Clinicopathological conference

24 February 2011

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Diagnostician: to be announced

ผู้ป่วยหญิงไทยอายุ 94 ปี ปัจจุบันไม่ได้ประกอบอาชีพ ภูมิลำเนาและที่อยู่ปัจจุบัน จ.กรุงเทพฯ
2nd admission ประวัติได้จากผู้ป่วยและผู้ดูแล เชื่อถือได้

Chief complaint:

กระดกข้อเท้าไม่ขึ้น 2 วันก่อนมา รพ.

Present illness:

3 ปีก่อนมารพ. ชาฝ่าเท้าทั้งสองข้าง ชาหนักๆ เดินทรงตัวไม่ค่อยถนัดแต่เดินไม่มีรองเท้าหลุด รู้สึกปวดแปล๊บๆที่ปลายนิ้วเท้า

มาตรวจที่รพ.จุฬา ตรวจร่างกายพบ decreased pinprick sensation from both feet up to mid-pretibial areas , motor gr IV of both tibialis anterior, gastrocnemius, peronii, and extensor hallucis longus muscles, areflexia of both ankle and knee jerks

Electrodiagnostic test:

Sensory nerve conduction study: No SNAP response of both sural nerves and Rt superficial peroneal nerve. No response of Rt medial plantar mixed nerve study. Borderline DSL and low SNAP amplitude of both median nerve when stimulating at wrist and unobtainable SNAP when stimulating at elbows. Normal DSL and SNAP amplitude of Rt ulnar nerve when stimulating at wrist and unobtainable SNAP when stimulating at elbow

Motor nerve conduction study: No CMAP response of both common peroneal nerves. Prolonged DML, low CMAP amplitude and slow MNCV of Rt median and Lt posterior tibial nerves. Low CMAP amplitude but normal DML and MNCV of Rt posterior tibial nerve. Normal DML, CMAP amplitude and MNCV of Lt ulnar nerve

Imp: Sensory and motor axonal polyneuropathy with Lt median sensory motor neuropathy at the wrist (CTS)

CBC: Hb 13.0 g%, Hct 39.9%, MCV 78 fL, RDW 15.8%, WBC 2,760 cells/mm^3 (PMN 59.8%, L 32.6%, M 6.5%, E 0.7%, B 0.4%), Plt 195,000 cells/mm^3

ได้รับการรักษาด้วย gabapentin มีอาการง่วงนอนมากจึงหยุดยาไป อาการชาและอ่อนแรงเท่าๆเดิมมาตลอด ช่วยเหลือตัวเองพอได้

3 เดือนก่อนมารพ. รู้สึกว่าเท้าทั้งสองข้างไม่ค่อยมีแรงมากขึ้น เดินยกเท้าไม่ค่อยขึ้นแต่ใส่รองเท้าไม่หลุด สองข้างเป็นพอๆกัน แขนสองข้างปกติดี อาการเป็นเท่าๆเดิม ช่วยเหลือตัวเองได้

3 วันก่อนมารพ. ลื่นล้มในห้องน้ำ ข้อเท้าขวาพลิก ปวดข้อเท้าขวา จับประคองเดินพอได้ ไปตรวจที่รพ. ตรวจร่างกายพบ swelling with tender at Rt. lateral malleolus

2 วันก่อนมารพ. รู้สึกเท้า 2 ข้างไม่ค่อยมีแรงมากขึ้น กระดกข้อเท้าไม่ขึ้น ปวดเท่าๆเดิม ชาเท่าๆเดิม ญาติเห็นขาไม่มีแรงมากขึ้นจึงพามารพ.

ไม่มีไข้ ไม่มีน้ำหนักลด

Past history:

- HT Dx 2540 ที่รพ.จุฬา จากตรวจสุขภาพ
- Dyslipidemia Dx 2540 ที่รพ.จุฬา จากตรวจสุขภาพ
- Osteoporosis Dx 2548 ที่รพ.จุฬา present ด้วยลื่นล้ม x-ray มี osteopenia, เคย work up BMD: L spine z-score -2.4, hip z-score -3.6
- ปี 2552 check up CXR, asymptomatic → พบ Rt hilar nodule → CT chest : multiple small bilateral paratracheal, prevascular, subcarinal and bilateral interlobular nodes, up to 1.1 cm, no mediastinal mass. Cardiomegaly with scatter small calcified wall of Rt coronary and Lt anterior descending artery are noted. Scattered small calcified wall of aortic arch and thoracic aorta, atherosclerotic change, are observed. The pericardium appears normal. The trachea and main bronchi are patent. Both lungs and pleural spaces are clear. There are 2 small hypodense lesions in thyroid gland, a 0.8-cm one in Rt lobe and a 0.2-cm one in Lt lobe. The limited study of upper abdomen shows few small hypodense lesions in the liver up to 1.3 cm. The spleen is enlarged with a 0.6-cm hypodense lesion in posterior aspect. There are 2 renal cysts, a 1.6x7.4-cm on in Lt kidney and a 0.4-cm one in middle pole of Rt kidney. Bilateral adrenal glands appear unremarkable. There is neither osteolytic nor osteoblastic lesion.
- Current medications: amlodipine(10) 1x1, metoprolol(100) 1x1, HCTZ(50) 1x1, Moduretic 1/2x1

Personal history:

- ไม่ดื่มสุรา
- ไม่สูบบุหรี่
- ปฏิเสธประวัติยาชุด,ยาหม้อ,ยาลูกกลอน

Physical examination

General appearance: an elderly Thai female patient, full consciousness

Vital sign: BT 37 °C, PR 82/min, RR 18/min, BP 120/70 mmHg

Skin: no skin lesion, no rash

HEENT: mildly pale conjunctivae, no icteric sclerae

Lymph node: no lymphadenopathy

RS: trachea in midline, good air entry, equal breath sound, no crepitation, no wheezing

CVS: apical impulse at Lt. 5th ICS, MCL, no heave, no thrill, normal S_1S_2, no murmur

Abdomen : no distention, normoactive bowel sound, soft, not tender, liver can't be palpated, spleen just palpable

Extremities: Rt. ankle swollen, tender, no deformity

Neuro exam:

Consciousness: fully conscious, oriented to time, place and person

CNs: pupil 2 mm RTL both eyes, full EOM, no facial weakness, tongue and uvula not deviated

Motor:

Muscle mass: no muscle atrophy

No fasciculation

Power	Rt	Lt
Deltoid	IV+	IV+
Bicep/tricep	IV+/IV+	IV+/IV+
Wrist F/E	IV+/IV+	IV+/IV
Handgrip	IV+	IV+
Iliopsoas	IV	IV
Gluteus maximus	IV+	IV+
Gluteus medius	IV+	IV+
Adductor maximus	IV+	IV+
Quadriceps	IV+	IV+
Hamstring	III+	IV
Tibialis anterior	II	I
Gastrocnemius	III	IV
Tibialis posteior	0	I
Peronii muscle	0	0
EHL	0	0

Tonicity: flaccid tone both legs

DTR: 0 except both tricep jerk 1+

BBK: plantar response bilateral, clonus: not sustaining

Sensory:

Pinprick sensation: decreased pinprick sensation at both legs and feet

Proprioception: impaired both feet

Meningeal signs: no neck stiffness

Cerebellar signs: no ataxia, normal finger to nose test, normal dysdiadokokinesia test

ANS: normal rectal sphincter tone, positive anal wink reflex

Investigation:

CBC: Hb 10.2 g%, Hct 29.4%, MCV 72 fL, RDW 15.7%

WBC 1,730 cells/mm^3 (PMN 65.3%, L 26%, M 8.1%, E 0%, B 0.6%)

Plt 185,000 cells/mm^3

PBS: RBC- hypochromic-microcytic anemia, no microspherocyte, no MAHA blood picture

WBC- decreased, PMN predominate, no abnormal cell, no blast, no plasma cell

Plt- adequate, no clumping

UA: sp.gr 1.040, pH 6.5, protein trace, glucose negative, WBC 5-10/HPF, RBC 1-2/HPF squamous epithelium 0-1/HPF, no cast

24hr urine protein 0.42 g/d

Fasting plasma glucose 85 mg/dL

BUN 11 mg/dL, Cr 0.44 mg/dL

Electrolytes: Na 122 mmol/L, K 3.5 mmol/L, Cl 93 mmol/L, HCO_3 24 mmol/L

LFT: total protein 6.1 g/dL, albumin 2.8 g/dL, TB 0.50 mg/dL, DB 0.30 mg/dL, AST 43 U/mL, ALT 19 U/mL, ALP 130 IU

Corrected Ca 8.78 mg/dl, Mg 0.76 mmol/L(0.7-1.7), phosphate 2.8 mg/dL(2.7-4.5)

LDH 538 U/L, uric acid 3.5 mg/dL

ESR 10 mm/hr

TSH 1.44 mU/mL(0.3-4.1), FT4 1.34 ng/mL(0.8-1.8), FT3 2.14 pg/mL (1.6-4.0)

Morning cortisol 33.8 ug%

Anti HIV: negative

HBsAg: negative, antiHBs: negative, antiHBc: negative

AntiHCV: negative

ANA: < 1: 80

Rheumatoid factor: negative

BMA:

Cellularity: mildly hypercellular

M:E = 3:1

Iron 0

Ringed sideroblast: negative

Megakaryopoiesis: normal

Granulopoiesis: promyelocyte 9%, myelocyte 21%, metamyelocyte 14%, PMN+band 45%, eosinophil 2%

Erythropoiesis: normal

Lymphopoiesis: normal

Abnormal cells: not seen

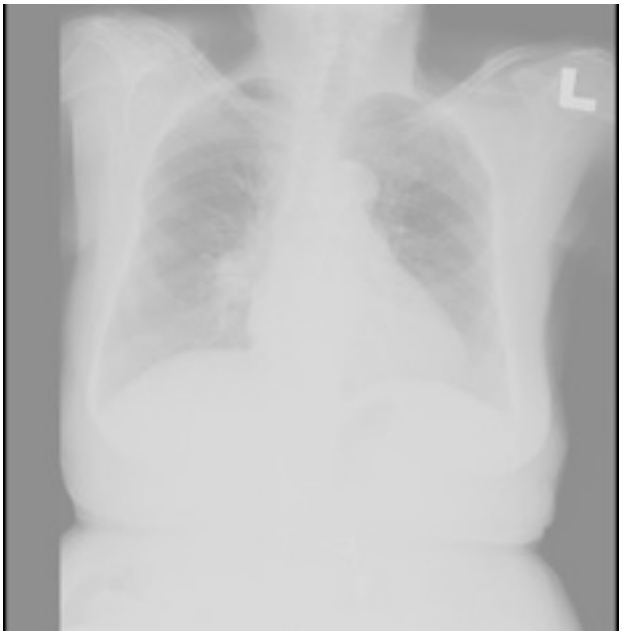
BM biopsy:

-hypercellular trilineage marrow with histologically normal maturation

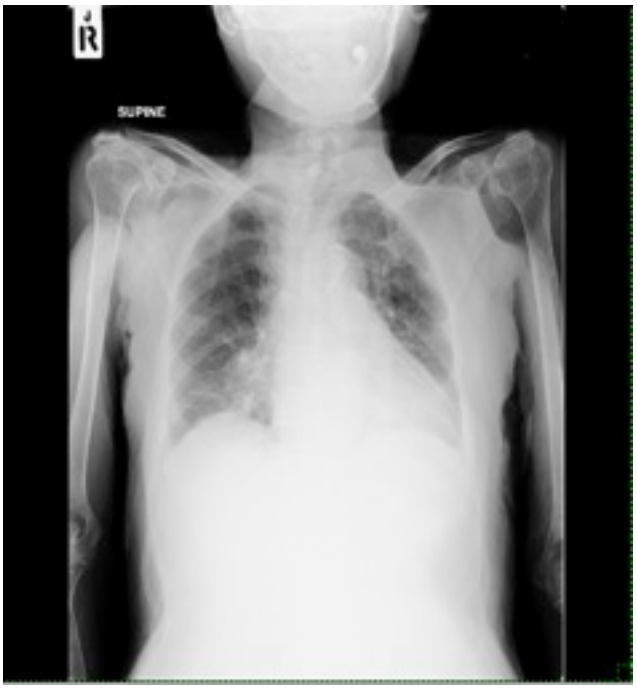
-no histological evidence of hematologic malignancy

-negative result of amyloid detected with Congo red stain under polarized light

CXR:

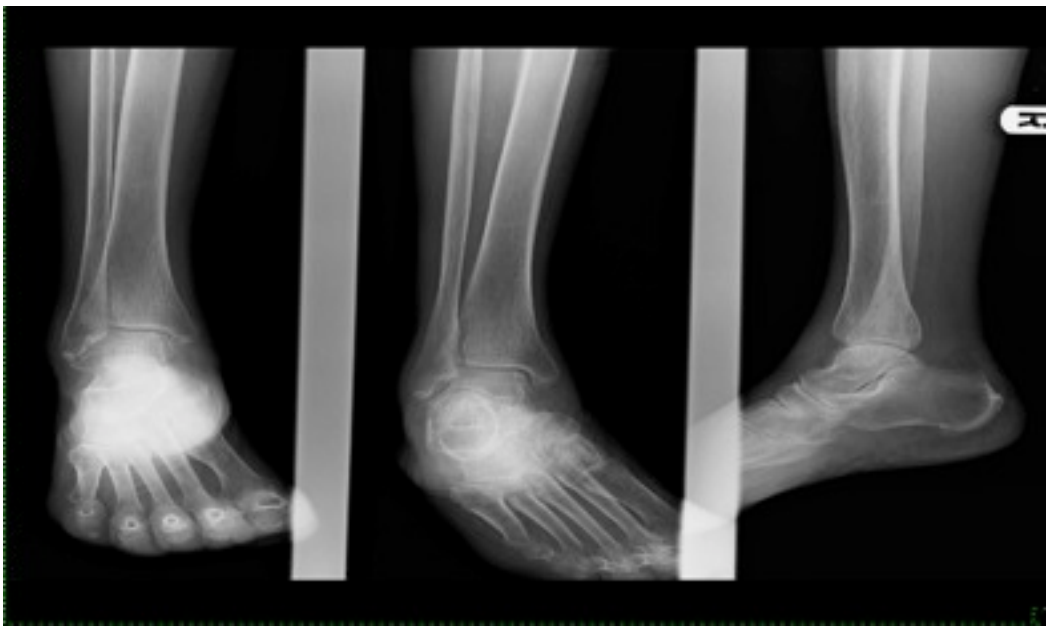


(CXR 2 years ago)



(at admission)

Rt. ankle:



CT chest:

There is subpleural reticulations and pleural thickening in both basal lungs, no significant change from Feb 2009

Recent segmental atelectasis is noted in LLL.

Recent minimal pleural effusions are seen, bilaterally.

No definite discrete pulmonary nodule is detected.

Trachea and main bronchi are patent.

Multiple nodes are seen at Lt prevascular, pretracheal, precarinal and bilateral hilar areas, size up to 1.2 cm at precarinal area.

No supraclavicular node is noted.

Minimal pericardial effusion is seen. Cardiomegaly is noted.

Few nodules are seen in both lobes of thyroid gland, size up to 1.3 cm on the Lt side.

Healed fracture of Lt posterior 10^{th} rib is noted

CT whole abdomen:

The study shows normal parenchymal attenuation. The are multiple cysts scattered in Rt and Lt hepatic lobe, size ranging 0.3-1.2 cm. No abnormal enhancement is seen. Small amount of ascites in gastrohepatic area is seen.

Bile ducts and CBD are not dilated.

Gallbladder is well-distended without gallstone.

Splenomegaly is seen with a 1.3 cm cyst at upper pole.

Pancreas and adrenal glands are unremarkable.

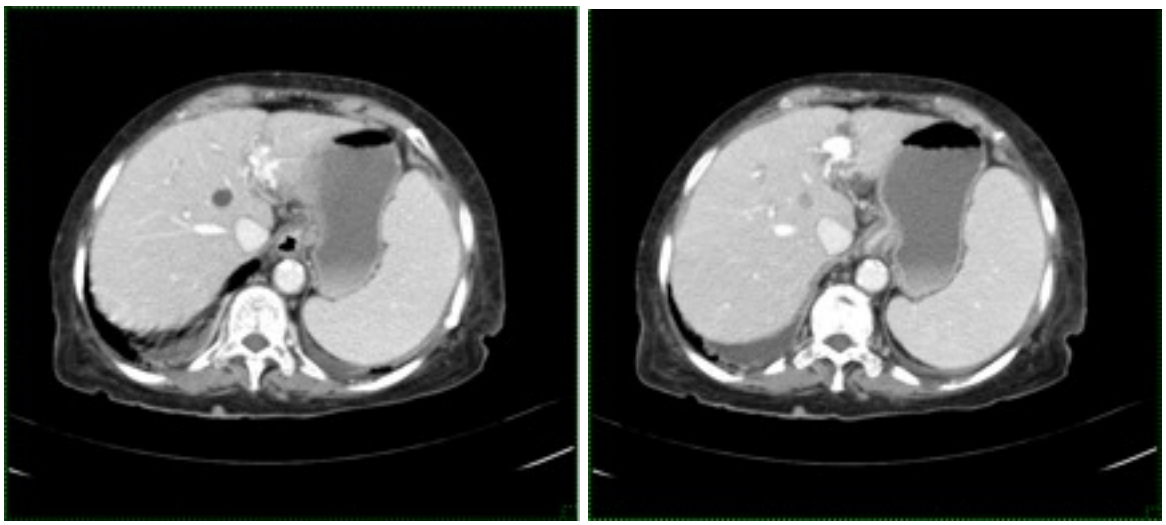
Cysts are seen in both kidneys, size up to 7 cm at Lt lower pole.

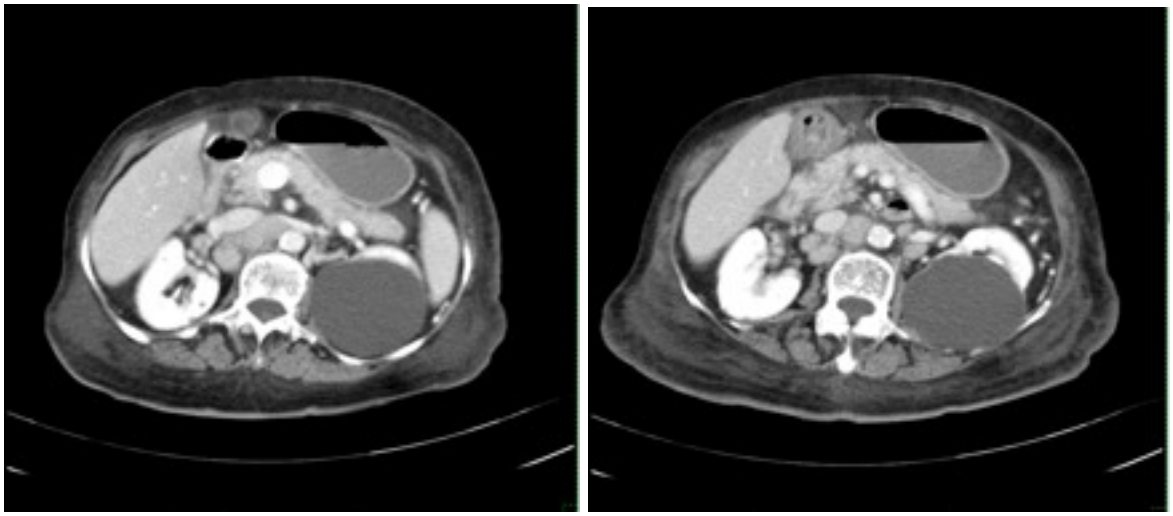
No hydronephrosis, stone or mass is seen.

Diverticulosis at sigmoid colon are noted.

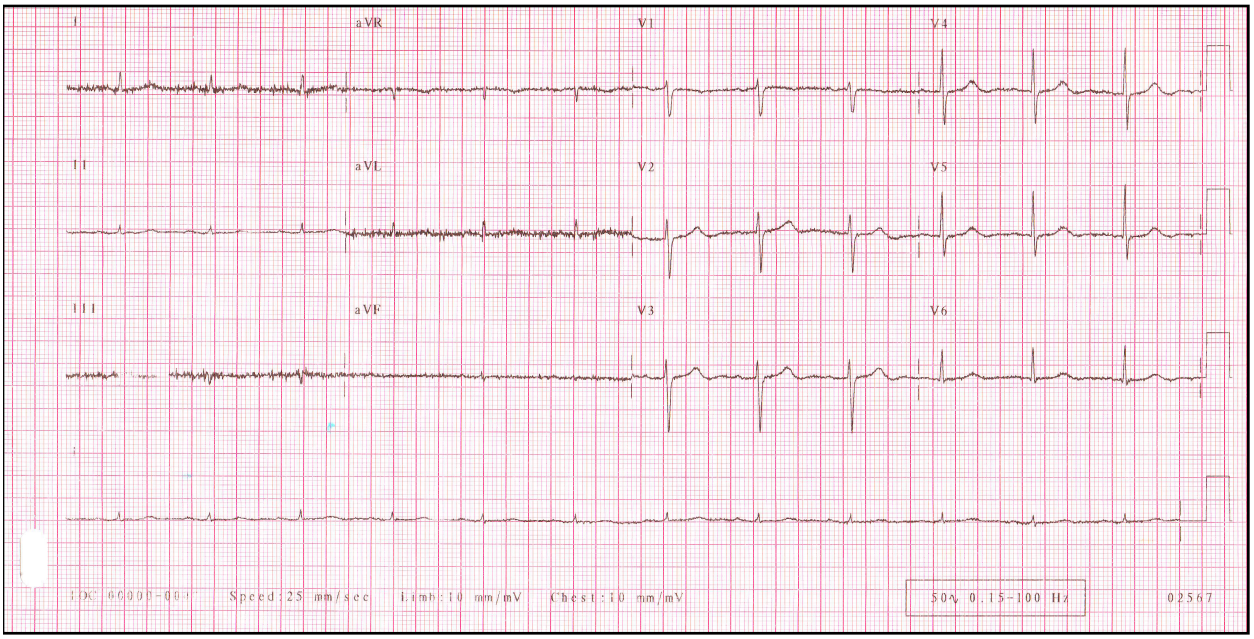
Stomach and bowel loops are unremarkable.

Multiple enlarged nodes are seen at aortocaval and precaval areas, size up to 1.7 cm in short axis





EKG 12 leads:


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
Speed:25 mm/sec
Limb:10 mm/mV
Chest:10 mm/mV
50Hz 0.15-100 Hz
02567

Electrodiagnostic test:

Sensory NCS

Nerve/sites	Rec. site	Onset Lat (ms)	Pk Amp (μV)	Response	Distance	Velocity (m/s)
Rt. ULNAR - Digit V						
1. Wrist	V			No		
Lt. ULNAR - Digit V						
1. Wrist	V	3.05	13.6		12	39.3
Rt. RADIAL - Wrist						
1. Forearm	wrist	2.85	5.7		12	42.1
Rt. SURAL - Lat Malleolus						
1. Calf	Lat Malleolus			No		

Motor NCS

Nerve/Sites	Rec. Site	Onset Lat (ms)	Amp (mV)	Total Dur (ms)	Velocity (m/s)	Distance (cm)
Rt. ULNAR - ADM						
1. Wrist	ADM	4.20	2.5	25.45		
2. B.Elbow		7.50	1.9	28.20	60.6	20
3. A.Elbow		9.75	2.0	80.00	44.4	10
Lt. ULNAR - ADM						
1. Wrist	ADM	3.30	6.8	28.30		8
2. B.Elbow		7.35	6.3	27.55	49.4	20
3. A.Elbow		9.40	5.5	27.90	48.8	10
Lt. TIBIAL - AH						
1. Ankle	AH		NR			
Rt. TIBIAL - AH						

Nerve/Sites	Rec. Site	Onset Lat (ms)	Amp (mV)	Total Dur (ms)	Velocity (m/s)	Distance (cm)
1. Ankle	AH		NR			
Lt. COMM PERONEAL - Tib Ant						
1. Fib head	Tib Ant		NR			
Rt. COMM PERONEAL - EDB						
1. Ankle	EDB		NR			
Lt. COMM PERONEAL - EDB						
1. Ankle	EDB		NR			

Needle EMG

	Spontaneous				MUAP			Recruitment
	1A	Fib	PSW	Others	Amp	Dur	PPP	Pattern
Rt. TIB ANTERIOR	N	None	+		N	Increased	N	Reduced
Rt. LUMB PSP lower L4L5 L5S1	N	None	None					N
Lt. LUMB PSP lower L4L5 L5S1	N	None	None					N
Lt. GASTROCN (MED)	N	None	+		N	Decreased	N	Reduced

Summary of finding

1. sensory nerve conduction study shows unobtainable Rt sural and Rt ulnar SNAP amplitudes, Rt superficial radial SNAP is of reduced amplitude and slow conduction velocity, Lt ulnar SNAP is of borderline low amplitude and slow conduction velocity
2. motor nerve conduction study shows unobtainable bilateral tibial and bilateral peroneal-EDB and Rt peroneal TA CMAP amplitude. Lt ulnar MNCS is normal. Rt ulnar MNCS shows low distal CMAP amplitude, prolonged distal latency and slow conduction velocity across the elbow

3. EMG of bilateral lower lumbar PSP muscles shows no spontaneous activities. EMG of Rt TA and Lt medial head of gastrocnemius shows rare spontaneous activity in the form of PSW. MUAPs are rarely recruited due to sever weakness

Interpretation:

The findings show electrodiagnostic evidences of large fiber sensory and motor axonal polyneuropathy. In addition, Rt ulnar neuropathy at the elbow is noted.

No electrodiagnostic evidences of lumbosacral radiculopathy. However, since this test is done at three days of weakness onset, thus, denervation potentials may not be demonstrated in this period. Follow up test may be considered.

Questions

1. What is the most likely diagnosis?
2. What is/are the investigation(s) leading to the final clinical diagnosis?