Case based presentations: Things that saw me... before I saw them

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No relevant conflicts for this talk
“I wouldn’t have seen it if I hadn’t believed it”
Marshall McLuhan
“Reality goggles”

• We all have perceptions of what is real
• We pre-interpretation what we see
• Plus: Gives us the ability to navigate life without having to analyze/critically dissect every interaction or experience we have
• Minuses: We misperceive or do not see certain things
• “A man should look for what is, and not what he thinks should be.” Einstein
Some illustrative cases

• 8 cases/case scenarios I have been confused or stumped
• Literature or colleagues given clues to solve the mysteries
• Emphasize that we can/do *always* see new things
• If you are not learning new stuff you are missing out
Some skin diseases where this applies

• EVHC
• Lateral Nail Fold Hyperplasia
• Idiopathic facial aseptic granuloma
• Idiopathic eruptive macular pigmentation
• Prurigo pigmentosum
• Asymmetric Periflexural Exanthem of Childhood
• Perianal pyramidal protrusion
• Clear-cell papulosis
Eruptive Vellus Hair Cysts (EVHC)

• First described by Esterly in 1977; now > 85 PubMed citations
• EVHC may be sporadic or inherited
  • Associated with ectodermal dyplasias and steatocystomas
• ? Defect in infundibular keratinization leading to vellus hair follicle occlusion
• Benign but difficult to treat.
Congenital hypertrophy lateral nail folds (CHLNF)

• Prospective study of 250 newborns and infants
• Most frequent abnormalities were onychoschizia in 2.4% and CHLNF with ingrown nails in 2.4% infants.

• Management:
  • Can remit spontaneously or with topical clobetasol
  • Occasionally surgery is needed

Idiopathic “facial aseptic granuloma”

• Case series of 30 children, mean age 3.8 yrs
• One or more acquired facial nodules >1 month mainly on cheek
• U/S hypoechoic without calcification
• Neg bact or AFB cultures, neg cat scratch serologies 70%; 4/26 responded to antibiotics
• Mean size 10mm (3-25mm)
• Spontaneous regression mean duration 11 mos
• Histology 5 cases: Chronic dermal inflammatory granuloma lymphocytes, histocytes, neuts, numerous foreign body type giant cells
Hyperpigmented macules on the face of young children: a series of 25 cases.

• Retrospective review of 25 children
• M=F; Age range 2 to 24 months (mean 12.2 months, median 6 months)
• Presented abruptly in the summer (12 cases), spring (5 cases), winter (5), and fall (2)
• Histopathology (3 cases) = PIPA.
• Lesions persisted in 19/25 with f/u ranging from 3 months to 4.5 years,

Just to confuse you a little...
IEMP with papillomatosis (IEMPP)

- Several reports/case series many from India
- Mainly in school-aged children
- Histology shows papillomatosis of the dermis with prominent pigmentation of the basal layer (pigmented papillomatosis) without significant dermal inflammation

Verma S, Thakur BK Indian Dermatol
What is this?

• Pattern looks like Confluent Reticulated Papillomatosis (CRP)
• Pruritic, not typical morphology
• Path does not show CRP; instead subacute spongiotic dermatitis with eosinophils
• Pathologist suggested possible diagnosis...
Purigo pigmentosa
(Nagashima’s disease)

• Presents as pruritic erythematous papules, papulovesicles, and vesicles appearing in a reticular pattern on the back, chest, or neck.
• Has been described in > 300 patients
• 3 stages: pruritic urticarial papules then fully developed then PIPA
• Relationship to CRP is debated
  • ? Inflammatory variant
  • Does respond to minocycline Rx
• Mainly young Asian women (20-30 years)
• Has been associated with several systemic conditions:
  • Anorexia nervosa, diabetes, prior bariatric surgery (relationship to ketosis is postulated)
  • Adult-onset Still’s disease
  • *H. pylori* infection
  • Sjögren’s syndrome

Asymmetric Periflexural Exanthem: Prospective Study

• 67 children with APEC
• No triggering factors, or inter-human transmission noted
• Further clinical findings:
  - Prodrome 61%
  - Fever 40% usually after onset of eruption
  - Adenopathy 70%
  - Persistence usually 3-6 weeks; 10% >7
Unilateral laterothoracic exanthem: Clinicopathologic study 48 patients

• Usually starts close to axilla, may evolve to bilateral but usually unilateral dominance
• Initially morbilliform; more eczematous over time with dusky center.
• Mean age 24 months; Duration 5 weeks; 2:1 females; Most in winter/spring
• Histology: Eccrine lymphocytic infiltrates
• No single infectious agent found

Ref: McCuaig CC et al. JAmAcad Dermatol 1996;34:979-84
Asymmetric Periflexural Exanthem: Histologic Features

- 9 skin biopsies during first 3 weeks of eruption
- Mild to moderate interface dermatitis with occasional necrotic keratinocytes
- Clear predominance around sweat glands
- Findings more specific than previously thought

Ref: Coustou Arch Derm 1999;135:799-803
Does APEC need a name change?

• Exanthem is prevalent in childhood but has been reported in adults.

• Not always unilateral and involve groin and legs – why “laterothoracic” has been mostly abandoned

• Endorse the title ‘superimposed lateralized exanthem’ suggested by Happle et al

• Hypothesis increased responsiveness of a polygenic predisposed side of the body to various infectious agents.

Infantile Perianal Pyramidal Protrusion

• Previous reports refer to these lesions as “skin tags”
• IPPP proposed as name based on characteristic presentation, prepubertal, almost always in girls
  • Classically just anterior to anus (can be posterior)
  • Most healthy – constipation risk factor, also Lichen sclerosis; DDx includes Crohn’s dz
• No Rx needed unless underlying cause – resolves spontaneously

Clear-cell papulosis

- Clear-cell papulosis (CCP) is uncommon to rare entity, initially described in 1987
- Asymptomatic hypopigmented macules or flat-topped papules on the lower abdomen and along the milk lines in young children
- DDX PIPA, vitiligo, PLC, flat warts, tinea versicolor etc.
- CCP can be pathologically misconstrued as normal skin, with biopsy often nondiagnostic
- Path should show Toker cells, (CEA positive)
- Benign? Spontaneouly remits