PEDIATRIC CUTANEOUS AUTOIMMUNE DISORDERS

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DISCLOSURES

• None
PRETEST QUESTION 1

- SLE:
  - A. More common in men than women
  - B. More common in Caucasians than AA or Asians or Hispanics
  - C. Can be diagnosed solely with one skin sign and one systemic involvement
  - D. NLE always progresses to SLE
  - E. None of the above are true
PRETEST QUESTION 2

• Juvenile Dermatomyositis
• A. can be a sign of malignancy in childhood dermatomyositis
• B. Rash specific to DM can include butterfly malar rash, scarring alopecia and mouth ulcers
• C. In childhood DM muscle involvement can be detected by MRI
• D. Anti Jo1 is specific for childhood DM
PRETEST QUESTION 3

• Morphea

• A. has a bad prognosis in children leading to systemic sclerosis

• B. Can be associated with other autoimmune disorders.

• C. Can present with thick indurated atrophic plaques

• D. Associated with organism causing RMSF

• E. Band C are correct
PRETEST QUESTION 4

• Childhood LSA
• A. limited to girls only
• B. Mostly in girls but boys can be affected
• C. Treatment with very mild steroids is sufficient
• D. Most cases are associated with sexual abuse
PEDIATRIC CUTANEOUS AUTOIMMUNE DISORDERS

- Lupus: Neonatal Lupus, Acute SLE, Subacute LE, Chronic LE
- Dermatomyositis
- Morphea/Localized Scleroderma
- Lichen sclerosus et atrophicus
LUPUS ERYTHEMATOSUS

• Chronic small vessel vasculopathy affecting the skin and most other organ system
• Unknown etiology; familial cases
• 15 -20% of all cases of SLE present in first 2 decades
• Except for NLE rarely seen before 3 yo
• After puberty F:M is 9:1
• Increased prevalence and severity in AA and Hispanics Asians
• Affected children more severe disease and increased risk of renal and CNS involvement
NEONATAL LUPUS ERYTHEMATOUS

- Seen in infants born to moms at risk for SLE RHA or mixed CTD
- Majority of moms are asymptomatic
- Face scalp lesions average onset at 6 weeks
- Raccoon eyes, annular, discoid, atrophic, telangiectasias
- Anti-Ro SSA, Anti-La SSB and anti-U1 RNP Ab
- 3rd degree heart block 15 to 30%
NEONATAL LUPUS ERYTHEMATOSUS

• NLE most frequent cause of congenital heart block
• If 3rd degree heart block, usually irreversible requiring pacemakers
• Some with 1st and 2nd degree heart block spontaneous resolution
• Also liver involvement, splenomegaly, lymphadenopathy, hematologic abnormalities
NLE

• Results from transplacental passage of anti Ro and anti La antibodies
• Need complete exam
• Confirmatory labs: Ab to Ro, La and U-RNP, CBC with platelets, liver fcn test
• Confirmatory bx
• Bradycardia /murmur: needs EKG and Echo
NLE

- Clearance of lesions 6 to 12 months of age.
- Tx: avoidance of sun
- Treatment topically?? Not shown to have effect on outcome of residual lesions
- Check mom and refer mom
- 22 % risk of NLE in second child
- Systemic corticosteroid to at risk moms before 16 weeks GA, Fetal echo 16 to 24 wks GA
- Pts with NLE has no increased risk of developing lupus and other autoimmune disorders, beyond that of their siblings. May have familial tendency.
NEONATAL LUPUS
NEONATAL LUPUS
NEONATAL LUPUS
MAKE SURE IT IS INDEED NLE!!
LUPUS ERYTHEMATOSUS

Dx : at least 4 criteria
LE

- Malar rash
- Discoid rash
- Photosensitivity
- Oral ulcers
- Arthritis
- Serositis
- Renal complications
- Neurologic complications
- Hematologic complications
- Immunologic complications
- Abnormal ANA titer
Skin signs of lupus

- Malar butterfly rash
- photosensitivity
- discoid lupus
- periungual telangiectasia
- Raynaud’s phenomenon
- Livedo reticularis
- scarring alopecia
- oral ulcers
ACTIVE DISCOID LUPUS
ACTIVE DISCOID LUPUS
DISCOID LUPUS
DISCOID LUPUS
SCLE
SCLE
BUTTERFLY RASH
DISCOID LUPUS
SCARRING ALOPECIA
SCARRING ALOPECIA
MOUTH ULCERS
DIAGNOSIS LABORATORY EVALUATION

- Pathology
- Serologic >1:160 ANA titer
- Pattern homogeneous
- Serum complement CH50 C3 C4
- Anti DS DNA Ab increased risk of renal dz
- Anti RNP Ab lower risk of renal dz
- Anti Smith Ab specific for LE and higher risk for renal dz
- Antiphospholipid antibodies can be elevated.
DRUG INDUCED LE

- Minocycline in adolescents treated for acne after 2 years of tx.
- Myalgia arthralgia arthritis livedo reticularis elevated LFT and antineutrophil ab
- No renal dz.
- Anti histone Ab elevated. Anti ds DNA and complement levels normal
THERAPY FOR CUTANEOUS LESIONS

• Avoidance of sun; trigger for skin and systemic involvement
• Sunscreens opaque blockers
• Topical steroids and TCI
• Antimalarial therapy
• CTCS systemic; vit d supplementation for osteopenia; growth retardation; avascular necrosis
JUVENILE DERMATOMYOSITIS

• 3 per million children per year
• Skin muscles internal organs
JDM CRITERIA

- Progressive symmetrical proximal mm weakness with or without dysphagia or resp weakness
- Mm bx evidence of myositis or necrosis
- Elevation of mm enzymes
- Evidence of myopathy on EMG or MRI in children
- Typical cutaneous signs in children
• Definite Dx: skin signs and 3 of the above
• Probable Dx: 2 of above plus 2 of above criteria
JDM

• 2x in Females than Males
• Bimodal distribution: 2 to 5 yo and 12-13 yo
• Malignancy not an association in pediatric patients.
• More inflammation necrosis of muscle more calcinosis more small vessel vasculopathy and lower mortality
• Insidious onset >50 % of children 30 % have fulminant onset
• Amyopathic form only in 3-5 % of children, most progress to develop myositis
DIAGNOSIS

• Clinical grounds
• Elevated CPK aldolase ALT LDH
• MRI scan with muscle inflammation.
• Skin biopsy not usually helpful
• Anti Mi 2 Anti Jo 1 Ab rarely elevated in kids. ANA + speckled nonspecific pattern
DERMATOMYOSITIS

• Photosensitivity
• Upper torso and extensor surface psoriasiform rash
• Nail fold telangiectasias
• Gower’s sign
• Dysphagia/dystonia/dyspnea
• Nodular calcification
• Constitutional Sx: fever fatigue
DERMATOMYOSITIS SKIN SIGNS

Gottron’s sign
Heliotrope
Shawl Sign V sign
Mechanic’s hands
Psoriasiform scaly scalp
Skin signs of Dermatomyositis

- Heliotrope
- Gottron’s Papules
- Dermatitis on V neck, elbows, knees
- Raynaud’s periungual telangiectasia and dermatitis
- calcinosis cutis
- vasculitis
PERIORBITAL AND CHEEK TELANGIECTASIAS
HELIOTROPE RASH
SHAWL SIGN: SUN EXPOSURE
TELANGIECTASIA AND ATROPHY, POIKILODERMA
DM AND CALCINOSIS CUTIS
PERIUNGUAL TELANGIECTASIAS
KNEE TELANGIECTASIA
PERIUNGUAL TELANGIECTASIAS
GOTTRON’S PAPULES
GOTTRON’S PAPULES
PERIUNGUAL TELANGIECTASIAS AND GOTTRON’S
Juvenile Dermatomyositis

- Does not always start with muscle enzyme elevation
- Can present with psoriasiform scalp dermatitis
- Can present as psoriatic plaques on the elbows knees
- Look for other more specific signs of DM
TREATMENT

• Referral to Rheum
• Hospitalization if acute and involving palatal resp mm
• Aspiration
• Physical therapy
• High dosage CTCS and KTX
• Biologics? Infliximab
SCLERODERMA

• Localized: limited to the skin
  • morphea plaque type morphea
  • Linear scleroderma: En coup de sabre and Parry Romberg syndrome
  • Pan sclerotic disabling morphea

• Systemic scleroderma
  • PSS
  • CREST
LOCALIZED SCLERODERMA

- Morphea plaque type
- Linear scleroderma
- Pansclerotic disabling morphea
INCIDENCE LOCALIZED SCLERODERMA

- 0.4/1 per 100000
- More common than systemic form of scleroderma
- 2to 3 :1 ratio F :M
- Mean age 10 yo.
- Familial
- Fhx of autoimmune disorders
MORPHEA

• Only systemic manifestation is arthralgia
• Can be asstd with other CTD
• Plaque type morphea most common and linear type more common in children than adults.
• Better prognosis
• Link to infection?? Lyme?? Trauma, Valproic acid.
PLAQUE TYPE MORPHEA
BURNT OUT MORPHEA
PATHOLOGY OF MORPHEA
EN COUP DE SABRE
EN COUP DE SABRE
PARRY ROMBERG SYNDROME
LOCALIZED SCLERODERMA
LOCALIZED MORPHEA
PAN SCLEROTIC DISABLING MORPHEA
PAN SCLEROSING DISABLING MORPHEA
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TREATMENT

• Difficult
• Topical TCI and Vitamin D
• PUVA, UVA 1
• Potent steroids
• CTCS and MTX
• Oral calcitriol - watch renal stones restricted calcium in diet
• Fillers
LICHEN SCLEROSIS ET ATROPHICUS

• Primarily female
• 1/900
• Onset before 13 years of age
• Most before age 7
CHILDHOOD LSA

• Sharply demarcated ivory white plaque vulva and perianal region
• Sx of severe itching, ecchymosis
• Dysuria discharge malodorous discharge constipation
• Figure of 8
• Can occur rarely in glans penis boys balanitis xerotica obliterans but less common
CHILDHOOD LSA

DDX

• Pinworm infestation
• Irritant contact dermatitis
• UTI
• Candidiasis
• Child abuse
CHILDHOOD LSA

- Rarely bx needed due to characteristic clinical appearance
- 60% Reported to improve or disappear at puberty
- Can be reactivated with pregnancy trauma or anovulatory drugs
CHILDHOOD LSA VULVAR
TREATMENT CHILDHOOD LSA

• Superpotent steroids 6 to 8 weeks
• Tacrolimus
• Boys circumcision
• Plastic surgery if labial fusion or scarring
• Look for secondary infection eg Strep if discharge or drainage
Vasculitis

- Medications
- Malignancy
- Serum Sickness
- Strep and other infections
- HSP
- Hep B
- CTD
- Cryoglobulinemia
VASCULITIS
OTHER NON SPECIFIC SKIN MANIFESTATIONS OF CTD

• Urticaria
• Erythema nodosum
• Erythema multiforme
• Livedo
• Panniculitis
• Pernio chilbains
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