

When Heavy Muscles Become Weak



Grand Round

A 17-year-old man with muscular hypertrophy and weakness.

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Declaration

- **There is no conflict of interest regarding this case presentation.**

Consent form

I [redacted] [Name] give my consent for information about myself/my child or ward/my relative (circle as appropriate) to be published in

[Name of journal, manuscript number and corresponding author]

I understand that the information will be published without my/my child or ward's/my relative's (circle as appropriate) name attached, but that full anonymity cannot be guaranteed.

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I have been offered the opportunity to read the manuscript.

Signing this consent form does not remove my rights to privacy.

Name [redacted]

Date [redacted]

Signed *ASX*

Author name [redacted]

Date [redacted]

Signed [redacted]

Please keep this consent form in the patient's case files. The manuscript reporting this patient's details should state that 'Written informed consent for publication of their clinical details and/or clinical images was obtained from the patient/parent/guardian/ relative of the patient. A copy of the consent form is available for review by the Editor of this journal.'

Case Scenario

A 17-year-old male

Student, non smoker, normotensive

permanent resident of Comilla

Admitted on 22nd April, 2015 with complaints of:

- **Tightness & swelling of all 4 limbs and chest for 3 months.**
- **Generalized weakness for same duration.**

Case Scenario (cont..)

- He was reasonably well until January,2016.
- Then he gradually developed painless swelling & tightness of both upper & lower limbs involving muscles of both arms, forearms, thighs & calves.
- Later involved:
anterolateral aspect of his chest wall
& neck region
- Patient was unable to flex his arms & legs to full extent, joints were unaffected.

Case Scenario (Cont..)

- **His weakness gradually increased, involving both proximal and distal muscles and hampered his daily activities.**
- **No H/O fever, joint pain, skin rash, cough, breathlessness, chest pain, palpitation, weight loss, night sweat, eye or ENT ailments.**
- **His bladder and bowel habits were normal.**
- **No family H/O similar type of illness.**

Case Scenario (Cont..)

- He has no H/O taking steroid or regular physical exercise.
- Treatment by local physician:
 - Tab. tolperisone (myolax)
 - Oral calcium & vitamins
- But his condition deteriorated.

General Examination

Anaemia

Jaundice

Cyanosis

Dehydration

Clubbing

Koilonychia

Leukonychia

absent

Weight- 54Kg

Height-162.5cm

BMI- 20.61kg/m²

Neck vein: not engorged

Thyroid: not enlarged

Edema – mild pitting edema in both legs

General Examination (cont'd)

Lymph node: Posterior cervical (Rt, size 2x2cm) & B/L axillary (3x2cm) lymph nodes were enlarged, firm, non-tender, not fixed to skin or underlying structures, no discharging sinus.

Skin- skin overlying the affected area was shiny. No pigmentation.

Pulse: 78 b/min

BP: 130/70 mmHg

Temp:98° F

RR: 16 breaths/min

Bedside urine revealed : albumin 1+

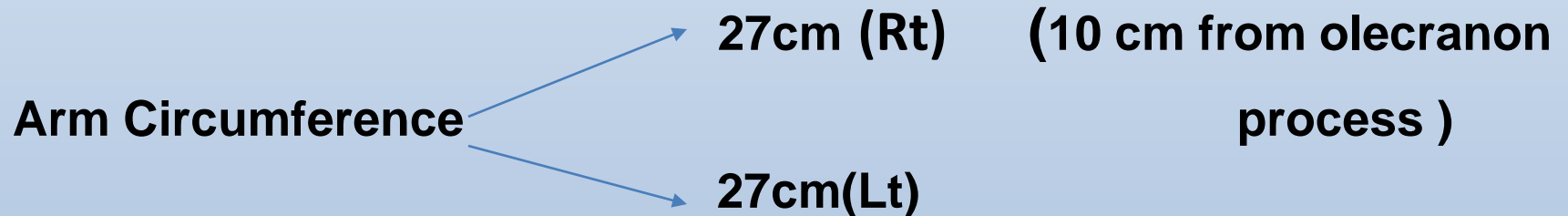






Musculoskeletal system

- **Gait- Myopathic gait.**
- **Arm -There is swelling of the muscle,which was non-tender**



Musculoskeletal system (Cont'd)

- **Legs-**

Circumference of legs :

→ **37cm(Rt.) (10 cm from tibial tuberosity)**
→ **35cm (Lt.)**

- **Spine- normal**

Nervous system

- **Mental Status : Normal**
- **GCS: 15/15**
- **Cranial nerves: intact including fundoscopy.**

Muscle Power

	Proximal	Distal
Upper Limb	4/5	4/5
Lower Limb	4/5	4/5

Muscle tone- diminished in all 4 limbs.

Reflexes

	Biceps	Triceps	Supinator	Knee	Ankle	Plantar
Right	++	++	++	+	+	flexor
Left	++	++	++	+	+	flexor

Sensory Examination

	Pain	Touch	Vibration	Joint Position
Upper limb	All modalities of sensation were intact			
Lower limb				

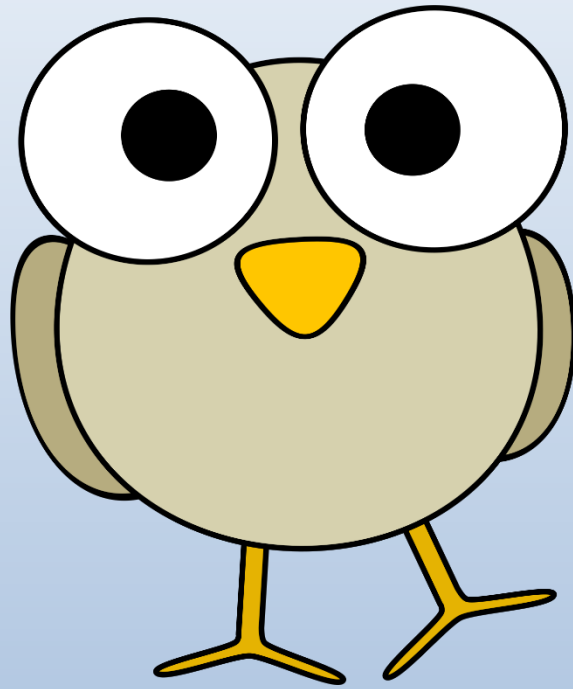
- **Examination of alimentary and cardiovascular system revealed no abnormality.**

**Muscle swelling and
tightness**

**Muscle weakness,
diminished
muscle tone &
reflexes**

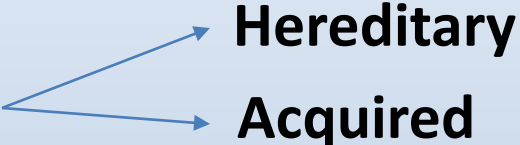
**Non-tender cervical
and axillary
lymphadenopathy**

**Pedal edema and
proteinuria**



??

Differential diagnoses

- **Lymphoma**
- **Sarcoidosis**
- **Myopathy** 

```
graph LR; Myopathy --> Hereditary; Myopathy --> Acquired;
```
- **Storage disorder**
- **Lipodystrophy**

Investigations

Hb: 12.2gm/dl

TC of WBC: 11, 200/ μ L

P: 52.4%, L:34.2%

M: 11%, E: 2.4%

Platelet: 4,06,000/ μ L

ESR: 10 mm in 1st hr

PBF:

RBC:Anisochromia with anisocytosis.

WBC- Mature with increased total count.

Platelet- normal

Urine for RME

albumin: +

pus cell: 2-3/HPF

RBC : 1-3/HPF

Granular casts: ++

S. albumin: 29.9 g/ L

24 hours UTP: 2.15 gm

CXR P/A VIEW

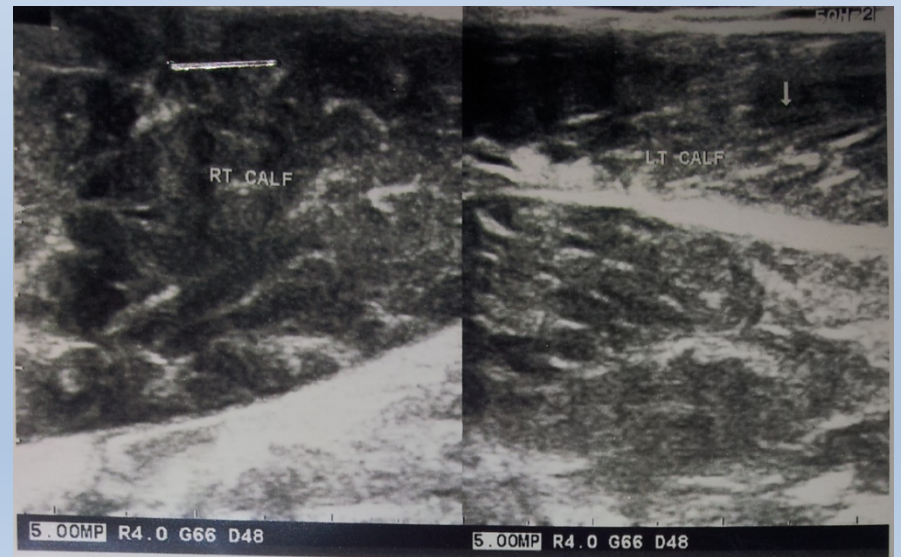
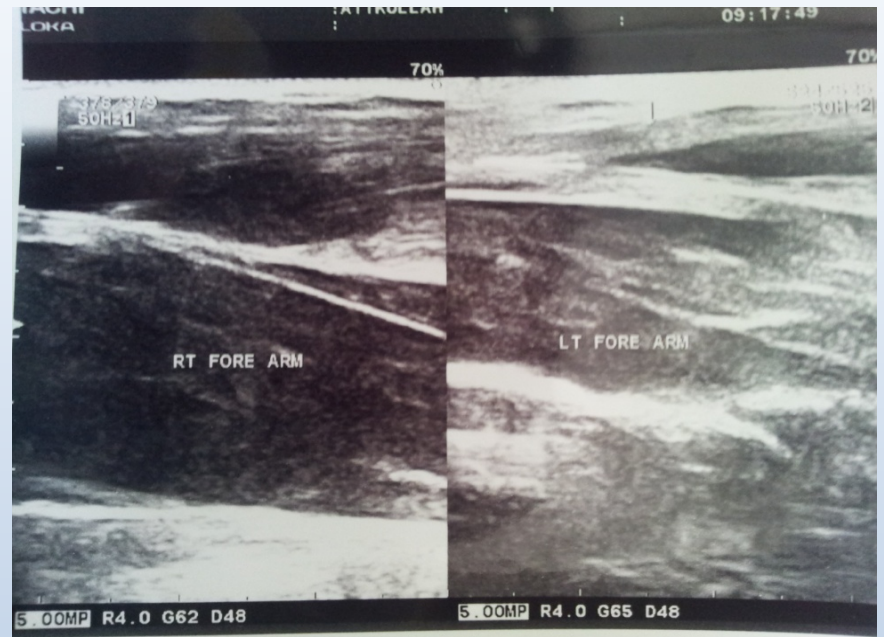


USG of affected muscles :

Fibres are not well separated, intermuscular septa are thin, echogenicity is altered, no calcification, no vascular abnormality.

Possibilities are:

- ✓ Storage disorder
- ✓ Muscular hypertrophy



USG Of Whole Abdomen



✓ Bilateral swollen kidney

S. creatinine : 0.8 mg/dl

ALT : 28 U/L

Na: 137mmol/L

K: 4.3 mmol/L

Cl : 101mmol/L

HCO₃ : 25mmol/L

RBS: 5.2 mmol/L

S. calcium: 9.48 mg/dl (corr.)

**S. Cholesterol- 150mg/dl, TG-
100mg/dl, HDL-55mg/dl, LDL-
60mg/dl**

CPK: 106 U/L

LDH- 383U/L

Thyroid function test:

FT4: 11.00 pmol/L

TSH : 4.88 μ IU/L

**Anti thyroid antibodies-
negative**

ANA : negative

Anti-Scl-70: negative

Consultations

Dept. of Surgery-for muscle & lymph node biopsy

Dept. of Nephrology-

For unexplained proteinuria & bilateral swollen kidney

Renal biopsy was advised

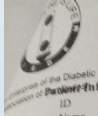
Dept. of Neurology- For evaluation of neurological symptoms.

NCS and EMG was advised.

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NCS and EMG

Sensory motor polyneuropathy (axonal) of both lower limbs with ongoing denervations. Muscle disease could not be excluded


 Bangladesh Institute of Research and Rehabilitation in Diabetes, Endocrine and Metabolic Disorders
 DEPARTMENT OF NEUROLOGY
 WHO Collaborating Center for Prevention and Control of Diabetes

Patient Information

ID	Neu-5873266	In/Out Patient	In
Name	Atikullah	Refer. Dept.	Unknown
Date of Birth		Physician	Unknown
Age	17Yrs	Examination Date	2015/05/04
Sex	Male	Examination No.	
Height		Examined by	
Weight		Patient Status	Walking
Complaint / Symptom		Weakness all limbs, Par. Four limbs Medical	

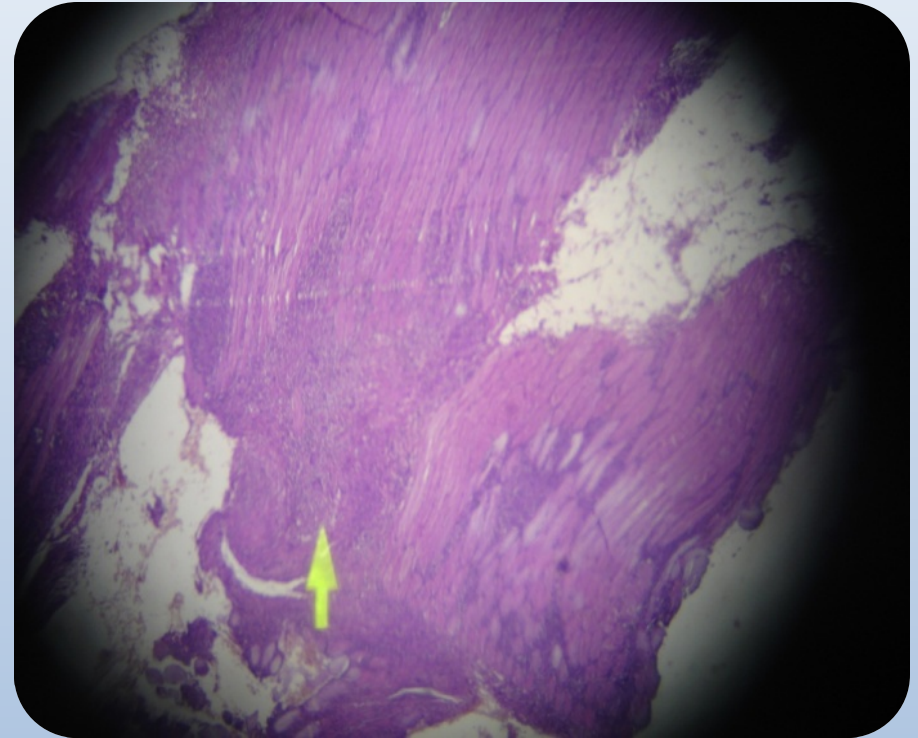
EMG Findings Summary

Muscle/Side	Ins. Act.	Fibs.	Pos. Wave	Fasc.	MYO Disch	Normal MUP	Poly	Low Amp	High Amp	Dur.	Recruit	Int. Patt.
Gastroc. Medial H	B	Incr.	+2	+1	0	0	++	+1			Normal	
Gastroc. Lateral H	B	Incr.	+2	+1	0	0	++	+2			Normal	
Soleus	B	Incr.	+2	+1	0	0	++	+2			Normal	
1st Dorsal Inter.	B	Normal	0	0	0	0	++	+2			Normal	
brachioradialis	B	Normal	0	0	0	0	N	+2			Normal	
Biceps Brachii	B	Normal	0	0	0	0	N	+2			Normal	

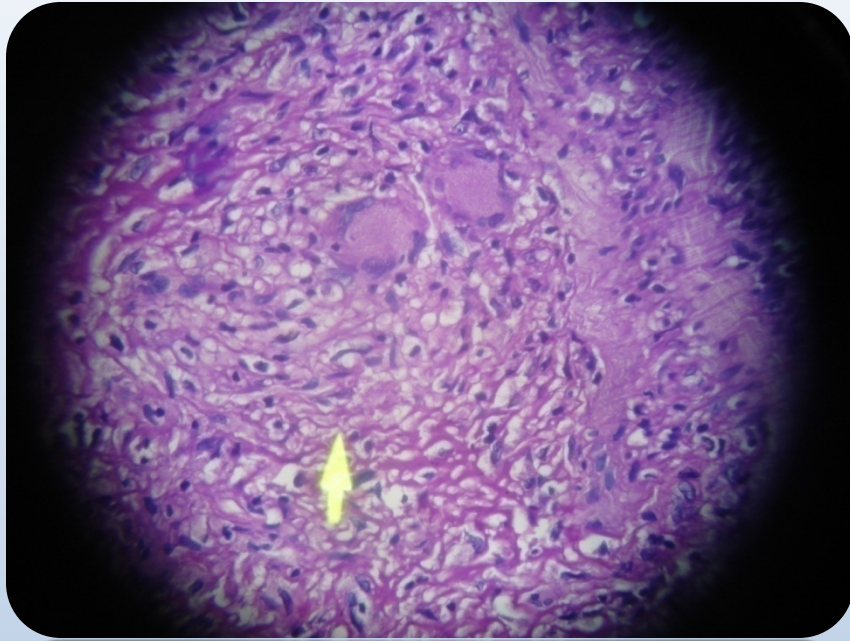
Motor Nerve Conduction Study

Site	Latency (ms)	Amplitude	Area	Segment	Distance (mm)	Interval (ms)	NCV (m/s)	NCV N/D
Median, R								
Wrist	3.52ms	6.020mV	7.901mVms	Wrist - Cub. Fossa	200mm	4.12ms	48.5m/s	
Cub. Fossa	7.64ms	5.780mV	7.622mVms					
Ulnar, R								
Wrist	2.74ms	5.530mV	10.54mVms	Wrist - Blw. Elbow	240mm	3.96ms	60.6m/s	
Blw. Elbow	6.70ms	5.270mV	9.869mVms	Blw. Elbow - Abv. Elbow	90mm	1.92ms	46.9m/s	
Abv. Elbow	8.62ms	5.080mV	9.937mVms					
Ulnar, L								
Wrist	2.06ms	7.250mV	12.94mVms	Wrist - Blw. Elbow	240mm	3.86ms	62.2m/s	
Blw. Elbow	5.92ms	6.140mV	11.69mVms	Blw. Elbow - Abv. Elbow	90mm	1.83ms	47.9m/s	
Abv. Elbow	7.80ms	5.390mV	10.58mVms					
Median, L								
Wrist	3.70ms	7.880mV	13.95mVms	Wrist - Cub. Fossa	230mm	4.16ms	55.3m/s	
Cub. Fossa	7.86ms	6.560mV	12.19mVms					
Peroneal, L								
Ankle				Ankle - Fib. Neck	240mm		0m/s	
Fib. Neck				Fib. Neck - Lat. pop. fossa	90mm		0m/s	
Lat. pop. fossa								
Tibial, L								
Ankle	5.72ms	2.490mV	5.977mVms	Ankle - Pop. Fossa	400mm	9.08ms	44.1m/s	
Pop. Fossa	14.80ms	1.940mV	4.029mVms					
Tibial, R								
Ankle	6.62ms	3.380mV	5.070mVms	Ankle - Pop. Fossa	400mm	10.54ms	38m/s	
Pop. Fossa	17.16ms	2.370mV	5.990mVms					
Peroneal, R								
Ankle				Ankle - Fib. Neck	240mm		0m/s	
Fib. Neck				Fib. Neck - Lat. pop. fossa	90mm		0m/s	
Lat. pop. fossa								

MUSCLE BIOPSY



LOW RESOLUTION



HIGH RESOLUTION

Dx: Granulomatous myositis

Findings

Epidermis: unremarkable

**Dermis: perivascular infiltration
of mononuclear cells.**

**Muscle: dense infiltration of
acute & chronic
inflammatory cells.**

- **Multiple non-caseating granuloma composed of aggregates of epithelioid cells & multinucleated giant cells.**

LN Biopsy

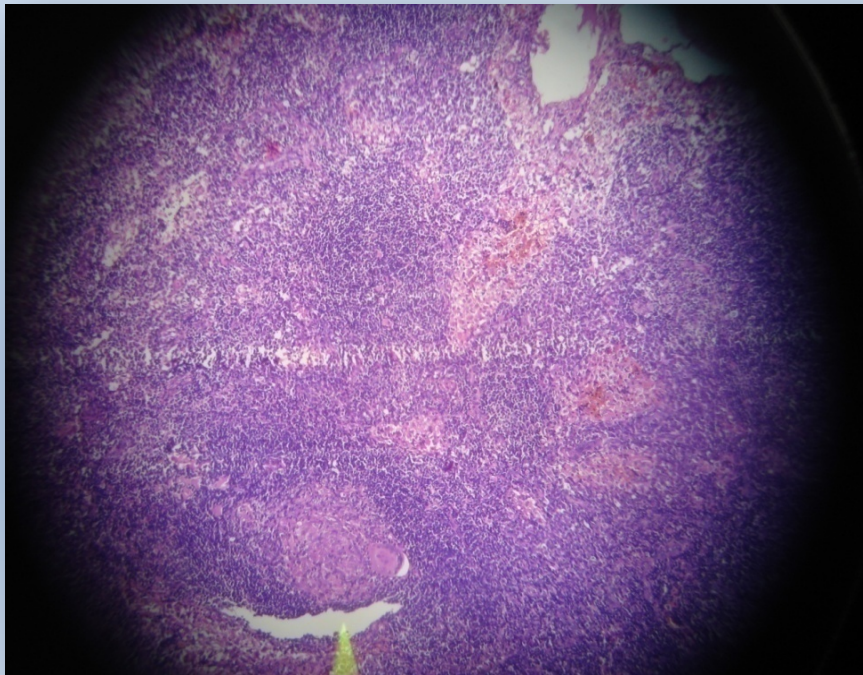


Findings

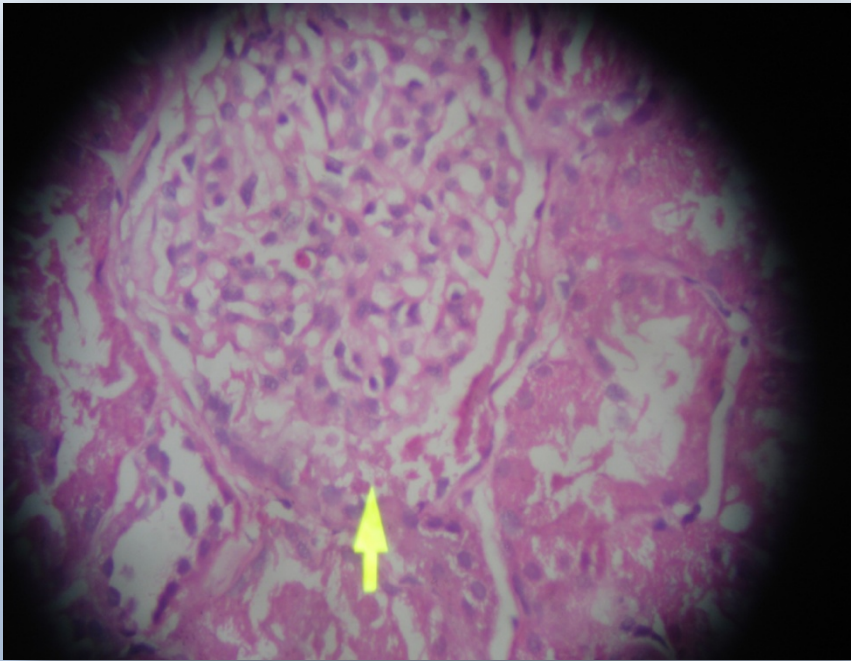
Multiple non caseating granulomas composed of aggregates of epithelioid cells & multi-nucleated giantcells

Dx:

Granulomatous lymphadenitis



Renal Biopsy



- H&E and PAS stain
All glomeruli are normal in respect to mesangial cellularity and basement membrane.
No granuloma present.
- DIF study
No deposit of IgA, IgM or C₃.

Dx:

Non proliferative glomerulopathy

MT



- **0 mm after 72 hrs.**

- **Serum angiotensin converting enzyme (ACE):**

417 U/L

(8-65 U/L normal value)

Grand Round

Final diagnosis:

Sarcoidosis

Treatment

- **Tab. Prednisolone : 1mg/kg body wt**
(S.F- 09.05.2015)
- **Cap.PPI (20mg) 1+0+1(before meal)**
- **Calcium, vitamin D**
- **DEXA scan advised**

Follow up after 15 days

Day -16 of steroid

Muscle tightness decreased

New involvement: both thenar

Paraesthesia: stocking pattern



Treatment

- **Dose of steroid was increased.**
- **Azathioprine was added.**

2nd FU
56th day of steroid



Wt: 58 kg

BP: 130/80 mmHg

Edema : absent

**Muscle weakness
was improving.**

Hb-15.6 gm/dl

FBS-4.9 mmol/L

RBS-5.2 mmol/L

Urine RME- P.C 0-2/HPF

E.C. 5-6/HPF

albumin : +

S. albumin: 34.1 g/ L

USG of KUB -Normal

3RD FU (5/11/2015)

6 months steroid intake



Steroid dose taper plan

- **Neuropathy : improved**

FBS-4.9 mmol/L

Random-5.3 mmol/L

- **Urine RME: P.C-0-2/HPF**
E.C. 1-3/HPF
albumin : nil

S. albumin: 46.2 g/ L

Before



After



Before



After



4th FU (27.01.2016)

8 months steroid intake

- **Patient is fully active & can do his daily activities without any problem.**
- **Neuropathy symptoms subsided. Size of lymph nodes decreased.**
- **Blood sugar profile normal.**
- **Oral prednisolone 45mg/day**
- **Advise: ACE level**
DEXA scan

Acknowledgements

- **Department of Surgery**
- **Department of Histopathology**
- **Department of Neurology**
- **Department of Nephrology**

Hasta La
Vista

