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Case report

Diencephalic storm in trauma patients: Is it really that rare

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Abstract

Introduction: Diencephalic storm is characterized by extreme episodic catecholamine release in the presence of a stressor and it is usually refractory to standard antihypertensives. The treatment of choice during the crisis is propofol and the best preventative measure is to remove the stressor (i.e. ventilator).

Case presentation: A 32-year-old male sustained 2nd and 3rd degree burn to 25 percent of the body surface area that included a severe inhalation component. The patient was admitted to the Burn Intensive Care Unit at the Arnold Luterman Regional Burn Center in Mobile, AL. The patient had frequent episodes (3-5 per day) of severe agitation that were accompanied by extreme tachycardia of > 200 beats per minute and hypertension (280/140). The inciting event was often endotracheal suctioning, but less noxious stimulation also resulted in similar episodes. During these episodes, the patient had significantly elevated catecholamine levels that improved after extubation. The patient's symptoms were refractor to standard antihypertensives but immediately resolved when given propofol. Further episodes of the diencephalic storm were treated successfully with propofol. Once the patient was removed from mechanical ventilation, there were no further episodes.

Conclusion: Diencephalic Storm may be difficult to diagnose due to a lack of familiarity with this rare entity. Any patient with severe agitation combined with the effects of episodic large catecholamine surges should be considered to have Diencephalic Storm. The standard immediate treatment is propofol due to the lack of responsiveness of standard antihypertensives and the removal of the stressor.

Introduction

The diencephalic storm is a rare syndrome characterized by the episodic release of catecholamines resulting in severe hypertension, tachycardia, tachypnea, and hyperthermia in response to external stimuli. It is known by various other terms, including paroxysmal sympathetic storm and diencephalic seizures. If appropriately diagnosed, there are effective treatment options previously described for this rare syndrome. Here we present a case of a diencephalic storm successfully treated with propofol and performed a review of the literature regarding the prevalence and treatment options.

Case presentation

A 32-year-old male was admitted to the Burn Intensive Care Unit at the Arnold Luterman Regional Brun Center after sustaining partial and full-thickness burns to 25 percent of his body surface area, which also included a severe inhalation component. The patient had frequent episodes (3-5 per day) of severe agitation that were accompanied by extreme tachycardia (>200 beats per minute) and severe hypertension (i.e., 280/140). The inciting event was often endotracheal suctioning, but less noxious stimulation also resulted in similar episodes. During these episodes, the patient had significantly elevated catecholamine levels that improved after extubation (Table 1). The patient's symptoms were refractory to standard antihypertensives but immediately resolved when administered propofol. Further episodes of the diencephalic storm were treated successfully with propofol. Once the patient was removed from mechanical ventilation, there were no further episodes suggesting that mechanical ventilation was the stressor initiating the syndrome.

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Table 1: Catecholamine levels during and after an episode of Diencephalic Storm.

	Dopamine	Norepinephrine	Epinephrine
Normal	<20	80-520	10-200
Symptomatic	49	1190	630
After Extubation	30	906	132

Discussion

The diencephalic storm is commonly associated with traumatic brain injury, hypoxic or ischemic brain injury, brain tumors, or hydrocephalus [1]. In the case presented herein, there was no evidence of common etiologies of this syndrome (i.e., brain injury). We believe the etiology of the diencephalic storm in our case was intra-tracheal suctioning while on mechanical ventilation. The diencephalic storm is thought to be dysfunction or interruption in certain autonomic pathways, particularly in the diencephalon, that lead to catecholamine releases and therefore, a hyperadrenergic state [2,3]. It is a very rare syndrome that is difficult to diagnose. Other conditions must be ruled out, including thyroid storm, pheochromocytoma, or drug-induced states. There is no definitive cause for its development, and it remains a diagnosis of exclusion.

Many different treatments have been described in the literature for the successful management of diencephalic storms. Currently, there is no standard therapy. Successful treatments previously described include the use of alpha-blockers, non-selective beta-blockers, opioids, and dopamine agonists [2-4]. However, none of these were able to resolve our patient's symptoms after multiple attempts with many of these agents, including opioids, beta-blockers, and calcium channel blockers. We discovered that intravenous propofol resulted in immediate resolution of each of the episodes of the sympathetic storm in our patient. The severe tachycardia and malignant hypertension were easily reproduced with any manipulation of the endotracheal tube, including suctioning the trachea-bronchial tree. Indeed, each of these episodes was immediately treated with propofol and the symptoms resolved immediately. Upon extubation, the patient had no further episodes of the diencephalic storm. Therefore, removal of the inciting stimulus is the definitive treatment for the signs and symptoms associated with diencephalic storms [1].

Conclusion

In conclusion, the diencephalic storm may be difficult to diagnose due to a lack of familiarity with this rare entity. Any patient with severe agitation combined with the effects of episodic large catecholamine surges should be considered to have a diencephalic storm. While there is no standard treatment, as shown in our case, propofol is a fast-acting agent that is effective in patients not responsive to standard antihypertensives. The definitive treatment is the removal of the stressor.

Contributions

Principal investigator: JDS. All authors participated in the development of this manuscript, including pertinent revisions.

Meeting: This paper was presented by CYQ at Southeastern Surgical Congress. Jacksonville, FL. February 2013.

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