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ACUTE CHEST SYNDROME IN A YOUNG ADULT WITH SICKLE CELL ANEMIA: A CASE REPORT FROM MAHARASHTRA

General Medicine

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ABSTRACT

Case Report- Herein we present a case of 22 year old male, resident of odisha , presented with acute chest pain, breathlessness since 4 days without fever ,cough, cold or any other cardiac complaints. On clinical examination pallor and icterus was present. On systemic examination crepitations was present in left infrascapular region. CXR has evidence of patchy infiltrates in left middle zone and lower zone. On blood investigation normocytic normochromic anemia was present and few sickeled cells seen on peripheral smear and there was indirect hyperbilirubinemia present. Sickling test was positive and HPLC has 70% HBS levels. Patient was diagnosed as a sickle cell anemia in crisis of acute chest syndrome and treated with exchange transfusion ,oxygen support , hydroxyurea and hydration. Patient responded dramatically to exchange transfusion therapy.

KEYWORDS

acute chest syndrome, HPLC, sickling test, exchange transfusion,

INTRODUCTION:

Sickle cell disease is an autosomal recessive disorder caused by point mutation in beta gene of haemoglobin found on chromosome number 11 with substitution of sixth amino acid from glutamic acid to valine. Sickle cell disease is the most common structural hemoglobinopathy in heterozygous form . In south east asia region, highest prevalance is in india with 20 million patients. Sickle cell crisis is a life threatening condition in these patients. May present as vaso occlusive crisis with painful joints/ aspetic necrosis , retinal hemorrhages, acute renal failure, splenic sequestration crisis , hand foot syndrome, stroke, priapism, acute chest syndrome. In this, acute chest syndrome is a medical emergency accounts for 25% of all deaths.

History:

22 yr old male , resident of odisha , a construction worker a known case of sickle cell disease since 2 years presented to CPR hospital casualty with chief complaints of breathlessness since 4 days sudden in onset initially present on exertion which progressed to at rest since 1 day. Associated with chest pain sudden in onset, sharp pain localised to left side of chest, non radiating pain ,increased on inspiration associated with generalised weakness since 4 days. No h/o fever, cough ,cold. No h/o similar complaints in past and no h/o similar complaints in family members.

Clinical Examination:

Patient was conscious and oriented to T/P/P.

Pulse :100/min, Regular BP:100/60 mm hg ,Spo2: 91 %on RA ,RR: 30/min
Pallor and icterus was present. No other significant finding.

RS: Coarse Crepts present in left infrascapular region . Air entry normal on right side.

CVS: S1S2 + No murmur
P/A: soft and non tender, No evidence of organomegaly.

CNS: wnl

Investigations:

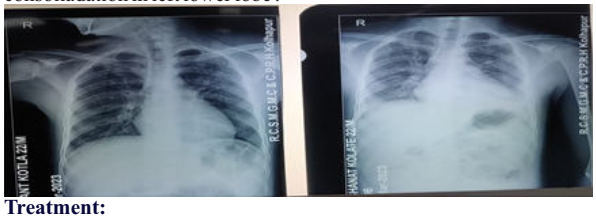
HB	9.8 gm%	BUN	13 mg/dl
HCT	27%	SR. CREAT.	0.5 mg/dl
MCV	82fl	T. B	11.7 mg/dl
MCH	28pg	D.B	0.5 mg/dl
MCHC	35gm/dl	I.B	11.2 mg/dl
RDW	15%	SGPT	58 IU/L
TLC	29600/cumm	SGOT	72 IU/L
DLC	82/14/02/02	SR. PROTEIN	7.0 gm/ dl
PLATELETS	4.1 lakhs/cumm	SR. ALBUMIN	4.1 gm/dl

PBS: Normocytic normochromic , few microcytes , few sickled cells, ovalocytes ,target cells seen.

Sr. LDH: 800 IU/L
Retic count: 0.6 %
ICT/DCT: Negative
USG (A+P): NAD
2D ECHO: LVEF 60% , Good biventricular function.
D-dimer : WNL
Sputum studies: No growth

For confirmation patient was evaluated further.
Sickling test : Positive
HPLC: 70 % HBS levels
CXR PA: Patchy infiltrates seen in left MZ and LZ
HRCT: Mild b/l pleural effusion more one left .

Passive atelectasis of lung parenchyma in bilateral lower lobes. Area of consoliadation in left lower lobe .



Treatment:

Oxygen support, Cap Hydroxyurea 500mg 2-1-1
Exchange transfusion protocol
Hydration(IV fluids) ,IV antibiotics and pain killers.

DISCUSSION :

Acute chest syndrome is a medical emergency that may require management in intensive care unit .Acute chest syndrome is thought to be due to in situ sickling within the pulmonary vasculature producing pain and temporary pulmonary dysfunction. Pulmonary infarction and pneumonia are frequent underlying or concomitant of this condition as reduction in arterial oxygen saturation promotes sickling on massive scale. Increased expression of adhesive molecules on endothelium and a role of HBS in scavanging of NO2 which in turn lead to raising pulmonary vascular tone are possible mechanism of acute chest syndrome.

CONCLUSION:

Acute chest syndrome is one of the sickle cell crisis is a medical emergency and it can be life threatening. High index of suspicion in known sickle cell disease presenting with acute respiratory symptoms

will lead to early diagnosis .If hematocrit <30% and saturation drops below 90% emergency exchange transfusion can be life saving in this patient so close monitoring and timely intervention is needed. Hydroxyurea is a drug of choice in this patient by increasing HBF levels ,RBC water content and altering adhesion of RBCS to endothelium , it plays important role in treating and preventing vasoocclusive crisis.

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