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Case Report

Thalassemia β Major in Confirmed Covid-19 Patient: A Case Report

Yosua Butar Butar[1,*], Puspa Wardhani[2]

ABSTRACT

The incidence of Thalassemia with confirmed Covid 19 is very rare. The aim of this study is to know the prognosis and evaluate the management of therapy in thalassemia β Mayor patients with COVID-19. This case study describes the progression of Thalassemia β Mayor with COVID 19 starting from how to establish the diagnosis of Thalassemia β Mayor and COVID 19 until how to treat thalassemia β Mayor patients with COVID-19 This case study result describes that COVID 19 aggravates thalassemia β Mayor. Infection can adversely affect thalassemia. Giving Blood transfusions must be done due to anemia in Thalassemia, but it will have an impact on the accumulation of iron in the body which will increase the severity of the infection. The administration of iron-chelation drugs is beneficial for Covid but on the other hand, it is contraindicated in Thalassemia patients.

Key words: Thalassemia, Infection, Preventable Death, COVID-19.

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INTRODUCTION

Thalassemia is a congenital hemolytic disorder caused by mutation of the globin chains that make up hemoglobin resulting in the deficiency of synthesis of part or all of the globin chains. Hematological abnormalities in thalassemia range from asymptomatic to various degrees of anemia, ineffective erythropoiesis, and hemolysis.[1]

Infection is the second most common cause of death in thalassemia major, as well as iron-chelation drug administration is not allowed in thalassemia patients with infection because it will make it easier for pathogens to get iron so that the pathogen will be more virulent. In early 2020, the world was shocked by the incidence of severe infections with unknown causes, which originated from a report from China. This virus can be transmitted from person to person and has spread widely in China and more than 190 other countries.[2-10]

Based on these data, it is known that Thalassemia patients with SARS-CoV2 have a high risk in their prognosis. The aim of this case study appointed to study further a case of Thalassemia β Major accompanied by confirmed SARS-CoV2 infection to determine the diagnosis, effective treatment management that resulted in better outcomes.

CASE REPORT

A 48-Years Old Man was admitted with a chief complain of fever three days admission to hospital. Complaints fever accompanied with short of breath, cough, cold, nausea. From medical have been diagnosed thalassemia in the past 4 years ago. History of hypertension, diabetes Mellitus was denied and History of Thalassemia in family was denied.

On Examination, patient was awake with GCS E4V5M6. The vital sign showed Blood pressure of 120/80mmHg, Heart rate 100 times per minute, Respiration rate 28 times per minute and body temperature 37.0^{0}C. The other examination such as Head and Neck, Thorax, Abdomen, Extremities was normal.

DISCUSSION

Diagnosis of COVID-19 pneumonia is based on examination of complaints, physical examination, and support. In these patients who meet the criteria according to WHO, fever, symptoms of shortness of breath, cough. In these patients, these are mild symptoms. WHO recommends that all patients with suspicion of COVID-19 undergo Reverse Transcriptase – Polymerase chain Reaction (RT-PCR) examination? The results of the RT-PCR examination in this patient were detected.[11]

Thalassemia β is characterized by reduced or absent production of β globin chains in the hemoglobin molecule which causes excess α globin chains, resulting in reduced hemoglobin in red blood cells, decreased production of red blood cells, and anemia. Examination of HB electrophoresis in this patient shows a decrease in HbA which reflects a decrease in β globin chain synthesis, then as compensation, there is an increase in the α globin chain which can be seen from the increase in HbA2 and HbF. The clinical symptoms of β thalassemia major are serious, long-term, transfusion-dependent anemia with hemolysis, microcytosis and hypochromia, hyperferemia and complex clinical manifestations. In this patient we can see for Blood smear evaluation sign of massive ineffective erythropoiesis such as normocyte, teardrop cells, target cells, ovalocyte, fragmentocyte.[12]

Individuals with thalassemia major usually come to the clinician in the first 2-years and require regular blood transfusions to survive. Patients are severely anemic and require regular transfusions. Transfusions are given up to Hb 9-10 g / dL). In this patient, clinical symptoms such as pale, conjunctiva

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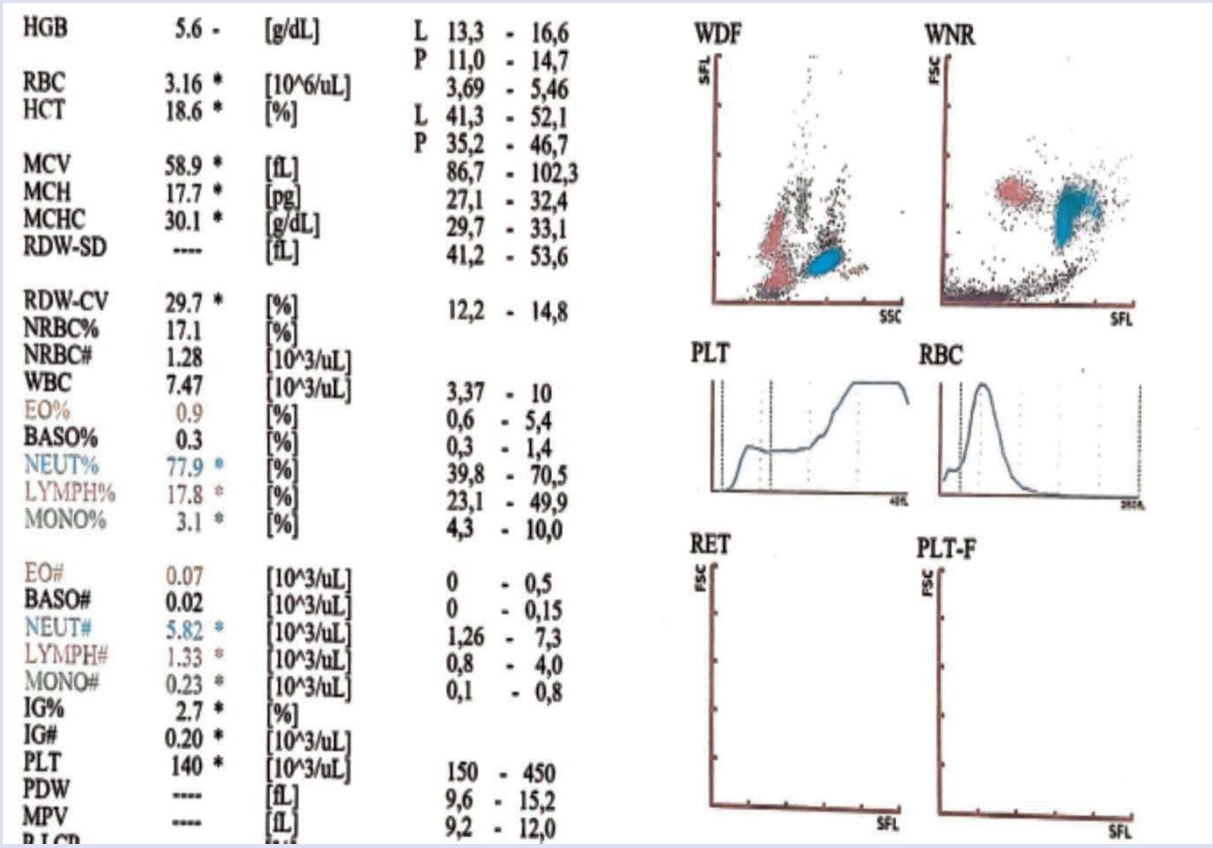

Figure 1: Scattergram.

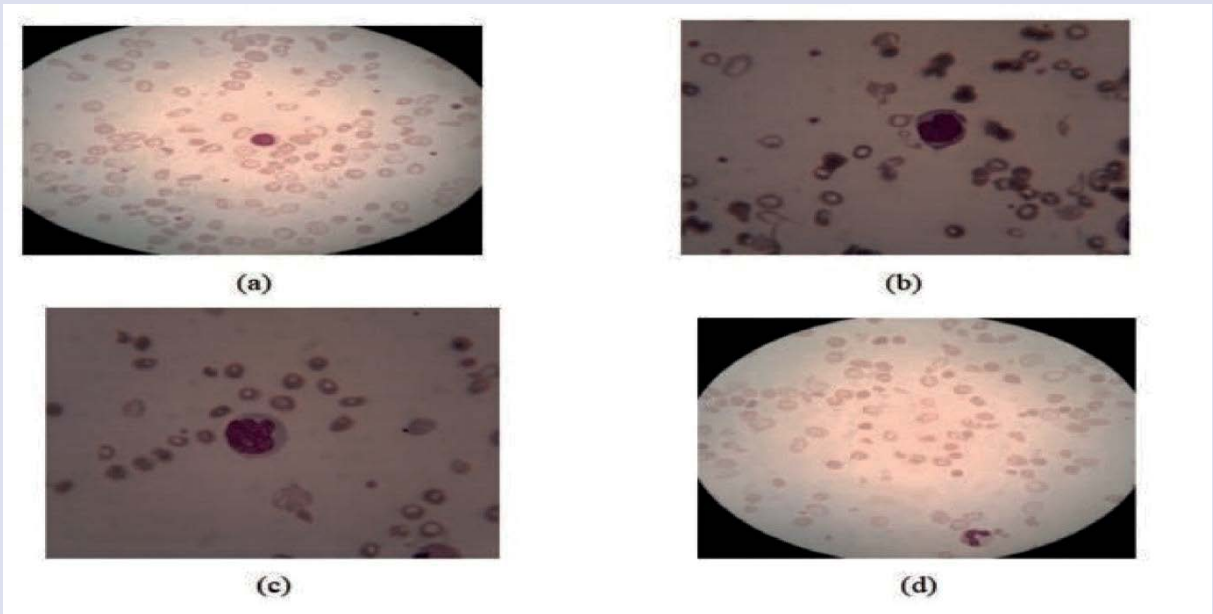

Figure 2: Blood smear evaluation a) Mature lymphocytes with morphology erythrocyte Hypochromic microcytic anisopoikilocytosis (normocytic, teardrop cells, ovalocytes, target cells, fragmentocytes, b) Immature Granulocytes (Metamielocytes), c) Atypical Lymphocytes, d) Segment neutrophils.

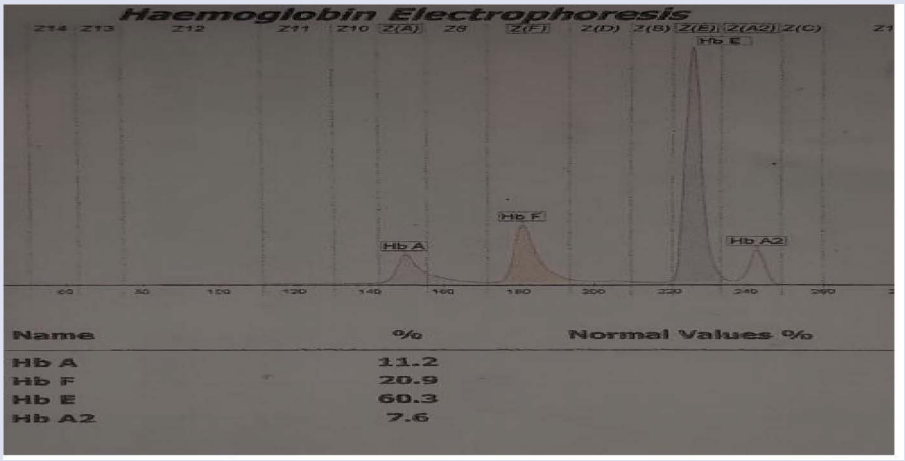

Figure 3: HB electrophoresis examination.

Table 1: Laboratory results.

	8/12/20	10/12/20	13/12/20	Reference range
Hematology				
Hb (g/dL)	5.6	5.4	9.6	13.3-16.6
RBC (10^6/µl)	3.16	3.19	4.74	3.69-5.46
Hct (%)	18.6	19.5	32.8	41.3-52.1
MCV (fl)	58.9	61.1	69.2	86.7-102.3
MCH (pg)	17.7	17.9	20.3	27.1-32.4
MCHC (g/L)	30.1	29.2	28.3	29.7-33.1
RDW CV (%)	29.7	29.1	33.9	12.2-14.8
WBC (10^3/µl)	7.86	7.93	7.82	3.37-10
% Eo	0.9	1.8	1.7	0.6-5.4
% Ba	0.3	0.5	0.4	0.3-1.4
% Neu	77.9	65.4	81.9	39.8-70.5
% Ly	17.8	28.9	12.5	23.1-49.9
% Mo	3.1	3.4	3.55	4.3-10
Plt (10^3/µl)	140	215	205	150-450
NLR	4.37	2.26	6.55	
Clinical Chemistry				
Na (mmol/L)		139	139	136-145
K (mmol/L)		4.8	5.2	3.5-5.1
Cl (mmol/L)		102	99	98-107
Coagulation test				
PPT (sec)		11	12.9	9-12
APTT (sec)		31.4	33	23-33
Fibrinogen	190			
D-Dimer	300	420	290	< 400
Immunology				
Procalcitonine (ng/ml)	0.14		0,19	< 0.5
Test Feritin (ng/ml)	8245			Men: 22-323 Women: 10-291
Blood Gas Analysis				
pH		7.37		7.35-7.45
pCO2 (mmHg)		45		35-45
pO2 (mmHg)		127		80-100
TCO2 (mmol/l)		24.6		23 - 30
Beecf		26		-3.5 - 2
SO2c (%)		-1.8		94- 98
A-aDO2		125		
%FiO2		99		
HCO3- (mmol/l)		21		22.0 - 26.0
Temp		37° C		
Serology				
Rapid Test Antibodi	Reaktif			
Swab PCR		Detected		

anemic, liver enlargement, and severe anemia with HB 5,6 so the patient require a blood transfusion.[13]

The problem in this patient is the controversy over the administration of iron chelation in this patient. On the one hand, iron chelation is believed to be useful for management in COVID-19 patients, with the mechanism: Inhibition of viral replication, Decrease of iron availability, Upregulation of B cells, Improvement of the neutralizing anti-viral antibody titer, Inhibition of endothelial inflammation, Prevention of pulmonary fibrosis and lung decline *via* reduction of pulmonary iron accumulation On the other hand, giving iron chelation to Thalassemia patients is not recommended for Thalassemia patients with the proven infection including SARS-COV 2 infection, because the iron chelation drug will cause leukopenia and will make it easier for pathogens to use iron as a food reserve.[13]

Treatment for thalassemia depends on the type and severity of the disorder. B minor thalassemia generally does not require therapy. Treatment for patients with β thalassemia major includes transfusion therapy, iron chelation, splenectomy, and allogeneic hematopoietic transplantation.[13]

CONCLUSION

This patient was diagnosed with Thalassemia β Major with SARS-CoV2 infection. The diagnosis of thalassemia was obtained from anamnesis history of thalassemia, clinical findings of conjunctiva anemic and hepatomegaly, and supported by hematologic laboratory and Hb electrophoresis examination. SARS-COV2 infection was diagnosed because the RT-PCR of the patient was detected. Management of this patient was given Packet Red Cell (PRC) transfusion to manage anemia, prophylactic antibiotics to prevent antibiotic, and did not provide iron-chelation drugs. The whole of this case study is in accordance with the existing theory so that it can be used in other case studies, because the diagnose of this case is rare, its suggested to do more case studies like this case so we can get more clinical information.

ETHICAL APPROVAL

This study approved by the Ethics Committee from General Hospital RSUD DR. Soetomo, Surabaya, Indonesia with reff number 1204/126/4/I2022.

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DISCLOSURE STATEMENT

The authors have declared that no competing interests exist.

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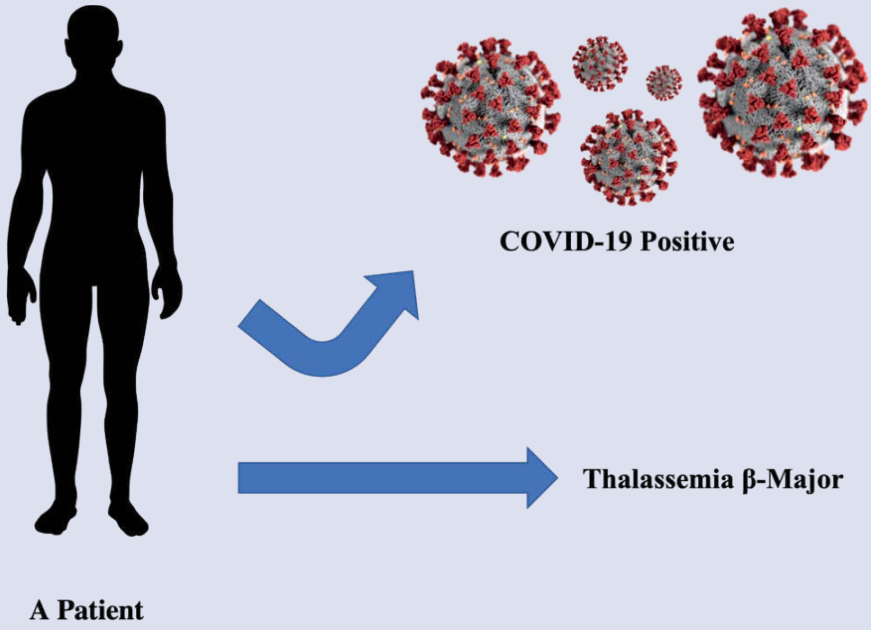
GRAPHICAL ABSTRACT



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