

Case report

# Sudden death due to sickle cell crisis during law enforcement restraint

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## Abstract

We report a case of vaso-occlusive sickle cell crisis in a young schizophrenic man with undiagnosed sickle cell trait who was restrained. Prior to being restrained he had locked himself in his apartment for two days without food or water. He was subsequently restrained, and transferred to hospital while handcuffed to the stretcher. He died suddenly during restraint. At autopsy, there was acute vaso-occlusive sickle cell crisis associated with hypernatremic dehydration. There were no injuries present. We conclude that the death was due to vaso-occlusive sickle cell crisis secondary to dehydration. It is important for the forensic pathologist to remember that death may occur suddenly during restraint from an unexpected mechanism other than excited delirium leading to cardiac arrhythmia or restraint asphyxia. © 2006 Elsevier Ltd and AFP. All rights reserved.

*Keywords:* Sickle cell trait; Sudden death; Restraint; Death in custody

## 1. Introduction

In the past decade, there has been increased awareness and investigation of sudden death during restraint. Excited delirium and restraint has been associated with sudden unexpected death. This topic is controversial and the exact mechanism of death in individual cases of excited delirium maybe unknown, or undecidable. However, different theories have been proposed to explain death in this circumstance, including excited delirium leading to cardiac arrhythmia and restraint asphyxia.<sup>1</sup>

Fatal cocaine intoxication may present as excited delirium in recreational cocaine users. In such cases, the individuals have clinical manifestations of intense paranoia, followed by bizarre and violent behavior necessitating forcible restraint. The symptoms are frequently accompanied by unexpected strength and hyperthermia. Cardiac dysrhythmia and fatal respiratory collapse may occur sud-

denly and without warning, rapidly after restraint. In a retrospective case study of 21 cases of unexpected death in people with excited delirium, all 21 cases of unexpected death associated with excited delirium had been restrained for violent agitation and hyperactivity. The restraint was either in the prone position (18 people [86%]) or by pressure on the neck (3 [14%]).<sup>1</sup> Another retrospective study of 11 cases and subsequent 21 cases of death during prone restraint are reported as examples of the common elements and range of variation in these apparent asphyxial events.<sup>2,3</sup>

However, it is also possible that death may occur suddenly during restraint from an unexpected mechanism such as cardiovascular disease or other lethal pre-existing pathology, drug intoxication, and neuroleptic malignant syndrome.<sup>4</sup> In most cases the causal issues are complicated and a definitive certification of cause of death may be difficult or controversial, even between forensic pathologists. Most decision-making difficulties arise when there is a definite or potential causal interaction between disease, injury, and circumstantial events.<sup>5</sup> Therefore, certification of death must not be precipitous despite high public interest in the death; the pathologist must exercise caution in ascribing

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a cause of death. All pre-existing natural conditions, toxicological findings, the findings of biochemical studies, and any other necessary ancillary investigations should be considered before giving a cause of death. Above all, the pathologist must be even-handed in fairness to all interested parties involved in the case including next-of-kin the law enforcement officers involved.

We report a case of sudden death during restraint of a young schizophrenic man with undiagnosed sickle cell trait who was restrained while in a catatonic state. This case illustrates many of the important dilemmas of a custodial death.

## 2. Case report

A 37-year-old man with a past history of schizophrenia was said to have been in a catatonic state in a locked apartment without eating or drinking for two days prior to the intervention by police and emergency medical personnel.

At the time of arrival of police and ambulance, he was conscious, alert and combative. He was then restrained by handcuffing and transferred to the ambulance in a supine position with a semi-agitated state. He was restrained from the time of arrest until he was brought to the emergency room where he was then handcuffed to the stretcher. He was given two separate intramuscular doses of 2 mg of midazolam while in the ambulance en route to the hospital. While he was supine on the stretcher he became unresponsive, and had no vital signs when brought into the emergency room. Resuscitation attempts were unsuccessful. His core temperature was normal in the emergency room.

There was past medical history of acute paranoid psychosis which was diagnosed in August 2004. Diazepam and olanzepine were present at the scene. However, it was unclear if he was taking these medications.

There was no history of drug abuse.

A postmortem examination was performed including layer-by-layer dissection of the ventral neck and face; dissection of the posterior neck; subcutaneous and musculo-skeletal dissection of the anterior and posterior torso; dissection of the extremities, wrists, ankles and testes. A complete histological examination of the major tissues was performed. Toxicological analysis of blood and urine was performed for cocaine, opiates, ethanol, benzodiazepine, olanzepine, medazolam and GC screen.

At autopsy, the body was that of a fully clad adult black male. There were no external injuries present on the body, and no ocular petechial haemorrhages. The neck was free of injuries. Subcutaneous dissection revealed focal subcutaneous haemorrhage on the ulnar surface of the right wrist, and overlying the right anterior superior iliac spine. There were no other significant injuries present on external or internal examination. Gross examination of the internal organs was unremarkable. The heart weighed was 360 g and was unremarkable with widely patent coronary arter-

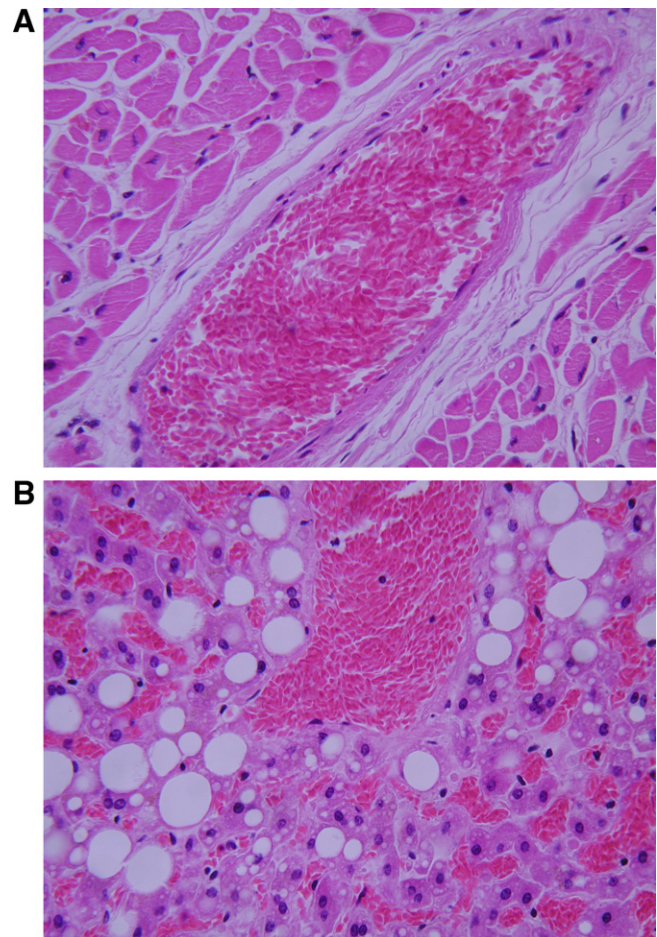


Fig. 1. (A). Vaso-occlusive sickling in the myocardium (H&E stain,  $\times 250$ ). (B). Vaso-occlusive sickling in the liver (H&E stain,  $\times 250$ ).

ies. The conducting system was not examined. No ante-mortem electrocardiogram was available.

Histological examination revealed widespread vaso-occlusive sickling of red blood cells in the brain, heart, liver, kidneys, adrenal glands, thyroid gland, intramyocardial coronary arteries, skeletal muscles, pancreas, testis and spleen (Fig. 1).

Vitreous biochemical studies showed findings compatible with hypernatremic dehydration with vitreous sodium of 150 mmol/L. Haemoglobin electrophoresis revealed a heterozygous phenotype for sickle cell anaemia, consistent with sickle cell trait. Toxicological analysis revealed only midazolam at therapeutic concentrations, compatible with administration during transport.

Death was attributed to vaso-occlusive sickle cell crisis due to hypernatremic dehydration in a man with undiagnosed sickle cell trait.

## 3. Discussion

Custodial deaths, particularly deaths during restraint, often raise public interest. Many stakeholders have a legitimate interest in knowing the cause of death. Therefore, it

is of paramount importance that a forensic pathologist perform a complete and careful autopsy by taking advantage of all necessary investigative avenues. Properly obtained information of the history of the incident will invariably guide the forensic pathologist. As performed in this case, the autopsy should often include a complete layer-by-layer dissection of the ventral neck and face after evisceration and removal of the brain to facilitate a bloodless field in the neck; dissection of the posterior neck; subcutaneous and musculoskeletal dissection of the anterior and posterior torso; dissection of the extremities, wrists, ankles; and dissection of testes with complete histological examination of tissues. Also, toxicological analysis and ancillary laboratory tests are mandatory considering the nature of these cases.

In this case, the absence of significant blunt force injuries excluded death due to assault. The absence of petechial haemorrhages in the conjunctivae and the absence of injuries in the neck exclude an asphyxial death due to an overt form of neck compression or strangulation. However, subcutaneous bruising of the wrist was indicative of handcuff restraint, which was not a variance with the history. The absence of gross macroscopic findings and the fact that the decedent was restrained in a catatonic state made the findings of ancillary investigations of paramount importance for deciding the cause of death after the gross autopsy. Despite this, the essentially negative autopsy raised the possibility of restraint or positional asphyxia, although the history was not particularly evocative of excited delirium.

Though some would disagree, positional asphyxia is a known complication when detainees are “hog tied” in the prone position in rear compartments of police patrol cars.<sup>6,7</sup> The decedent in our case was transported by ambulance rather than by police, and was handcuffed in the supine position rather than hog-tied or restrained in the prone position. Therefore, it is unlikely that positional asphyxia played a role as a cause of death, or as a contributory factor for the cause of death in this case. Similarly, the absence of cocaine or other sympathomimetic drugs indicates that the death was not due to excited delirium induced by chemical agents. The presence of midazolam in blood was within therapeutic range, and originated from medical intervention. Therefore, we were able to exclude that death was due to intoxication or drug overdose.

One other possible cause of death in a psychiatric patient in this circumstance is the so-called neuroleptic malignant syndrome. The alternative diagnosis of “acute exhaustive mania” has also been given to the sudden death of psychotic patients from apparently natural but undetermined causes.<sup>8,9</sup> This condition is poorly understood and occurs in people who are taking antipsychotic agents such as phenothiazines, butyrophenones, and thioxanthenes. However, the absence of neuroleptic medications as well as the presence of an alternative lethal cause (sickle cell crisis) reasonably excludes such a mechanism.

Since the decedent had locked himself in his apartment for two days without food or water in a catatonic state, he was dehydrated. Hypernatremia in the vitreous fluid supported this history. The decedent was a black male and it has been reported that the heterozygous condition occurs in approximately 8% of the American black population. Unlike the homozygous state (sickle-cell disease), sickle-cell trait is not widely recognized as a cause of life-threatening illness. Nevertheless, under unusual circumstances such as dehydration, hypoxia, acidosis and physical exertion serious morbidity or mortality can result from complications related to polymerisation of deoxy-hemoglobin S.<sup>10</sup>

In this case, histologic sections showed widespread sickling with vaso-occlusion. Although rare, exertional collapse and sudden death are the most serious potential complications of sickle-cell trait due to sickle cell crisis. Dehydration, hypoxia, acidosis and physical exertion are known aggravating factors for sudden death in sickle-cell trait due to vaso-occlusive sickle cell crisis. The vitreous sodium level was 150 mmol/L, which is indicative of hypernatremic dehydration. Since the autopsy was performed within a relatively short postmortem interval the reliability of the postmortem sodium level is reasonable. The hypernatremic dehydration could be due to abstaining from food and water for two days due to his catatonic state. Therefore, we can conclude that dehydration likely triggered vaso-occlusive sickle cell crisis with the subsequent violent outburst and/or the physical exertion during restraint possibly contributing to the sickling process.<sup>11–14</sup>

In summary, we have presented a case of sudden death from vaso-occlusive sickle cell crisis due to hypernatremic dehydration in a schizophrenic patient with sickle cell trait. This case indicates that not all deaths occurring during restraint are necessarily due to mechanical asphyxia or excited delirium leading to cardiac arrhythmia, but may be due to a previously undiagnosed condition that is fatal.

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