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Case Report

The Emotional Impact of Sickle Cell Disease Symptoms: A Case Review

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Abstract

Background: Sickle Cell Disease (SCD) is a chronic and debilitating disease that can affect almost any organ system in the body; hence, it is associated with a broad range of complications and comorbidities that may have a profound impact on a patients' quality of life through medical and mental health consequences. Psychosocial and affective comorbidities are prevalent in SCD and could considerably influence disease outcomes; however, treatment options for SCD psychosocial sequelae are suboptimal, and evidence on their effectiveness is limited.

Case Report: We report a 21-year-old African American female with a history of SCD and recurrent hospital admissions due to acute SCD complications, who was referred to the consultation-liaison psychiatry service for evaluation of psychiatric comorbidities and management recommendations.

Conclusion: This case demonstrates the complexity of the relationship between somatic complications and neuropsychiatric disorders in SCD and highlights the importance of early identification and management of mental health conditions in this patient population. We emphasize treatment challenges and the need for large scale research on pharmacotherapies and nonpharmacological interventions in treating psychosocial comorbidities associated with SCD.

Keywords: Sickle cell; Mental disorders; Adjustment disorders; Chronic pain

Background

Sickle Cell Disease (SCD) is an early age genetic condition that affects approximately 100,000 Americans [1]. It is a chronic and debilitating disease that can affect almost any organ system in the body; hence, it is associated with a broad range of complications and comorbidities that can impair patients' medical and mental health over time and may have a profound impact on their quality of life [2]. Vaso-occlusive crises (VOC) are hallmark symptoms in SCD, and acute pain episodes are the most common types of VOC. These episodes of severe and unpredictable pain are responsible for a large number of hospital encounters, and repeated episodes can lead to frequent hospitalizations [3]. Psychosocial and affective comorbidities are prevalent in SCD and can considerably influence disease outcomes, particularly outcomes associated with pain, including pain intensity and frequency, emergency department visits, and hospital admissions [4]. Anxiety, mood symptoms, negative thoughts, depression, neurocognitive deficits, and substance use disorders have been repeatedly reported to have an association with SCD [4,5]. While management of psychiatric symptoms is an important component of SCD treatment, many patients with psychiatric comorbidity remain unrecognized and untreated [2]. Moreover, treatment options for SCD psychosocial sequelae are suboptimal, and evidence on their effectiveness is limited [5].

Case Presentation

A 21-year-old African American female with a history of Sickle Cell Disease (SCD) and recurrent hospital admissions due to Vaso-Occlusive Crises (VOC) was referred to the consultation-liaison psychiatry service of Kern Medical Center due to the concern regarding the disproportionate number of admissions and ongoing mood symptoms and social isolation.

The patient is a known SCD patient with hemoglobin SS genotype diagnosed at two weeks of age. She has been treated with hydroxyurea and had a history of multiple hospital admissions due to vaso-occlusive pain episodes, Acute Chest Syndrome (ACS) requiring exchange transfusions, Idiopathic Intracranial Hypertension (IIH), and infectious episodes. She was unemployed and received significant support from her mother. More recently, there was a notable increase in the severity of pain episodes and frequency of hospital admissions, with more than ten hospitalizations over the past three months. Pain management during recent admissions required parenteral hydromorphone at escalating doses. The patient also presented with symptoms suggestive of depression. This led to the concern of potential suboptimal management of psychiatric sequela that might have also contributed to the pain experiences and frequent hospitalizations. Consultation-liaison psychiatry team was consulted for evaluation of psychiatric comorbidities and management recommendations.

On psychiatric assessment, the patient was uncooperative, withdrawn, and emotionally labile. She demonstrated maladaptive emotional and behavioral symptoms, including low mood, irritability, anger, and pain catastrophizing. She was overly concerned about her pain and medical condition and expressed being worried that she

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cannot attend school consistently and plan her future. She reported an escalation of her emotional symptoms over the past few weeks. She denied feeling depressed, stating that "it's not depression, it's more that I just don't want to do anything." She also denied changes in sleep or appetite; however, she endorsed decreased energy and general weakness secondary to her increased pain. She did not report loss of interest, describing, "I make plans to do things, but I can never get to do them." She denied suicidal thoughts but reported occasional passive thoughts of "not wanting to be alive" secondary to her extreme pain and the reduced quality of life caused by her disease. Personal and family history of mental illness was negative, and the patient did not have a history of drug abuse. The mental status examination was unremarkable. The Becks Depression Inventory and the Hamilton Rating Scale of Depression were administered as screening instruments, and the results were not conclusive. The diagnosis of adjustment disorder unspecified was made according to the fifth edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5). Duloxetine was initiated with 30 mg per day and increased to 60 mg per day over one week. Cognitive-Behavioral Therapy (CBT) with psychoeducation techniques were recommended and discussed with the patient. The patient was scheduled for frequent outpatient mental health visits to build trust and establish plans of care. However, she refused therapy and outpatient care, therefore the efficacy of the recommended treatment remained unclear.

Discussion

Sickle cell disease remains a constant treatment challenge for patients and health care professionals, despite the advances in medicine that have significantly improved the survival of patients with SCD. Multiple sources of biopsychosocial pathology leave patients with SCD prone to poor psychosocial adjustment and at increased risk for psychiatric illnesses [6]. The pain associated with vaso-occlusive episodes is unpredictable, severe, and debilitating. These episodes are more frequent in patients with higher disease severity and have a significant psychological impact on patients [2]. Physical disabilities, frequent hospitalizations, a constant fear of complications, educational difficulties, financial problems, lack of environmental resources, [7] racial prejudice, and stigmatization are other potential stressors that can contribute to the mental health burden on these individuals [8].

Pathophysiology of neuropsychological deficits associated with SCD is often attributed to the severe anemia, silent cerebral infarcts, and chronic inflammation, although the precise mechanisms are unclear [9]. Impairments in cognitive functioning are highly prevalent in patients with SCD [9]. Martin et al. examined certain cognitive domains in thirty symptomatic adults with SCD and used their unaffected siblings with sickle cell trait as a comparison group. They found that patients with symptomatic SCD were significantly slower in processing speed and demonstrated more executive functioning difficulties than their unaffected sibling counterparts [10]. Psychological symptoms, including social withdrawal, depression, anxiety, aggression, and feelings of helplessness, are also highly prevalent among children and adults with SCD [2]. The prevalence of clinical depression is estimated to be at least 20 percent [11]. Depression may also be associated with a higher rate of healthcare utilization and disease severity [12]. Taken together, debilitating somatic complications coupled with cognitive impairments and psychosocial disturbances can substantially impair Health-Related Quality of Life (HRQoL) in this patient population. A recent review demonstrated that patients with SCD have lower HRQoL scores than the general population in physical, psychological, and social domains [13]. While it has long been known that somatic complaints associated with chronic diseases can lead to neuropsychiatric disorders, a growing body of evidence suggests that this link in patients with SCD is bidirectional [7]. Psychiatric symptoms resulting from somatic complaints in SCD may exacerbate patients' physical symptoms through a psychosomatic component, and further decrease their HRQoL. Namely, somatic complications may lead to depression and anxiety, which can further worsen the pain and increase the risk of somatic complications [7]. This pathologic circle has been the subject of research in the past two decades, and findings suggest that the management of psychosocial and affective comorbidities are essential, when treating sickle cell disease [4].

Adjustment disorder is often prevalent in consultation-liaison psychiatry services [14]. A 2011 meta-analysis found a prevalence rate of 15 to 19 percent in oncology-related settings [15]. According to DSM-5, [16] functional impairment and/or disproportional emotional distress that occur within three months of, and in response to an identifiable stressor are key components of adjustment disorder. Though our patient's reaction might be proportional to her recently worsening pain and repeated hospitalizations, significant impairment of functioning in different life domains happening shortly after the worsening of her medical condition warrants the diagnosis of adjustment disorder. Katz et al. demonstrated that Adjustment disorder is the most appropriate diagnosis for patients with chronic pain, who are excessively concerned about their pain [17]. The patient also showed various maladaptive behaviors, of which, catastrophizing was conspicuous. Catastrophizing is defined by a negative cognitive-affective response to stressful events, and represents with rumination, magnification, and helplessness [18]. Pain catastrophizing is common in individuals with SCD, and negatively affects their response to pain and their HRQoL [4]. Numerous studies suggest that rumination is a strong risk factor for the development of depression and anxiety [19]. The patient's catastrophizing manifested as magnification of pain-related symptoms, feeling of helplessness, and rumination about her pain by repetitive passive focusing on the causes and consequences of her pain without active engagement in problem-solving and coping strategies. She refused therapy of any form in spite of multiple efforts to convince her otherwise, underlining the need for further multidisciplinary treatment approaches. Occasional thoughts of not wanting to live in this patient, though in the context of being overwhelmed secondary to pain, further highlights the need for mental health care interventions.

Despite strong evidence in the literature on co-treating somatic and neuropsychiatric sequelae in SCD, evidence on the effectiveness of interventions and target treatments for psychosocial sequelae are sparse and mixed. In addition, patient's noncompliance to treatment and lack of resources can pose further challenges to treating physicians. Reports on pharmacotherapies are limited, and CBT is the most reported nonpharmacological treatment with promising results [5]. Early screening and intervention is important and can improve disease outcomes [20]. It is essential for healthcare providers to become more familiar with psychosocial

comorbidities associated with SCD, which can help with early identification of patients who may benefit from nonpharmacological interventions or pharmacotherapies [4,11]. More research is needed on pharmacotherapies and nonpharmacological interventions in treating psychosocial comorbidities associated with SCD.

Conclusion

Despite the high prevalence of neuropsychiatric sequelae in SCD, treatment options are suboptimal and evidence on the effectiveness of available interventions and treatments are limited. We emphasize the importance of early identification and management of mental health conditions in patients with SCD and demonstrate the treatment challenges and limitations for patients and providers. We also highlight the need for more research on pharmacotherapies and nonpharmacological interventions in treating psychosocial comorbidities associated with SCD.

Disclosure

The authors report no proprietary or commercial interest in any product mentioned or concept discussed in this article.

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