A RARE PRESENTATION OF PRIMARY POLYCYTHEMIA

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A 72 year old male presented to the emergency department of Chengalpattu medical college with complaints of chest pain and shortness of breath for a day.

HISTORY OF PRESENTING COMPLAINTS:

Chest pain for a day

Compressive in nature, retrosternal, radiating to left arm and neck,

Associated with sweating and palpitations

Patient Complaints of intermittent headache for past 1 month

Hemicranial involving right side , stabbing type of pain

Patient also gives history of tinnitus

No history of fever, cough

No history of abdominal pain / vomiting

PAST HISTORY:

Not a known case of SHTN / TYPE 2 DM

Not a known case of CAD

No history of any malignancy

PERSONAL HISTORY:

Mixed diet

Sleep and appetite – Normal

Bowel and bladder habits – Regular

Not a smoker or alcoholic

GENERAL EXAMINATION

Patient consious and oriented

Afebrile

Face : congested

Ear : No diagonal ear lobe crease

No pallor , icterus, cyanosis, clubbing

No pedal edema or lymphadenopathy

VITALS

BP – 130/90 mmHg, Recorded in all four limbs

PR – 84/min, regular in rythum

RR- 18/min

SPO2- 98 % in RA

SYSTEMIC EXAMINATION

CVS : S1 S2 Heard, No murmur

RS: Bilateral NVBS present,
No added sound

P/A : Soft, non tender

CNS: No neurological deficits

ECG taken at emergency department shows:

Normal sinus rhythm

HR - 75/min

Left axis Deviation

ST segment Elevation with T inversion in V1,V2,V3

ST depression in II,avF

Suggestive of AWMI

Kilip class –I

Cardiologist opinion : Thrombolysis with streptokinase

TREATMENT:

Loading dose of Aspirin, Clopidogrel and Atorvastatin

Inj Streptokinase 1.5 million IU in 100ml NS over 1 hr

Inj Heparin 5000IU IV Q6H

Inj Rantidine 50mg IV BD

T Aspirin 150mg 0-1-0

T Clopidogrel 75mg 0-1-0

T Atorvastatin 10mg 0-0-4

T Enalapril 2.5mg 1-0-1

T Metoprolol 12.5mg 1-0-0

INVESTIGATIONS

Complete Blood count

WBC - 34000 / cu.mm

RBC - 6.5 M / cu.mm

HB - 20.1 g/dl

HCT - 58 %

MCV - 88 fl

MCH - 30 pg

MCHC - 34 g/dl

PLT – 3.4 Lakh / cu.mm

Neutrophil - 54 %

Lymphocyte - 43 %

1. RFT & Electrolyte - Normal
2. LFT – Normal
3. CKMB - 97 IU/L
4. Chest X ray – Normal

PROVISIONAL DIAGNOSIS

ACUTE CORONARY SYNDROME/ STEMI/ ANTERIOR WALL MYOCARDIAL INFARCTION

POLYCYTHEMIA UNDER EVALUATION

Day 2

USG abdomen and pelvis:

Mild splenomegaly(12.5 cm)
Normal sized kidney with grade I RPD

** Patient c/o headache
Analgesic and adequate hydration given

Day 3

ECHO : Hypokinesia of Anterior wall
Mild LV dysfunction
EF : 54%

PS report : Normocytic Normochromic RBC
Increased number of RBC
Increased mature WBC
Erythrocytosis with leucocytosis

Day 4

- Serum uric acid – 9.2 mg/dl (Increased)

  Started on Tab. Allopurinol 200 mg
- BT – 3 min

  CT – 5 min

  PT – 14.2 sec

  INR – 1.1

Primary Vs Secondary Polycythemia

Serum erythropoietin : 5 mIU/ml (5 -15)

Primary

Day 7

BONE MARROW ASPIRATION:

Cellular marrow with trilineage hyperplasia

Megakaryocytes showing morphological abnormalities

Day 8

JAK 2 MUTATIONS : detected +

Diagnostic criteria (WHO)

MAJOR:

- Hemoglobin >16.5 g/dL in men and >16 g/dL in women or
  Hematocrit > 49% in men and > 48% in women or
  Red cell mass >25% above mean normal predicted value

- Bone marrow biopsy showing hypercellularity for age with trilineage growth (panmyelosis) including prominent erythroid, granulocytic and megakaryocytic proliferation with pleomorphic mature megakaryocytes

- Presence of JAK2V617F or JAK2 exon 12 mutation

MINOR:

- Serum erythropoietin level below the reference range for normal

Diagnosis of PV requires:

The presence of either all three major criteria or the first two major criteria and the minor criterion.

In our case:

All the 3 major criteria are fulfilled i.e.,

1) Hb - 20.1 and Hct - 58%

2) Bone marrow hypercellularity with trilineage hyperplasia

3) Presence of Jak 2 mutations

DIAGNOSIS

① PRIMARY POLYCYTHEMIA VERA

② MYOCARDIAL INFARCTION

TREATMENT GIVEN

- Phelebotomy – 300ml of blood taken
- Tab Hydroxyurea 200mg
- Tab Rantidine 150mg BD
- Tab paracetamol SOS
- Tab Aspirin 75mg and other cardiac drugs

THANK YOU