CLINICAL PEARL

A 56 YEAR OLD MAN WITH SWALLOWING AND SPEECH DIFFICULTY.

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PARTICULARS OF THE PATIENT

Name: Anwar Hossain.

Age: 56 Years

Sex: Male

Religion: Islam

Marital Status: Married

Occupation: Businessman.

Address: Badda, Dhaka.

Date of Admission: 04.09.2017

Date of examination: 04.09.2017

CASE SUMMARY

Mr. Anwar Hossain, 56-year-old, Married, muslim, businessman, hailing from Badda, dhaka was admitted in DMCH on 04.09.2017 with the complaints of

- Swelling and enlargement of the tongue for one year.
- Low back pain for one year.
- Shortness of breath for one year.

According to the statement of the patient he was reasonably alright about one year back. Then he developed gradual swelling and enlargement of the tongue. He complained repeated accidental tongue bite because of this enlargement. He also reported having difficulty in swallowing, eating, sleeping and had disturbed speech manifested as lisping but he denied sialorrhoea or noisy breathing.

- After several days he noticed multiple ulcerations all over the tongue which were small and painful. The pain was so intense that he developed difficulty in swallowing and slurring of speech. He visited several local physicians for this problem but his condition did not improve.
- For last one year he had been experiencing lower back pain which was gradual in onset, localized, continuous and dull aching in nature.

 Pain was aggravated by movement and relieved after taking medications. Pain was not radiating to thigh, leg or feet, not associated with tingling or numbness. He had long standing bilateral knee joint pain and walking difficulty. Sometimes both knees became so painful that he needed to use walking aid. No history of pain in any other joints.

- He mentioned having exertional breathlessness for last one year which was initially mild but gradually increasing in severity. He also noticed occasional orthopnea and paroxysmal nocturnal dyspnea. He developed ankle swelling for same duration which was persistant but decreassed after taking medications.
- On query, he stated having occasional passage of loose stool without any blood which resolved spontaneously

- He has no history of any kind of trauma to the tongue prior to this illness. No headache, fall or vision problem.
- No fever, weight loss, cough, palpitation, jaundice, cold intolerance, constipation, hoarseness of voice and contact with TB patient.
- No bleeding from any site.
- His bladder habit and sleep pattern was normal.

- He was hypertensive for Last 12 years but nondiabetic and non asthmatic.
- He had IHD and PCI was done to LAD and RCA about 5 years back.
- He had vitiligo for last 24 years.
- He was taking anti ischemic medications and diuretics, analgesics and antihypertensive drugs regularly.

- He is not known to be allergic to any medication or substances.
- He was not fully immunized according to EPI schedule.
- He is a smoker for last 30 years (15 pack year) but nonalcoholic.
- He belongs to middle class family.
- All of his family members are in good health

PHYSICAL EXAMINATION

GENERAL EXAMINATION

- Appearance:
 - The patient was ill looking but normal facial appearance.
 - Body built : Average.
 - He was cooperative.
 - Decubitus: On Choice

GENERAL EXAMINATION (CONTINUED...)

OTHER GENERAL EXAMINATION FINDING:

- Anaemia: Mild
- Cyanosis : Absent
- Clubbing: absent
- Koilonychia: absent.
- Leukonychia : Absent
- Edema: Bilateral pitting ankle edema.
- Dehydration: Absent

GENERAL EXAMINATION (CONTINUED...)

- Skin condition: Multiple hypopigmented macules were present all over his body and around oral cavity which were painless.
- Hair: Normal.
- Nail: Normal.
- Neck vein: Not engorged.
- Thyroid gland: Not enlarged
- Lymph Node: Not palpable.

GENERAL EXAMINATION (CONTINUED...)

VITAL SIGNS:

- Pulse: 80 beats /min, regular.
- Blood Pressure: 130/80 mm Hg, No postural drop.
- Temperature 98 F.
- Respiratory Rate- 18 breaths/min.
- Bed side urine examination : Positive.

ALIMENTARY SYSTEM EXAMINATION

Examination Of Tongue and Oral cavity:

The tongue was red on dorsal and ventral surface. Dorsal surface was rough due to overgrowth and swelling of fingerlike papillae.



ALIMENTARY SYSTEM EXAMINATION:

- There was generaliged enlargement of the tongue with indentation on the lateral border due to pressure from tooth. There was no fasciculation and tongue deviation. Multiple small, nodular and plaque like ulcers were present all over the tongue which were painful.
- Angular stomatitis was absent. There was no tooth spacing.

ALIMENTARY SYSTEM EXAMINATION:

PER ABDOMEN

- Abdomen was soft, nontender, nondistended.
 There was no palpable mass.
- □ No other organomegaly.
- Shifting dullness was absent.
- Bowel sound was present.
- □ D/R/E : Normal.

MUSCULOSKETAL SYSTEM EXAMINATION

Exam of Knee Joints:

- □ **LOOK:** No muscle wasting, no visible swelling or colour change of the skin over the joints.
- FEEL: There was mild tenderness and crepitations were palpable over both knee joints.
- MOVE: Flexion and extension of both knees were restricted due to pain.
- Examination of back and other joints revealed normal findings.

RESPIRATORY SYSTEM EXAMINATION

Shape of the chest and movement- Normal & symmetric. Trachea- Central,
Vocal fremitus - Normal
Percussion note was resonant.
Breath sound - Vesicular with few **bilateral**

basal crepitations.

Vocal resonance- Normal.

NERVOUS SYSTEM EXAMINATION

- Patient was fully conscious, oriented to time, place and person.
- His memory and intelligence was normal. Cranial nerve examination including fundoscopy revealed normal findings.

NERVOUS SYSTEM (CONT....)

Motor examination

Muscle power 5/5 in all four limbs with normal deep tendon reflexes.

All modalities of sensation were intact and coordination was normal.

Gait : Antalgic gait

Examination of Other Systems Reveal No Abnormality.

PROBLEM LISTS:

HISTORY

- Enlargement of tongue.
- Multiple painful tongue ulcers.
- Low back pain
- Exertional breathlessness
- Leg swelling
- Occasional loose stool.
- Knee joint pain

EXAMINATION

- Anaemia
- Ankle edema.
- Enlarged, red tongue with Multiple ulceration.
- Bilateral basal crepitations.
- Vitiligo.

Expert suggestion:



- Provisional diagnosis?
- What should be our plan of Investigations?

COMPLETE BLOOD COUNT

COMPLETE BLOOD COUNT	RESULT.
Hb level	10.3 g/dl
RBC	3.6 million/ cumm
WBC	8360/cumm, N- 59.4 %, L – 34.3%
	M – 33.1 %, E – 1.6 %, B- 0.1 %
Platelet	60,000 / cu mm
MCV	93 fl
PCV	33.8 %
ESR	71 mm in 1st hour.

PERIPHERAL BLOOD FILM

- RBC : Normocytic normochromic anaemia.
- WBC : Mature with normal count and distribution.
- Platelet : Reduced.
- Comment: Normocytic normochromic anaemia with thrombocytopenia.

RENAL FUNCTION TEST

TEST	RESULT	REFERENCE
Serum creatinine	1.06 mg/dl	0.18 -0.94 mg/dl
Serum urea	16 mg/dl	7-20 mg/dl
Urine R/M/E	protein +	

INVESTIGATIONS(contd.....)

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S.Calcium : 9.0 mg/dl
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Serum electrolytes: Na - 135 mmol/l

K - 5.1 mmol/l

Cl - 110 mmol/1

INVESTIGATIONS(Contd.....)

S.Uric acid : 4.60 mg/dl

♦ S.TSH : 2.6 mIU/L

Serum T3 : 90 ng/dl

Serum T4 : 1.5 ng/dl

RA test : Negative.

CRP: 1.0 mg/dl

ANA : Negative

Anti CCP Ab: Negative

LIVER FUNCTION TEST

TEST	RESULT	REFERENCE
Serum Bilirubin	0.8 mg/dl	0.18 -0.94 mg/dl
SGPT	88 U/L	Up to 40
Alkaline phosphatase	45 U/L	40-125 U/L
Prothrombin time	14 sec	12 - 16
INR	1.46	
Serum albumin	4.00 gm/dl	3.6 - 5.20

INVESTIGATIONS

ECG: Normal Study.

Chest X ray: Normal study.

Skull X ray: Normal study

X-ray lumbo sacral spine: Normal findings.

X-ray Knee (Right and Left): Joint space narrowing and formation of osteophytes.

Suggestive of Osteoarthritis.

X-RAY SKULL:



ECHOCARDIOGRAPHY

- Septal, Anterior & Lateral wall hypokinesia (OMI)
- Moderate LV systolic dysfunction.
- Borderline concentric LVH.
- □ **EF** 38%

INVESTIGATIONS(Contd.....)

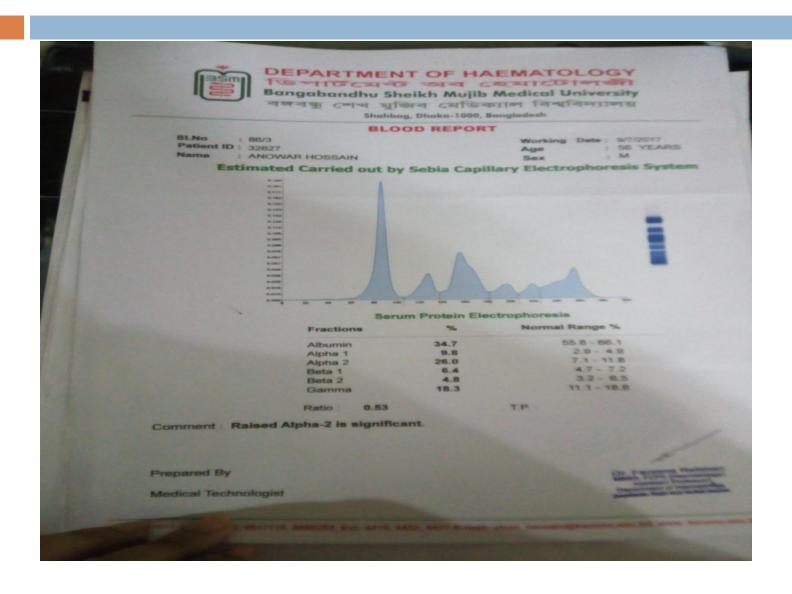
- USG of Whole Abdomen : Normal
- USG of tongue
 - i) Soft tissue structure is enlarged
 - ii) Echotexture is non-homogenous
 - iii)Multiple tiny hypoechoic nodules are seen in the musculature

Comment: Macroglossia

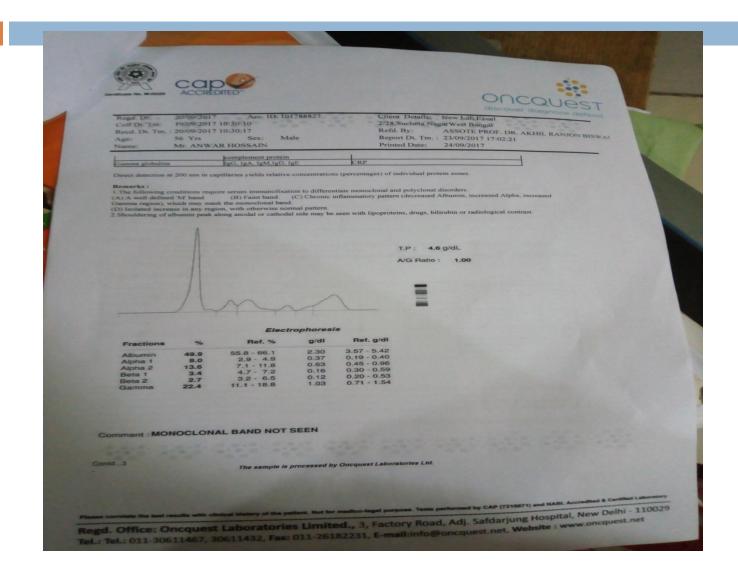
INVESTIGATIONS(Contd.....)

Plasma protein electrophoresis:
 Raised Alpha 2 is significant (Bangladesh)
 Monoclonal band not seen. (India.)

PLASMA PROTEIN ELECTROPHORESIS



PLASMA PROTEIN ELECTROPHORESIS



TONGUE BIOPSY

Microscopic Description:

Sections show a small piece of tissue lined by Stratified Squamous Epithelium. The sub epithelial tissue shows focal hyalinization with deposition of eosinophilic structure less substances morphologically resembling Amyloidosis.

Comment: Amyloidosis

Congo red stain Of biopsy material with polarized light: **Positive.** (Apple green amyloid under microscopy)

FINAL DIAGNOSIS

Primary Systemic Amyloidosis with Osteoarthritis of both knee joints with IHD.

TREATMENT

Treatment:

Correction of anaemia

Other symptomatic treatment.

Specific: Chemotherapy with

Bortezomib with

Dexamethasone (VD)

Cyclophosphamide(VCD)



Thank you