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RESEARCH ARTICLE

LIVING LONG WITH SICKLE CELL BETA THALSSEMIA: A-71-YEAR-OLD SURVIVOR: A CASE REPORT

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ABSTRACT

Sickle cell disease (SCD) is an inherited hemoglobinopathy associated with reduced life expectancy due to recurrent vaso-occlusive crises, progressive organ damage, and life-threatening infections. Although genotype is a major determinant of disease severity and mortality, advances in preventive strategies and comprehensive care have significantly improved survival. However, survival into the seventh decade remains uncommon in patients with sickle-β-thalassemia. We report the case of a 71-year-old Bahraini male with sickle-β^{+}-thalassemia who demonstrated a remarkably mild clinical course over seven decades. This case illustrates that survival in SCD is influenced not only by genotype but also by preventive strategies, adherence to vaccination and penicillin prophylaxis, regular follow-up at specialized SCD centers, and multidisciplinary care. Comprehensive management can substantially modify disease progression and improve long-term outcomes, even in patients with genetically severe forms of SCD.

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INTRODUCTION

Sickle cell disease (SCD) is an inherited blood disorder characterized by the presence of abnormal hemoglobin, leading to distortion of red blood cells into a sickled shape and subsequent membrane damage. This results in significant acute and chronic complications and reduced life expectancy. [1] SCD encompasses a group of inherited hemoglobinopathies caused by hemoglobin S (HbS). In Arab Gulf countries, including Bahrain, homozygous sickle cell anemia (HbSS) is the predominant genotype, followed by HbS/β-thalassemia, while HbSC is less common. [2] The presence of β-thalassemia modifies the clinical phenotype of SCD. HbS/β^{0}-thalassemia is clinically similar to HbSS, whereas HbS/β^{+}-thalassemia is generally associated with a milder disease course depending on residual β-globin production.[3 4] Patients with SCD experience both acute and chronic complications. Acute complications include vaso-occlusive crises, acute chest syndrome, stroke, priapism, and severe infections. Chronic complications include pulmonary hypertension, chronic kidney disease, avascular necrosis, retinopathy, and cardiomyopathy, all of which contribute to reduced life expectancy. [5] Although patients with HbS/β^{+}-thalassemia typically have a milder clinical course, survival into the seventh decade remains uncommon. In this report, we describe a 71-year-old patient with sickle-β^{+}-thalassemia who underwent splenectomy and remained free from severe infections while on prophylactic penicillin. This case highlights the influence of genotype and long-term management on survival in SCD.

CASEREPORT

A 71-year-old Bahraini male with sickle-β^{+}-thalassemia presented with a remarkably mild disease course over seven decades. His medical history included type 2 diabetes mellitus, hypertension, and hyperlipidemia. He underwent splenectomy 20 years earlier, vitrectomy 13 years earlier, and laparoscopic cholecystectomy 2 years prior, which required blood transfusion. He reported infrequent vaso-occlusive crises, minimal hospitalizations, and no history of acute chest syndrome, avascular necrosis, or stroke. He remained physically active and independent in activities of daily living. The patient attended regular follow-up visits at both the non-communicable disease clinic and a specialized sickle cell clinic. His medications included metformin 500 mg daily, atorvastatin 20 mg daily, a combination of perindopril 5 mg and indapamide 1.25 mg daily, aspirin 81 mg daily, and penicillin V 250 mg daily as prophylaxis. He was fully vaccinated against coronavirus disease 2019 (COVID-19) and had received recommended post-splenectomy vaccines, including pneumococcal and meningococcal vaccines. Hemoglobin electrophoresis revealed HbS 53.2%, HbA 24.2%, HbF 14.3%, and HbA_2 5.2% (Table 1). His baseline

hemoglobin ranged from 11 to 12 g/dL. Reticulocyte count was 4.2%, white blood cell count was 8.12 × 10^9/L, and ferritin level was 82.3 µg/L (Table 2). Markers of hemolysis showed elevated total bilirubin of 42 µmol/L, with direct bilirubin 18 µmol/L and indirect bilirubin 24 µmol/L. Lactate dehydrogenase (LDH) was 357 U/L (Table 3). Peripheral blood smear demonstrated anisocytosis and poikilocytosis (++), hypochromia (+), polychromasia (+), macrocytosis (+), target cells (++), ovalocytes (+), and occasional sickle cells (Table 4).

Renal function was preserved, with serum creatinine of 64 µmol/L and uric acid of 483 µmol/L. Liver function tests were within normal limits, including alkaline phosphatase (ALP) 95 U/L, alanine aminotransferase (ALT) 18 U/L, and gamma-glutamyl transferase (GGT) 12 U/L. Glycated hemoglobin (HbA1c) was 48 mmol/mol (approximately 6.5%), indicating good glycemic control. Echocardiography demonstrated preserved left ventricular function with an ejection fraction of 60% and grade I diastolic dysfunction. The aortic valve showed nodular calcification without stenosis. Right ventricular size and function were normal, with an estimated right ventricular systolic pressure of 28 mmHg and no pericardial effusion.

Table 1. Hemoglobin Electrophoresis Results with Reference Ranges

Parameter	Patient Value	Normal Range
HbA	24.2%	95-98%
HbS	53.2%	0%
HbF	14.3%	<2%
HbA_2	5.2%	2-3.5%

Table 2. Complete blood Count and Iron studies

Test	Patient Value	Normal Range
Hemoglobin	11-12 g/dL	13.5-17.5 g/dL
Reticulocytes	4.2%	0.5-2.5%
White Blood Cells Count	8.12 x 10^9/L	4.0-11.0 x 10^9/L
Ferritin	82.3 µg/L	30-400 µg/L

Table 3. Hemolysis Markers

Test	Patient Value	Normal Range
Total Bilirubin	42µmol/L	5-21 µmol/L
Direct Bilirubin	18 µmol/L	0-7 µmol/L
Indirect Bilirubin	24 µmol/L	3-17 µmol/L
Lactate dehydrogenase (LDH)	357 U/L	140-280 U/L

Table 4. Peripheral Blood Smear findings

Finding	Patient Finding	Normal
anisocytosis and poikilocytosis	++	Absent
Hypochromiaand Polychromasia	+	Absent
Macrocytosis	+	Absent
Target Cells	++	Absent
Ovalocytes	+	Absent
Sickle cells	Occasional	Absent

DISCUSSION

Patients with SCD typically have reduced life expectancy due to recurrent vaso-occlusive crises, progressive organ damage, and increased susceptibility to severe infections. Earlier studies reported median survival in the fourth to fifth decades, particularly in patients with HbSS genotype. Recent evidence, however, indicates improved survival with advances in medical care.[6] The prolonged survival observed in this patient may be partly explained by genotype-specific factors. HbS/β^+-thalassemia is associated with a milder phenotype due to residual β-globin production and the presence of hemoglobin A, which reduces hemoglobin S polymerization and vaso-occlusion.[7] Splenectomy increases susceptibility to infections, particularly from encapsulated organisms. However, vaccination programs, including pneumococcal, Haemophilus influenzae type b, and meningococcal vaccines, significantly reduce infection-related morbidity and mortality.[8]

Prophylactic penicillin also plays a critical role in reducing severe pneumococcal infections.[6] Regular follow-up in specialized sickle cell centers further contributes to improved outcomes and increased life expectancy.[9] In addition, lifestyle and socioeconomic factors, including access to healthcare, treatment adherence, patient education, and social support, are associated with better clinical outcomes.[7] This case highlights that survival in SCD is multifactorial and influenced not only by genotype but also by preventive care, adherence to vaccination and prophylactic antibiotics, regular monitoring, and multidisciplinary management. Despite historically reduced life expectancy in severe genotypes, comprehensive modern care can significantly alter disease progression and improve prognosis.

PATIENT'S CONSENT: Verbal informed consent for publication was obtained from the patient. Identifying information has been removed to ensure confidentiality.

COMPETING INTEREST: The authors declare no competing interests.

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