



Paediatric Case presentation

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Objective

- Case presentation
- Discussion around the case
- Take home message



*Presenting
Complaint*

7-year-old Chinese boy

3 months history of

1. Painful/weak legs
2. Easily tired

Background History

Diagnosed with

- Eczema - RAST positive for egg and milk – avoiding
- Hay fever

Medication

- Topical steroid
- Moisturizer
- Daily cetirizine



Painful/weak leg

3 months

- Thigh pain
- Worse with exercise
- Difficulty getting up from sitting/lying down & getting dress
- Getting tired – refuse to walk long distance
- Slow with eating but no difficulty swallowing
- No joint swelling

Rash

- Ezcema – diagnosed early this year, no previous
- No Fhx athopy
- Not responding to moisturizer or steroids
- Middle of the year stopped egg and milk as RAST +ve - no difference
- Previously tolerating egg and milk



Other history

- No recent illness or weight loss
- No family history of Muscle or Rheumatological illness
- Still able to go to school

Examination

- Weight - 50th centile and Height - 50th centile .
- Pain walking & difficulty climbing up onto the examination bed
- Heart sounds, chest and abdomen normal
- pGALS examination:

He had thigh pain on knee flexion

- Other joints- no obvious swelling and full range of movements.
- Walk on his toes but refuses to walk on his heels.
- Difficulty getting up after squatting down.
- Positive Gower's sign.
- Generalized muscle weakness on both his upper and lower limb especially his proximal muscle.
- The rest of his upper and lower limb neurology normal. (tone, reflexes, plantar)
- Normal eye movement





Diagnosis

1. Significant weakness/pain

- Muscle origin

Muscle inflammation/destruction

Chronic

All muscle especially proximal

- Rash

Eczema? Consider distribution

Relation to muscle inflammation?

Investigations

- Normal Full blood count
- LFT - elevated ALT 111
- CK was elevated 380
- Anti CCP <1
- Normal renal function
- RF <9
- LDH elevated 336

	KIU(A)/L	Class

Codfish	<0.05	0
Cows Milk	3.96	3+
Egg (white)	2.23	2+
Peanut	<0.05	0
Soybean	<0.05	0
Wheat	0.33	0

A dark blue, irregularly shaped graphic with a splatter effect, containing the text "Take home message". The graphic has a rough, hand-painted appearance with various shades of blue and white splatters around its edges. The text is centered within the dark blue area.

Take home
message

<u>Disease</u>	<u>Upper motor neuron</u>	<u>Anterior horn cell</u>	<u>Neuromuscular junction</u>	<u>Peripheral nerve</u>	<u>Muscle</u>
		<ul style="list-style-type: none"> SMA Poliomyelitis 	<ul style="list-style-type: none"> Myasthenia gravis infant botulism 	<ul style="list-style-type: none"> Hereditary Sensory Muscle Neuropathy 	
Tone	↑	↓↓	variable	↓	N/↓↓
Distribution	hemiparesis paraparesis etc	variable, asymmetric LL>UL prox > distal	fluctuating cranial n involved	distal>proximal	proximal>distal (except myotonic dystrophy)
Reflexes	↑	↓/ normal	normal	↓/ absent	N/↓
Babinski	Upgoing	-	-	-	-
Special clinical features	↓ cognition atrophy late	fasciculations atrophy no sensory involvement	fluctuating course	sensory involved atrophy occ fasciculations	no sensory loss
Other		<ul style="list-style-type: none"> CK – normal/ ↑ 	<ul style="list-style-type: none"> anti-Ach antibodies (+ve in 90%) ANA, immune complexes 	<ul style="list-style-type: none"> CK – normal Nerve biopsy – ↓ large myelinated fibres 	CK –↑

The pGALS musculoskeletal screen		
Screening questions: <ul style="list-style-type: none"> Do you (or does your child) have any pain or stiffness in your (their) joints, muscles or back? Do you (or does your child) have any difficulty getting yourself (themselves) dressed without any help? Do you (or does your child) have any problem going up and down stairs? 		
FIGURE	SCREENING MANDIBLES	WHAT IS BEING ASSESSED?
	Observe the child standing (from front, back and sides)	<ul style="list-style-type: none"> Posture and habitus Legs/ankles – e.g. postural Deformity – e.g. leg length inequality, leg alignment (valgus, varus at the knee or ankle), scoliosis, joint swelling, muscle wasting, flat feet
	Observe the child walking and "Walk on your heels" and "Walk on your tiptoes"	<ul style="list-style-type: none"> Ankles, subtalar, midtarsal and small joints of foot and toes Foot posture (note if presence of normal longitudinal arches of feet when on tiptoes)
	Hold your hands out straight in front of you	<ul style="list-style-type: none"> Forward flexion of shoulders Elbow extension Wrist extension Extension of small joints of fingers
	"Turn your hands over and make a fist"	<ul style="list-style-type: none"> Wrist supination Elbow supination Flexion of small joints of fingers
	"Place your index finger and thumb together"	<ul style="list-style-type: none"> Manual dexterity Flexion/extension of small joints of index finger and thumb and hand/wrist/forearm grip

FIGURE	SCREENING MANDIBLES	WHAT IS BEING ASSESSED?
	"Touch the tips of your fingers"	<ul style="list-style-type: none"> Manual dexterity Flexion/extension of small joints of fingers and thumbs
	Squeeze the metacarpophalangeal joints for tenderness	<ul style="list-style-type: none"> Metacarpophalangeal joints
	"Put your hands together palm to palm" and "Put your hands together back to back"	<ul style="list-style-type: none"> Extension of small joints of fingers Wrist extension Elbow flexion
	"Reach up, 'touch the sky' and 'look at the ceiling'"	<ul style="list-style-type: none"> Elbow extension Wrist extension Shoulder abduction Neck extension
	"Put your hands behind your neck"	<ul style="list-style-type: none"> Shoulder abduction External rotation of shoulders Elbow flexion

FIGURE	SCREENING MANDIBLES	WHAT IS BEING ASSESSED?
	"In and touch your shoulder with your ear"	<ul style="list-style-type: none"> Cervical spine/lateral flexion
	"Open wide and put three (child's own) fingers in your mouth"	<ul style="list-style-type: none"> Temporomandibular joints (and check for deviation of jaw movement)
	Test for effusion at the knee (patella tap, or cross-flexures)	<ul style="list-style-type: none"> Knee effusion (small effusion may be missed by patella tap alone)
	Active movement of knees (flexion and extension) and test for crepitus	<ul style="list-style-type: none"> Knee flexion Knee extension
	Passive movement of hip (knee flexed to 90°, and internal rotation of hip)	<ul style="list-style-type: none"> Hip flexion and internal rotation
	"Stand forwards and touch your toes"	<ul style="list-style-type: none"> Forward flexion of thoracic/lumbar spine (and check for scoliosis)

Distribution of AD by Age

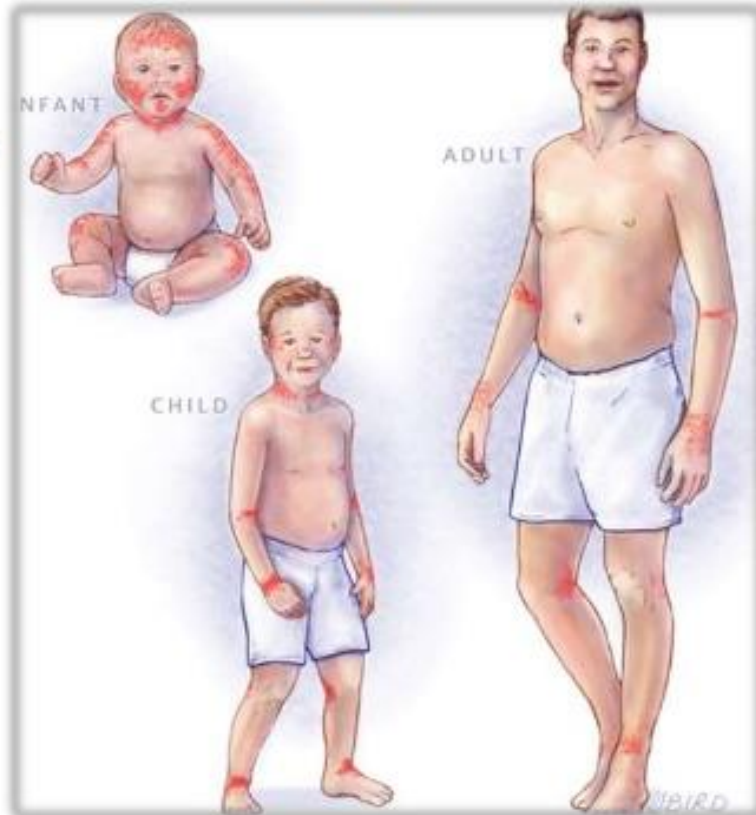
Infant

(birth-2 years)

Face (cheeks),
scalp, ears

Extensor
extremities

Seborrheic
dermatitis
overlap



Childhood

(2 years-puberty)

Face (cheeks)
Flexural extremities

Teenager-Adult

Localized flexural
extremities

Hands, dorsum feet

Be familiar with
typical distribution

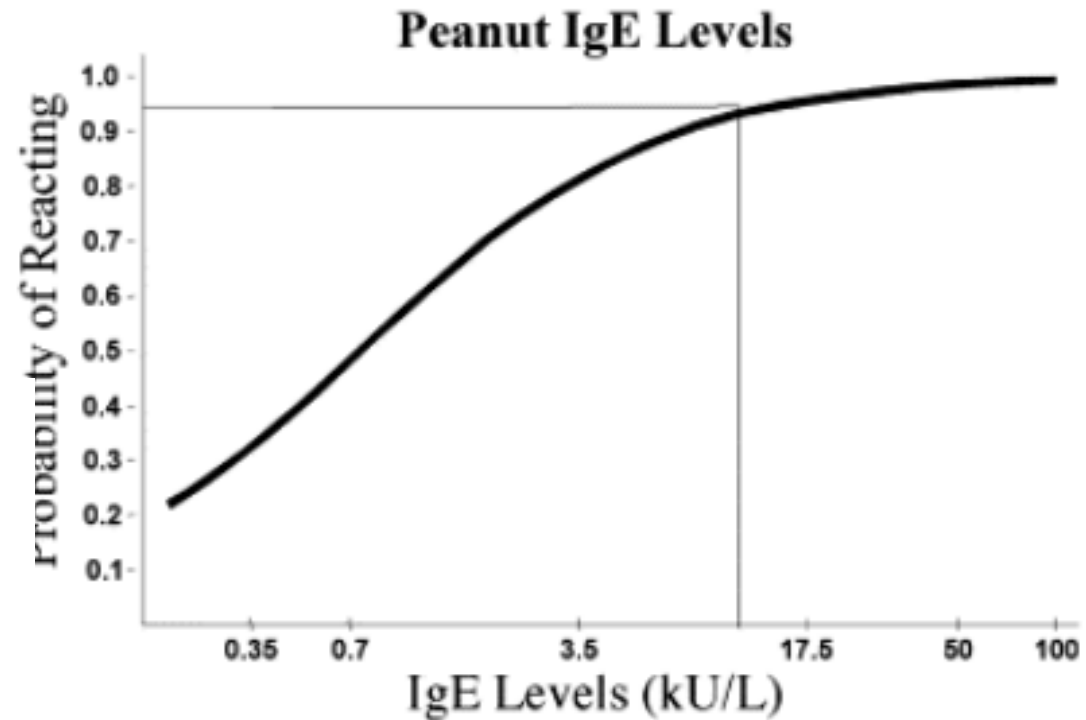
Past history and
Family history of
atopy

95% Predictive Level

Allergen	[kU _Δ /L]	PPV
Egg	7	98
- Infants ≤ 2 yrs ⁺	2	95
Milk	15	95
- Infants ≤ 2 yrs ⁺⁺	5	95
	KIU(A)/L	Class
Codfish	<0.05	0
Cows Milk	3.96	3+
Egg (white)	2.23	2+
Peanut	<0.05	0
Soybean	<0.05	0
Wheat	0.33	0

- + Boyano MT, et al. Clin Exp Allergy 2001; 31(9):1464-9.
 ++ Garcia-Ara C, et al. J Allergy Clin Immunol 2001; 107(1):185-90.
 +++ Clark AT, Ewan P. Clin Exp Allergy. 2003; 33(8):1041-5

PPV = Positive predictive value



Increasing probability of clinical reactivity with increasing level of food-antigen specific IgE value; note: values <0.35 do not exclude allergic reactivity

For both SPT and sIgE it is possible to have weakly positive tests associated with clinical allergy, and strongly positive tests associated with tolerance. Neither predicts the severity of the reaction



Thank you