A Case Report on Vaso-occlusive Crisis in Complex Case of Sickle Cell Disease with Beta Thalassemia and Sexually Transmitted Disease

Twinkle Rathod[1], Mohit Buddhadev[2], S P Srinivas Nayak[2,*], Gunosindhu Chakraborthy[1]

[1]Parul Institute of Pharmacy and Research, Parul University, Vadodara, Gujarat, INDIA.
[2]Department of Pharmacy Practice, Parul Institute of Pharmacy and Research, Parul University, Vadodara, Gujarat, INDIA.

ABSTRACT

Painful episodes also known as sickle cell crisis are the key symptoms of patients suffering from Sickle cell disease. The presence of other life-threatening conditions such as Beta-thalassemia and Sexually transmitted disease can increase the risk of developing complications and premature deaths in patients. Our case study encountered a 17-year-old male having Sickle cell crisis with Beta-thalassemia and Sexually transmitted disease. Our finding has demonstrated that the etiologic interactions among these conditions were very much dependent upon genetics and transmission via contaminated blood. Managing such a complicated conditions can often bring challenges while planning for therapeutic management. Hence, the foremost goal for managing the patients is to control the signs and symptoms promptly and safely, ultimately improving the quality of life of the patients and decreasing further number of hospitalizations.

Key words: Sickle cell disease, Vaso-occlusive crisis, Beta-thalassemia, Sexually transmitted disease, Case report.

INTRODUCTION

The vaso-occlusive crisis, also known as sickle cell crisis, begins and is well assisted by the interactions between sickle cells, endothelial cells and plasma constituents.[1] Vaso-occlusion causes some of the complications such as pain syndromes, stroke, leg ulcers, spontaneous abortion and renal insufficiency in patients with sickle cell disease.[1] About 5.2 percent of patients suffering from sickle cell disease experiences 3 to 10 episodes of severe pain every year based upon epidemiologic data.[2] In most patients, a pain crisis resolves within five to seven days. Though, severe crisis may be presented with pain persisting for weeks to months.[2] Few factors, like dehydration, infection, strenuous exercise or activity, cold weather, serves as precipitating factors for acute Crisis.[3-5] Also, it is difficult to determine actual cause in many patients.[3] In this case, it is suspected with non-medical Adherence and HIV infection. The clinical severity of sickle cell-beta thalassemia depends on the percentage of the normal hemoglobin A, which can vary between 3% and 25%.[1-2] Eighty percent of cases of beta thalassemia in African-Americans is due to promoter region mutations that result in a phenotype in which HbA accounts for 18% to 25% of total hemoglobin.[6-8] Unlike those individuals with sickle cell disease who suffers from splenic infarction especially in childhood these patients continue to have splenomegaly in adulthood and are therefore susceptible to the development of the splenic sequestration crisis.[9]

DOI: 10.5530/ijopp.15.2.29

Address for correspondence:
Prof. S P Srinivas Nayak,
Assistant Professor, Department of Pharmacy Practice, Parul Institute of Pharmacy and Research, Parul University, Vadodara, Gujarat, INDIA.
Email id: spnayak843@gmail.com

www.ijopp.org

The Interaction between Sickle Cell Disease and HIV Infection: Human immunodeficiency virus (HIV) and sickle cell disease (SCD) are considered as endemic in overlapping geographic areas; however, in most of the countries very scanty of data and disease burden exists based upon the interaction between HIV and SCD. HIV prevalence in SCD patients varies between 0% and 11.5% in published studies. SCD has been suggested to reduce disease progression of HIV into AIDS.[10]

CASE PRESENTATION

A 17-year-old male patient was admitted with the complaints of pain in joints of upper and lower limbs since 1 day, chest pain since 1 day and back pain for 2 days. On second day of admission patient had a complaint of constipation.

PHYSICAL EXAMINATION

Upon physical examination, patient had mild pallor and icterus. On General examination the patient was fair, cooperative and coherent; CNS: Conscious and oriented; CVS: S1, S2 (positive), murmurs noted; RS: BLAE (+); GIT: soft, non-distended.

VITALS

Respiratory rate (RR): 16 breaths/minute; Pulse rate (PR): 89 beats/minute; Temperature: 98°F; Oxygen saturation (SpO_2): 98%.

FAMILY HISTORY AND HISTORY OF PRESENT ILLNESS

Elder brother was normal. Patient was diagnosed with Sickle cell disease (SCD) with Beta thalassemia at 6years of age. Patient was on Tab. Hydroxyurea and tablets got over, hence he stopped taking it 10 days ago. Patient was diagnosed with Human immunodeficiency virus (HIV) when he was 14years old. Patient was on Tab. Abacavir 60mg/ Tab. Lamivudine 30mg (4-0-4) and Tab. Efavirenze 200mg (2tab).

LABORATORY FINDINGS

A. DIAGNOSTIC TESTS

- RBC morphology – Mild microcytic hypochromic, mild anisopoikilocytosis.
- Coagulation profile – Normal.

Table 1: Laboratory findings.

Parameters	Obtained value	Normal range	Interference
Hemoglobin	7.60g/dl	12-17.5 g/dl	Anemia
RBCs	3.41 mill/cumm	4.2-6.1 mill/cumm	Vitamin B_6, B_{12} or folate deficiency
WBCs	16400 cells/cumm	5000-13000 cells/cumm	Infection/ Inflammation
Neutrophils	78%	40-80%	Normal
Lymphocytes	20%	20-40%	Normal
Monocytes	1%	2-12%	Monocytopenia
Eosinophils	1%	1-6%	Normal
Mean corpuscular hemoglobin	22.20pg	27-33pg	Iron deficiency anemia
Mean corpuscular hemoglobin concentration	32.90g/dl	31-37g/dl	Normal
Mean corpuscular volume	68fL	80-96fL	Microcytic Anemia
Packed cell volume	23.10%	40-56%	Nutritional deficiency (iron, folate and vitamin B_{12} deficiency)
Red cell distribution width	17.80%	11.6-14%	Iron/folate deficiency Anemia
Serum glutamic-oxaloacetic transaminase	56IU/L	5-40IU/L	Liver damage
Serum glutamic-pyruvic transaminase	21IU/L	7-56IU/L	Liver damage
Serum Alkaline phosphate	325IU/L	44-147IU/L	Liver damage
Total Bilirubin	0.80mg/dl	0.2-1.2mg/dl	Normal
Direct Bilirubin	0.30mg/dl	0.1-0.4mg/dl	Normal
Indirect Bilirubin	0.50mg/dl	0.2-0.8mg/dl	Normal
Total Protein	7.60g/dl	6-8g/dl	Normal
Serum Albumin	4.10g/dl	3.5-5.5g/dl	Normal
Serum Globulin	3.50g/dl	2.3-3.6g/dl	Normal
Albumin/ globulin ratio	1.17	1-2	Normal
Sodium	130mEq/l	135-145mEq/l	Normal
Potassium	4.40mEq/l	3.5-5mEq/l	Normal
Urea	29mg/dl	7-20mg/dl	Dehydration/ Kidney damage
Creatinine	0.78mg/dl	0.5-1.2mg/dl	Normal

Table 2: Drug Management.

Sr.	Drug	Dose	Route	Frequency
1.	Inj. Tramadol	2ml	IV	TID
2.	Inj. Pantoprazole/ Ondansetron	40mg/2ml	IV	BD/TID
3.	Inj. Multivitamin B complex	10ml	IV	BD
4.	Inj. Sodium chloride	10cc	IV	Per hour
5.	Tab. Abacavir/ Lamivudine	60mg/30mg	PO	4-0-4
6.	Tab. Efavirenze	200mg	PO	2 OD
7.	Tab. Sodium bicarbonate	500mg	PO	2-2-2-2
8.	Tab. Folic acid	5mg	PO	OD
9.	Syp. Laxose	2tsp	PO	TDS
10.	Cap. Hydroxyurea	250mg	PO	OD
11.	Tab. Deferasirox	200mg	PO	BD

- 2 pint PCV and 370ml Blood was transfused.

- CD4 lab report – 348cells/cells/mm^3 (low count) (500-1200cells/mm^3), 27%.

Other laboratory investigations are discussed in Table 1.

FINAL DIAGNOSIS

Correlating subjective and objective data along with past history, final diagnosis was made to be – Vaso-occlusive crisis in case of Sickle cell disease with Beta thalassemia and Human immunodeficiency virus.

Therapeutic management of the patient including drugs, dose, frequency and route has been briefly discussed in Table 2.

PLAN OF ANALYSIS

DISCUSSION

A 17-year-old patient presented to the hospital with the symptoms of vaso-occlusive crisis such as bilateral upper and lower limbs and back pain. The patient was previously diagnosed with SCD with Beta-Thalessemia and had 26-28 times blood transfusions where he might have infected with contaminated HIV blood. Hence, tested positive for HIV/AIDS without any family history. Patient had complaint of chest pain and constipation on 3rd day of admission. Soon after blood transfusion and proper management, pain reduced and compliant of constipation was resolved and patient became stable. Correlation between low levels of Hemoglobin and pain in SCD patients is well known. Managing low levels of hemoglobin in such patients becomes crucial. The use of Hydroxyurea has increased in sickle cell diseased patients focusing its action on preventing future blood transfusions and sickle cell crisis by changing the shape of sickled cells and increasing levels of fetal hemoglobin. While having conversation with the patient, he seemed depressive (sad and unhappy). Even after being joyful around the patient, he seemed uninterested. Thus, it is necessary to closely observe any signs and symptoms of psychiatric conditions in patients having such complicated conditions, since children are more likely to develop depression and anxiety. HIV weakens one's immune system, thereby decreasing the quality of life along with increasing risk of mortality among the patients. On the other hand Beta-Thalessemia is genetic incurable condition with no specific therapeutic agents to be treated with. Managing all the above condition in patient is challenging hence, closely monitoring for any fatal signs and symptoms becomes significant.

CONCLUSION

Sickle cell disease with Beta thalassemia and HIV/AIDS when interacts in a single patient, risk can be elevated for developing life-threatening complications such as avascular necrosis, stroke, sepsis and much more. Such underlying incurable diseases or disorders can affect one's mental health leading to psychiatric conditions such as depression. Moreover, there is limited data available on proper management of such comorbidity, that it even becomes difficult for the clinicians to minimize the dose and maximise effectiveness of the therapy. Hence, thorough research on risks and management of these diseases is much needed to ensure well-being of patients facing these complicated comorbid conditions.

ACKNOWLEDGEMENT

We would like to thank Principal Dr. Gunosindhu Chakraborthy, Principal and Professor PIPR, Parul University, all the Authors.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

ABBREVIATIONS

AIDS: Acquired immunodeficiency syndrome; **BID:** Twice a day; **BLAE:** Bilateral air entry clear; **CD4:** Cluster of differentiation 4; **CVS:** Cardiovascular system; **CNS:** Central nervous system; **GIT:** Gastrointestinal tract;

HbA: Hemoglobin A; **HIV:** Human immunodeficiency virus; **IV:** Intravenous; **OD:** Once a day; **PCV:** Packed cell volume; **PO:** Per oral; **PR:** Pulse rate; **RBCs:** Red blood cells; **RR:** Respiratory rate; **RS:** Respiratory system; **SCD:** Sickle cell disease; **SpO_2:** Oxygen saturation; **TID:** Thrice a day; **WBCs:** White blood cells.

REFERENCES

1. Steinberg MH. Management of sickle cell disease. N Engl J Med. 1999; 340(13):1021-30. doi: 10.1056/NEJM199904013401307, PMID 10099145.
2. Platt OS, Thorington BD, Brambilla DJ, Milner PF, Rosse WF, Vichinsky E, *et al*. Pain in sickle cell disease. Rates and risk factors. N Engl J Med. 1991;325(1):11-6. doi: 10.1056/NEJM199107043250103, PMID 1710777.
3. Shapiro BS. The management of pain in sickle cell disease. Pediatr Clin North Am. 1989;36(4):1029-45. doi: 10.1016/s0031-3955(16)36735-9, PMID 2666928.
4. Ballis SK, Carlos TM, Dampier C, Guidelines Committee. Guidelines for standard of care of acute painful episodes in patients with sickle cell disease. Harrisburg, Pa.: commonwealth of Pennsylvania Department of Health; 1996.
5. Serjeant GR, Ceulaer CD, Lethbridge R, Morris J, Singhal A, Thomas PW. The painful crisis of homozygous sickle cell disease: Clinical features. Br J Haematol. 1994;87(3):586-91. doi: 10.1111/j.1365-2141.1994.tb08317.x, PMID 7993801.
6. Gonzalez-Redondo JM, Stoming TA, Lanclos KD, Gu YC, Kutlar A, Kutlar F, *et al*. Clinical and genetic heterogeneity in black patients with homozygous beta-thalassemia from the southeastern United States. Blood. 1988;72(3):1007-14, PMID 2458145.
7. Gonzalez-Redondo JM, Kutlar A, Kutlar F, McKie VC, McKie KM, Baysal E, *et al*. Molecular characterization of Hb S(C) beta-thalassemia in American Blacks. Am J Hematol. 1991;38(1):9-14. doi: 10.1002/ajh.2830380103, PMID 1897518.
8. Christakis J, Vavatsi N, Hassapopoulou H, Angeloudi M, Papadopoulou M, Loukopoulos D, *et al*. A comparison of sickle cell syndromes in northern Greece. Br J Haematol. 1991;77(3):386-91. doi: 10.1111/j.1365-2141.1991.tb08589.x, PMID 2012764.
9. Al-Salem AH, Naserullah Z, Qaisaruddin S, Al-Abkari H, Al-Faraj A, Yassin YM. Splenic complications of the sickling syndromes and the role of splenectomy. J Pediatr Hematol Oncol. 1999;21(5):401-6. doi: 10.1097/00043426-199909000-00012, PMID 10524454.
10. Owusu ED, Visser BJ, Nagel IM, Mens PF, Grobusch MP. The interaction between sickle cell disease and HIV infection: A systematic review. Clin Infect Dis. 2015;60(4):612-26. doi: 10.1093/cid/ciu832, PMID 25344542.