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Case Report

A challenging case of severe sickle cell crisis: a case report on vaso occlusive crisis in sickle cell disease with multi organ failure

Prasanta Dihingia, Rima Moni Doley, Arniban Borah, Anwesha Phukan, Karthik Ilangovan*, Akshay Saha

Department of Medicine, Assam Medical College Dibrugarh, Assam, India

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***Correspondence:**
Dr. Karthik Ilangovan,
E-mail: karthikrani1234@gmail.com

ABSTRACT

Sickle cell disease involves recurrent hemolysis and vaso occlusive crisis, which occurs mainly due to abnormal Hemoglobin S polymerization and sickling of erythrocytes. These sickled erythrocytes are prone for damage in micro capillaries, escalating into vaso occlusive crisis presenting as acute pain episodes and recurrent hospitalisations. Severe vaso occlusive crisis can precipitate multi organ dysfunction and failure, significantly increasing morbidity and mortality. This report presents a 19-year-old male with known sickle cell disease who developed a severe vaso occlusive crisis complicated by multi organ failure, including acute chest syndrome, renal impairment, and liver dysfunction. The patient presented with generalized joint pain, chest discomfort, dyspnea, and oliguria. Clinical evaluation revealed pallor, mild icterus, hypoxemia, and hemodynamic instability. Investigations demonstrated bilateral pulmonary infiltrates, pleural effusions, and deranged liver and renal function tests. Management included aggressive intravenous hydration, oxygen therapy, blood transfusion, hydroxyurea, broad spectrum antibiotics, anticoagulation, and supportive care. Despite clinical deterioration necessitating intensive care and non-invasive ventilation, the patient responded favourably to multidisciplinary intervention, with gradual resolution of organ dysfunction and return to baseline status. This case underscores the critical importance of early recognition, prompt resuscitation, and comprehensive management in sickle cell disease patients with vaso occlusive crisis and evolving multi organ failure. Timely intervention can be lifesaving and is essential for improving outcomes in high-risk sickle cell disease populations.

Keywords: Sickle cell disease, Vaso occlusive crisis, Multi organ failure, Acute chest syndrome, Organ dysfunction, Hemolytic anemia, Thrombotic events, Intensive care, Acute kidney injury, End organ damage

INTRODUCTION

Sickle cell disease occurs due to substitution of single nucleotide in beta globin gene, glutamate to valine which leads to abnormal Hemoglobin S under deoxygenated conditions, polymerization of HbS occurs leading to the formation of sickle shaped, rigid erythrocytes. These sickle shaped erythrocytes are vulnerable to damage, rigid and has decreased life span leading to long term hemolysis and intermittent vaso occlusive episodes.[1,2] Vaso occlusive crises are the most common and catastrophic manifestation of sickle cell disease, characterized by tissue ischemia, Joint pain and severe inflammatory responses.[3] The clinical spectrum of vaso occlusive crises ranges from localized pain to severe complications involving vital organ systems. In some instances, vaso occlusive crises can escalate into multi-organ dysfunction or failure leading to catastrophic effects.[4] This includes mainly liver dysfunction, acute chest syndrome, acute kidney injury and encephalopathy. Factors such as infection, dehydration, anemia, hypoxia, and acidosis exacerbate the crisis, promoting widespread vascular occlusion and ischemia.[5] Despite constant improvement in the sickle cell disease management through hydroxyurea, chronic transfusions, and supportive care, multi organ failure

remains a critical and underreported complication. Early identification and aggressive multidisciplinary management are crucial to improving outcomes in these high-risk cases.[6] This case report presents a young male with sickle cell disease who developed a severe vaso occlusive crisis and multi organ failure, emphasizing the importance of timely intervention in the critical care setting.

CASE REPORT

A 19-year-old male who is a diagnosed case of sickle cell anemia for past 8 years, presented to emergency room with complaints of bilateral knee, ankle, elbow, shoulder joint pain for 3 days duration. Patient also complains of fever with chills, dysuria, chest pain, difficulty breathing for 3 days duration, nausea for 2 days duration, decreased urine output for 1 day duration. No history of conjunctival congestion/alcohol intake / herbal medication intake/ hepatotoxic drug intake/cough/intravenous drug abuse/tattoo/high risk behaviour. Patient is receiving PRBC transfusion regularly for every three months for past eight years. He was not a known case of diabetes, hypertension or thyroid disorder.

On general examination, he was conscious, oriented, afebrile and dehydrated. Pallor and mild icterus present at presentation. His vital signs revealed a pulse rate of 116 bpm and blood pressure of 112/ 74 mmHg with Mean arterial pressure of 80 mmHg. Oxygen saturation in room air was 90%. And respiratory rate 22 per minute. No features of petechiae, purpura, ecchymoses or gum bleeding were found.

Investigations

The patient's blood and urine samples were sent for routine investigations and culture sensitivity. Chest X-ray showed bilateral infiltrates with bilateral pleural effusion. USG whole abdomen-bulky left kidney taking increased vascularity on colour doppler suggest possibility of pyelonephritis, non-visualised spleen, bilateral raised renal echotexture. Echocardiography–normal study LVEF 60%. HRCT thorax–cardiomegaly with reversed aorto pulmonary ratio and bilateral pleural effusion. ECG-normal sinus rhythm.

The laboratory investigations revealed several abnormalities consistent with multi organ involvement in severe sickle cell crisis. Hemoglobin levels were markedly reduced indicating severe anemia. On day 1 Hb was 6.3 gm/dl, after receiving 4 units PRBC, on day 10, Hb was 8.6 gm/dl, while the total leukocyte count was elevated with 23200 cells/mm^3 on day 1, reflecting an ongoing inflammatory or infectious process. USG whole abdomen revealed left sided pyelonephritis with bilateral raised renal echotexture. After receiving antibiotics, on day 10, total leukocyte count came down to 9400/mm^3. Platelet counts went down upto 40,000/mm^3 on day 5, and normalised to 1.2 lakhs/mm^3 on day 10. Renal function tests showed elevated blood urea and serum creatinine, confirming acute kidney injury. Liver function tests demonstrated increased serum bilirubin, predominantly direct hyperbilirubinemia, along with raised transaminases (AST and ALT), suggesting hepatic dysfunction. On day 1 total bilirubin, direct bilirubin was 32 and 29 mg/dl respectively. On day 10, it came down to 6.7 and 5.5 mg/dL respectively. Serum lactate dehydrogenase was also elevated (1120 U/l), indicative of ongoing hemolysis. Electrolyte analysis revealed mild hyponatremia 120 mg/dL and hyperkalemia 5.95 mg/dl on day 1, consistent with renal impairment, at day 10, electrolytes came to normal.

The patient's D-dimer levels were significantly raised (8548 ng/ml), pointing to a hypercoagulable state and increased risk of thrombotic complications. Anticoagulants were added injection LMWH for 5 days followed by oral anticoagulants in view of hyper coagulable state. Urinalysis showed proteinuria and microscopic hematuria, further supporting renal involvement. Microbiological investigations, including blood culture revealed MRSA sensitive to Doxycycline and urine culture and sensitivity revealed *Klebsiella pnemoniae* sensitive to Doxycycline, Gentamicin and Linezolid. These findings collectively confirmed the diagnosis of multi-organ failure, involving hematologic, renal, hepatic, and coagulation systems. The laboratory profile guided the multidisciplinary management approach, emphasizing the need for aggressive supportive care, transfusion, infection surveillance, and organ specific interventions to reverse the evolving dysfunction and improve patient outcomes.

Treatment and follow up

The patient was admitted in medicine unit 4 male ward of Assam medical college hospital on 16/06/2025 with sickle cell anemia in vaso occlusive crisis and was resuscitated with intravenous fluid 0.9% NaCl 1 litre over 2 hours followed by 0.9% NaCl 1.5 l per day, oxygen inhalation with face mask 10 l O_2, 1 unit PRBC transfusion on day 1 of admission, tablet hydroxyurea 500 mg one tab twice daily, Tab folvite 5 mg one tab once daily, tab deferasirox 400 mg one tab twice daily, injection piperacillin tazobactam 2.25 grams intravenously thrice daily, injection doxycycline 100 mg intravenously twice daily, injection furosemide 20 mg intravenously twice daily, injection LMWH 40 mcg subcutaneously once daily, injection tramadol infusion. Patient condition deteriorated on day 3 of admission, patient was shifted to intensive care unit, and was put on non-invasive ventilation and was managed with above mentioned treatment. Patient vitals improved, and was shifted back to male ward. On day 6, inj LMWH was stopped and changed to Tab dabigatran 150 mg twice daily. His LFT RFT parameters came back to normal, urine output improved, jaundice resolved, liver enzymes came back to normal on day 10 of admission. However, D-Dimer values remained high, with continuation of oral anti coagulants.

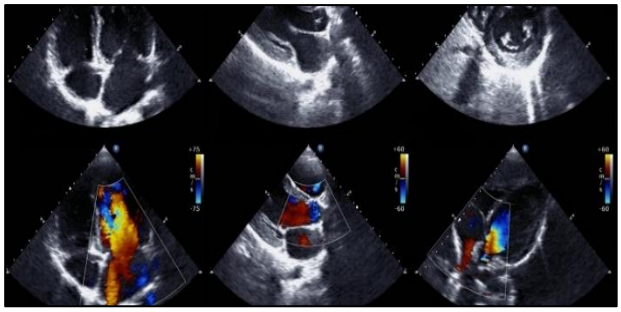

Figure 1: 2D Echocardiography showing normal study LVEF 60%.

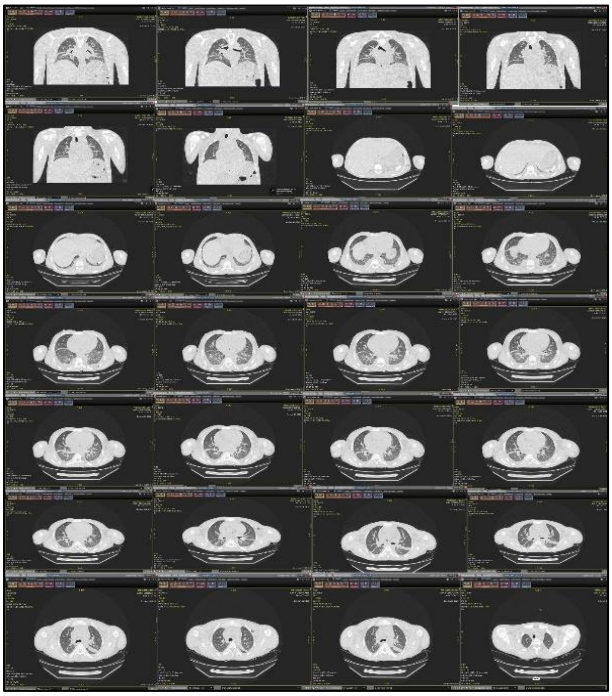

Figure 2: HRCT Thorax showing cardiomegaly with reversed aorto pulmonary ratio and bilateral pleural effusion.

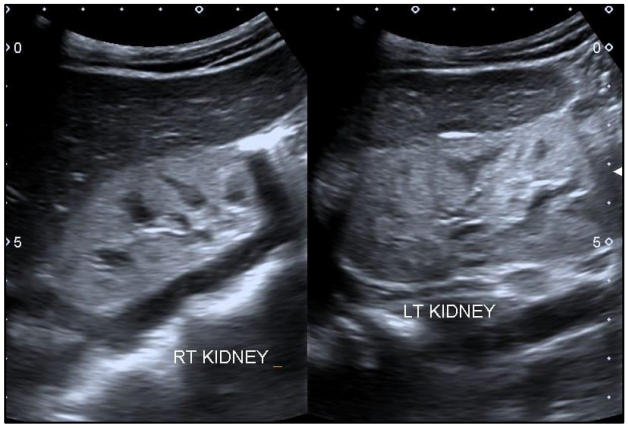

Figure 3: USG Whole abdomen showing bulky left kidney taking increased vascularity on colour doppler suggest possibility of pyelonephritis, non-visualised spleen, bilateral raised renal echotexture.

DISCUSSION

This case exemplifies the life-threatening potential of vaso occlusive crisis in sickle cell disease, demonstrating rapid progression to multi-organ failure. Previous case reports suggest that patients with sickle cell disease are at increased risk of having vaso occlusive crisis, The patients present with chest pain, breathing difficulty, hypoxemia. These patients are at increased risk of acute coronary syndrome.[7] Patients with Sickle cell disease can also present with leukoencephalopathy primarily occurring due to sickling of red blood cells inside cerebral microvasculature.[8]

In this case patient's acute chest syndrome (bilateral infiltrates, hypoxia) and renal impairment (oliguria, elevated creatinine) highlight systemic ischemia from microvascular occlusion. Hepatic involvement (elevated transaminases) and hematologic stress (severe anemia) further underscore the pathophysiological cascade: sickled erythrocytes obstructing blood flow, triggering inflammation, endothelial damage, and tissue infarction. Aggressive hydration, oxygen, and transfusion mitigated crisis progression, while antibiotics addressed potential infectious triggers. The ICU escalation emphasizes vaso occlusive crisis volatility and the need for early detection and management to reduce morbidity and mortality.

CONCLUSION

Vaso occlusive crisis associated with sickle cell disease demands immediate, comprehensive intervention to prevent irreversible organ damage. This case reinforces that rapid hydration, oxygenation, transfusion, and infection control are critical in multi organ failure scenarios. Prophylactic hydroxyurea and regular blood transfusions remain essential for long-term crisis prevention. Early recognition of evolving complications - particularly acute chest syndrome, hepatic injury and renal injury - is paramount to reduce mortality in high-risk sickle cell disease patients.

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