

From: "ROOT" <root@sctimst.ac.in>
To: "ROOT" <root@sctimst.ac.in>
Date: 30/10/2024 07:53 AM
Subject: CPC Clinical Protocol 30.10.2024

Dear All,
Season's Greetings.

The next Wednesday CPC of the session will be held on **October 30, 2024** at 08.00 hours (IST) in **Lecture Theatre 1**, Nehru Hospital, PGIMER, Chandigarh.

The session will also be available on the Webex platform. Kindly follow the link below to join.
<https://telemedicine.webex.com/telemedicine/j.php?MTID=mb8f5c93d0efe86e2ec7df721b3c21227>

In case you join in thru WebEx, kindly **ensure that your microphone and camera are switched off** and **PLEASE DO NOT SHARE YOUR SCREEN**.

The Clinical handout of the case to be discussed is attached herewith.
The clinical protocol will be discussed by **Dr. Gaurav Prakash, Department of Internal Medicine**. Radiology will be discussed by **Dr(s). Manphool Singhal/ Dr. Madhurima Sharma**. Autopsy pathology will be presented by **Dr. Ritambhra Nada**.

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Yours sincerely,

Staff CPC 30-10-2024

Patient: SS, 53/M

Labourer from Punjab

DOA:22/7/24, **DOD:** 27/724

Chairperson

Clinicals by:

Radiology by:

Dr Sanjay Jain

Dr Gaurav Prakash

Dr Manphool Singhal/ Dr.

Madhurima Sharma

Dr Ritambhara Nada

Clinician I/C: Dr Pankaj Malhotra

Pathology by:

First admission Under Nephrology services

Presenting complaints (1st March 2024-20th March 2024) Progressive bilateral pedal oedema for 1 year associated with periorbital swelling and frothy urine. He reported decreased urine output (decreased for last 3 months 200-300 ml/24 hour) along with history of progressive breathlessness on exertion (NYHA I→III). It was insidious in onset and associated with cough without expectoration, no chest pain, no orthopnea or PND. Took indigenous medicines for 2- 3 months before admission. Prior to this admission, he was evaluated in cardiology OPD and found to have DCMP. Coronary angiography was normal, hence he was sent to Nephrology OPD.

Past History: No history of diabetes, hypertension, TB, or any other hospitalization, major surgery or allergy in the past.

Personal History: Predominantly vegetarian diet, Alcohol, tobacco smoking and chewing; occasional for 15-20 years

Examination: BP min 86/56 mmHg, max100/74mmhg, RR 22/minute, HR 110/minute, SpO2 – 98%@RA. Oral Cavity – normal, generalized edema++, scrotal edema+

Liver palpable- 3-4 cm below costal margins, Spleen-NP; No S3 or Cardiac murmur

Investigations (In the first admission)

Inv	11/3/24
Hb, g/dL	8.8
MCV	60
TLC, (*10 ⁹ /L)	8120
DLC (N/L%)	57%/23.8%
Platelets, (*10 ⁹ /L)	213
BU/Creat, mg/dL	88/2.29
Bilirubin, mg/dL	0.27
Prot/Alb, mg/dL	4.7/2.72
AST/ALT, U/L	29/22
Alk Phos, U/L	153
Ca/PO4, mg/dL	8.42/5.73
Magnesium, mg/dL	2.2
TC: 275, LDL: 196, TG: 156,	

URE- pH 7.5, Protein 4+, Glu- nil, RBC-nil, Pus cells 3-5/hpf

24h UP: 5.08 gm/day,

AntiPLR2 antibody- negative

ProBNP: 15771

SPEP:0.15g/dl

SIFE: IgA Lambda

S.FLCA: $\kappa:\lambda=74.69:180$ Ratio-0.41

RENAL BIOPSY (5.3.24, S-6527/24): AL amyloidosis, λ restricted

RECTAL BIOPSY (13.3.24, S-7244/24): non-specific inflammation

BM biopsy (19.3.2024, A-693/24): 4% plasma cells with amyloid deposits

Cardiac MRI (18.3.24, PG248611): s/o Amyloidosis

FBG= 80mg/dl, HbA1c=5.9g/dl

ANA-neg, Sputum AFB- neg

HPLC- Beta Thalassemia trait

Imaging:

2D echo (March24) – E>>A, LVEF-45-50%, mild MR, TR, RVSP=RAP+20mmhg, Inter atrial septum 1.6 cm

CECT(chest +abdomen)(7/3/24) Early centrilobular emphysematous involvement in bilateral lung

Mild pericardial effusion (thickness 9 mm)

liver is enlarged measures 18.9cm with normal attenuation. No IHBRD, HV and PV are normal

Spleen- 11.9 cm normal attenuation

RK-10.4,LK9.5 shows normal outlines

Cardiac MRI- Concentric LV RV hypertrophy with EF 39%, Diffuse subendocardial enhancement

Treatment -

For AL amyloidosis, he was treated with VCD (Bortezomib, cyclophosphamide &Dexamethasone) and referred to the Department of Clinical Hematology and Medical Oncology. He completed 4 cycle of treatment (each cycle consists of weekly doses of all three agent).

Second admission (22/7/24-27/7/24) (Last admission) under CHMO services

Presenting complaint-

1. Loose stools, watery in consistency (Bristol Stool Scale 6-7). Over the past ten days

2. Followed by oliguria and worsening shortness of breath (mMRC grade from II to IV), along with anasarca and swelling in both lower limbs with skin changes.

No H/o blood in stool or undigested food particles. No history of chest pain

On Examination-Sick, tachypneic, pallor+, Anasarca+, PR 94/min, BP-90/60mmhg, RR 26/min

Chest- B/L Crepitations, Abdomen- hepatomegaly, no splenomegaly, bowel sounds+

ECHO- (May-2024) LV wall thickness 11mm, EF 50%, Mod MR,

Severe TR, RSVP=20+RAP

ECHO- (23-7-2024) Global Hypokinesia (LAD>LCx),visual EF 40-45% No MR AR TR, Mild Pericardial effusion

Date	23.07.2024	24.07.2024	25.07.2024	26.07.2024
Hb (gm%)	4.7	5.1	4.9	6.1
TLC($\times 10^9/L$)	3.8	2.6	2.95	5
Platelets($\times 10^9/cu.mm$)	18	11	6	14
Na / K	141/5.17	134/3.89	135.9/4.06	129/3.28
Urea/Cr	327/4.28	177/2.43	188.7/2.73	101/1.50
Ca/Ph/Mg	7.5/-	-	7.52/5.34/2.50	-
Bilirubin (T/C)	0.89/0.44	1.32/0.60	1.07/0.76	1.52/0.77
TP/Alb	4.3/1.82	4.7/1.84	4.28/2.03	4.9/2.05
OT/PT/ALP	95/75/-	88/63/-	71.5/52.1/193	106/55/-

DATED	24.7.24	27.7.24
pH	7.421	7.506
pCO2	34.7	27.3
pO2	26.3	40
HCO3	22.1	21.1
Lactate	5.47	4.89

Blood culture (23-7-24) collected after death of the patient. –

- (1) *Acinetobacter baumannii* (Pan-resistant:- Carbapenem, Colistin, Pip-Taz) ,
- (2) *Enterobacter faecium* (S- only Linazolid)

Imaging:

CXR- Rt Pleural effusion, Cardiomegaly

HRCT chest – Right sided pleural effusion with underlying consolidation in right lower lobe .

Uric acid 13mg/dl (on 25-7-2024)

Coagulogram- PT 16s (10-13s), PTI 72%, aPTT- 26

CRP- 28 mg/L

URE (27-7-2027)

- Protein +++
- Blood +

CKMB- 18.6 U/L – (7-25)

Hemolytic w/u

- Plasma Hb- nil
- Urine Hb- nil
- Haptoglobin 195(36-195)
- DCT- Negative

Course and management

In March 2024, a 53-year-old male presented with adult-onset nephrotic syndrome, heart failure, and hepatomegaly. He was diagnosed with systemic AL amyloidosis, with cardiac and renal involvement, and received four cycles of Bortezomib, Cyclophosphamide, & Dexamethasone. In July 2024, He was admitted again with complaints of diarrhoea, oliguria, and shortness of breath. Upon admission, his creatinine level was 4.28mg/dl, and his urea level was 328mg/dl. Due to low urine output and low BP, noradrenaline was started and dialysis support was provided. HRCT chest revealed right lower lobe consolidation with pleural effusion. IV antibiotics: cefoperazone with sulbactam in renal modified doses, changed to Imipenem and Vancomycin considering septic shock and indwelling dialysis catheter.

However, there was little improvement. He continued to require intravenous norepinephrine support and dialysis was continued with conservative fluid management. On July 27, 2024, the patient deteriorated, his sensorium worsened and he became hypoxic. He was intubated, and a second inotrope was introduced.

At 2:00 PM, the patient's vital signs became unrecordable. Cardiopulmonary resuscitation (CPR) was initiated. The patient was declared clinically dead at 2:36 PM.

Unit's final diagnosis- Primary AL Amyloidosis, with AOCKD, Amyloid related cardiac dysfunction, Sepsis

Cause of death- Septic shock