic ancestry received a diagnosis of systemic lupus erythematosus (SLE) in June 2021, based on polyarthralgia/itis, malar rash, proteinuria of 1400 mg/24 h, positive antinuclear antibodies, anti-double-stranded DNA antibodies (anti-dsDNA), with hypocomplementemia. Lupus anticoagulant, anticardiolipin and anti-β_2-Glycoprotein-I antibodies were negative. Her SLEDAI score was 16. A kidney biopsy was performed showing a focal proliferative glomerulonephritis (Class III), with a score of 12 and 0 of activity and chronicity, respectively. She was treated with hydroxychloroquine 400 mg/day, prednisone 20 mg/day, and mycophenolate mofetil 3000 mg/day as induction therapy.

In September 2021 she came to the emergency room due to persistent tachycardia, dyspnea on moderate exertion, and chest pain. At admission she presented elevated ESR and C-reactive protein level, normal kidney function tests, proteinuria of 350 mg/24 h, and positive anti-dsDNA, with low C3 and C4. During hospitalisation she presented fever, and worsening dyspnea, for which she required oxygen therapy. The electrocardiogram showed sinus tachycardia and the echocardiography a systolic dysfunction and a hypokinetic left ventricle (inferior and lateral walls) with an ejection fraction of 40%. Troponin T and brain natriuretic peptides were elevated. The SARS-CoV2 RT-PCR was positive. With the suspected diagnosis of acute myocarditis in the context of SARS-CoV2 infection, the patient was treated with methylprednisolone pulses, IVIG, and respiratory support.

Learning Objectives
- Describe the different myocardial manifestations in a patient with SLE.
- Discuss complications and differential diagnosis with allied diseases.
- Discuss the treatment of myocarditis in a patient with SLE.

15 MANAGEMENT OF CARDIOVASCULAR INVOLVEMENT IN SLE

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Case 1: A 40-year-old woman with myocardial infarction

A 40-year-old female was diagnosed with systemic lupus erythematosus (SLE) at the age of 30 based on malar rash, arthritis, positive antinuclear antibodies, anti-double-stranded DNA antibodies, hypocomplementaemia, and biopsy-proven Class IV lupus nephritis. She was treated with glucocorticoids (GC), hydroxychloroquine (HCQ), and intravenous pulses of cyclophosphamide followed by mycophenolate mofetil (MMF) achieving complete remission 6 months later. Four years later, she suffered from a second SLE flare in the form of Class IV lupus nephritis as well as arthritis, receiving induction treatment with GC and MMF and achieving complete renal response 8 months later. She remained in lupus low disease activity for the next 5 years with prednisone 2.5 mg/day, HCQ 300 mg/day, and MMF 500 mg/12h. She was a current smoker, and her previous history included arterial hypertension and dyslipidaemia treated with enalapril 10 mg/day and atorvastatin 20 mg/day.

At the current admission, she presented at Emergency Department with thoracic pain and shortness of breath. She was diagnosed with myocardial infarction. Coronary angiography showed an atherosclerotic plaque in anterior descending coronary artery that required percutaneous coronary intervention and stenting. The patient was discharged without acute complications under treatment with dual platelet antiaggregation.

What could we have done to avoid this outcome?

Learning Objectives
- Discuss the general management of cardiovascular risk factors in patients with SLE
- Discuss the usefulness of different scoring tools to assess the atherosclerotic cardiovascular disease in SLE patients and the potential utility of imaging
- Discuss the objectives of treatment (primary prevention) of the different cardiovascular risk factors (hypertension, dyslipidaemia, tobacco) in SLE patients and the indications of aspirin in primary prevention

16 MANAGEMENT OF LUPUS NEPHRITIS

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The therapeutic armamentarium of the lupologist is expanding, notably when faced with one of the most frequent severe systemic lupus erythematosus (SLE) organ manifestation: Lupus nephritis (LN). Based on real-practice case studies, we will address the latest guideline recommendations for the management of LN. With novel agents at hand, we will make a journey through several treatment strategies for LN and provide key learnings by interactive discussions.