



Review Paper

**CASE REVIEW: DRUG ADDICTION IN SICKLE CELL DISEASE, A
POSSIBLE ONGOING CHALLENGE IN THE MANAGEMENT OF PAIN ?**

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INTRODUCTION

Sickle cell anemia was first described as a disease entity that deserved much more attention than it was receiving at the time along the west African subregion in a publication in the British medical journal by J O Mabayoje as far back as 1956¹. In this presentation he gave various ways sufferers of the disease could present clinically. The disease actually first earlier described in the United States of America (USA) was of J. B Herrick in Chicago in 1910. It is a disease inherited in a mendelian fashion. What has perplexed scientists and medical personnel for years is the protean manifestations of the disease suggesting that there is a host of factors that contribute to the characteristics and presentation of the disorder.

A major feature is the persistent anaemia often worsened by recurrent crisis which could be brought on by a variety of factors. These include infection, extremes of temperature, stress, and low circulating oxygen in ambient atmosphere e.g. high altitude. In a significant number of cases no precipitating factor or factors can be identified². The victims of the disease from early on in life have retarded growth, recurrent fever, bone pain (vasoocclusive crisis) with joint pain/swelling and effusion. Jaundice and haemolysis are common features. It appears that no system is safe from affliction. These include kidney disease, pulmonary hypertension, leg ulcers, acute chest syndrome, retinopathy and priapism³.

The *sine qua non* of sickle cell anaemia is the glutamine to valine substitution in the sixth amino acid of the β -globin gene. This results in pathophysiologic processes that result in the clinical phenotype that extends beyond the red cell. These include aggregates of haemoglobin that encourage polymer growth leading to vascular obstruction^{4,5}. The loss of intracellular potassium and water resulting in cellular dehydration has also been implicated^{6,7}. Nitric oxide⁸, abnormal cell adhesiveness⁹, inflammation with abnormal activation of neutrophils and monocytes, increased proinflammatory mediators like tumour necrosis factor (TNF)- α , interleukin (IL)6, and IL- β ^{10,11,12,13}, and activation of the coagulation system have all been suggested to play vital roles in the pathophysiology of the disease. It is interesting to note that despite the vast and intimate understanding of the pathophysiology of this condition, many patients continue to undergo untold hardship and suffering at the mercy of this disease. More so in resource limited countries (RLC) as is ours.

The hall mark of sickle cell anaemia is the pain of vaso-occlusive crisis. It is the most common clinical presentation. Pain crisis may affect any tissue, but patients typically complain of pain in the chest, lower back, and extremities. Phases of vasoocclusive episodes have been described in children and adults¹⁴.

The pain could be acute, chronic or both. Unfortunately there is a tendency among caregivers to underrate the pain in its intensity, patients being seen as drug seekers or addicts.

We present the case of Patient AO, a 34 year old single male presently without a job. He gave a 6 year history of self medication with Pentazocine initially intravenously, converting to intramuscularly when all the veins in his right forearm, the only site where he did all the injecting were destroyed. He said it was easier to conceal the injection marks which was why he chose to use only one region of his anatomy, the right forearm. The last time he actually had a crisis was 2 years ago. He however agreed to injecting himself to feel 'high' and relaxed even if he had no symptoms of pain. He commenced injecting himself with one ampoule 8 hourly but over time this increased to as much as four hourly. He declined to reveal the source of his drugs. The patient lives alone and does not encourage any form of socialization with either family or friends. He denied having ever had any relationship with a significant other of the opposite sex. He admitted his only source of pleasure was Pentazocine. The patient went out of his way to ensure isolation from other people, and it appears he efficiently employed conditions available to him to successfully achieve this aim.

On examination he was a young man, pale ill looking and wasted. He was anicteric, conscious alert and coherent. Well orientated in person time and place. Fairly well groomed with good motor function. His memory was good both long and short term, however judgment appeared to be impaired. A review of his systems were essentially normal. There was no organomegaly. The arm itself revealed reduced muscle bulk, the forearm revealing a round circumscribed ulcer around the middle third of the forearm with dirty sloughs and swollen edges. The ulcer measured 10cm in length. There were swollen palmar and dorsal surfaces of the hand including the fingers.

Lab Findings revealed PCV 24%, WBC15,400/mm³, neutrophils 81%, eosinophils 04%, lymphocytes 15%, electrolytes and urea were essentially within normal limits.

Culture of the wound yielded growth of *Staphylococcus aureus* and was placed on appropriate antibiotics. He has been referred to the plastic surgeons who are considering stages of skin grafting due to the size of the lesion. The patient has also been referred to the psychiatrists for further evaluation and treatment.

The x-rays of the right forearm (injection site) revealed osteomyelitis and cellulitis.





DISCUSSION

Pentazocine is an (opiod) narcotic analgesic. It works on the brain and nervous system to reduce pain. It is metabolized in the liver and eliminated by the kidneys. It is useful for mild to severe pain such as is encountered in sickle cell disease(SCD). Patients with SCD have acute pain, chronic pain or both. Some times unfortunately pain as a symptom is often underrated in its intensity by caregivers. Patients are often perceived as drug seekers or drug addicts. However it said that less than 10% of patients are addicted , a number which is comparable to other diseases. Studies have shown that frequent emergency department use by these patients does not necessarily imply drug abuse or addiction.¹⁵

Acute pain can be managed with opiods , nonsteroidal anti-inflammatory drugs (NSAIDS), acetaminophen , or a combination of these medications. Immediate pain assessment and frequent reassessment with appropriate application of medications until pain relief is important. This is of considerable importance as it has been suggested that sub optimal treatment of painful episodes could actually be an important contributing factor to the development of drug abuse through self-medication.

The prevailing consensus for now appears to be that particular care and attention should taken before labeling a patient addicted . As already mentioned all too frequently the pain is underrated. This is due to the reluctance of physicians to give these patients adequate doses of analgesia due to concerns about addiction ,tolerance and side effects. The evidence suggests that a high percentage of sickle cell patients are perceived to be opioid dependent when in fact the percentage of this population of patients that are dependent is no higher than in the general population.¹⁶ However it should be noted that addiction to pain medication in sickle cell disease has been reported. Searches revealed Alao et al reported the case of a 38 year old female sickle cell anaemia patient , though the drug of choice in this instance was cocain.¹⁷Others have also reported addiction with cocain.¹⁸ This would tend to support the view that opioid addiction in sickle cell disease patients may probably be over exaggerated. However having said this Aghanwa et al reported the case of a young girl with Sickle cell disease who was addicted to Phernegan and pentazocine¹⁹.

CONCLUSION

Objective and careful regular assessment of sickle cell patients presenting with painful episodes should be carried out by experienced health caregivers. Each case should be taken on its own merit. It should be remembered that more and more patients have considerable knowledge about pain medication especially since the emergence and widespread use of the internet and should not be taken as an indication of possible drug addiction.

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