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Short Communication

Adrenal crisis during a trip in a young child with septo-optic dysplasia

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Highlights

- Long-distance trips may be a precipitating factor for AC.
- Young children with SOD are at high risk of trip-associated AC.
- Prophylactic administration of a stress dose of hydrocortisone during trips may prevent AC.

Key words: adrenal crisis, septo-optic dysplasia, trip

Introduction

Adrenal crisis (AC) results from an absolute or relative cortisol deficiency (1). Previously identified precipitating factors for AC encompasses infection, surgery, and psychological stress (1). While a previous study indicated trip as a potential precipitating factor for AC (2), there is a lack of information regarding individuals susceptible to trip-associated AC. Herein, we report a case involving a young child with septo-optic dysplasia (SOD) who developed AC during a trip.

Case Report

The patient was a 16-mo-old Japanese diagnosed with SOD and complicated by deficiencies in adrenocorticotropin and thyrotropin, along with intellectual disability. The patient resided in Ehime prefecture and had no history of AC during the four previous visits to Keio University Hospital in Tokyo between the ages of 10 and 15 mo. He was referred to our hospital for craniosynostosis surgery. Perioperative management during cranioplasty for craniosynostosis and detailed medical information from infancy have been previously reported in the case of this patient (3).

At 16 mo of age, the patient demonstrated the ability to sit unaided and utter a few meaningful words, including “mama” and “papa”, but was unable to pull himself up. At that time hydrocortisone 12 mg/m^2/d and levothyroxine 1.6 μg/kg/d wee prescribed to him. Two days before his scheduled visit to our hospital, he traveled from Ehime to Tokyo for 7 h (by train, airplane, and car). On the day preceding his visit, the patient and his parents stayed in a hotel room all day. On the subsequent morning, namely on the visit day, his mother observed that he had become unresponsive, prompting an emergency visit. The patient presented with a Glasgow Coma Scale score of 7, blood pressure of 84/75

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mmHg, heart rate of 117/min, and a body temperature of 36.5°C. Peripheral coldness was noted during physical examination. Laboratory data indicated normoglycemia, normal electrolyte balance, and mild elevations in neutrophils (7,770/μL) with a white blood cell count of 14,000/μL, neutrophils at 55.5%, and lymphocytes at 36.0%. The C-reactive protein (CRP) level was measured at 0.58 mg/dL. A serum cortisol level of 20.3 μg/dL was observed two hours after the administration of 3 mg (equivalent to 5.6 mg/m^2) of hydrocortisone. We suspected AC and administered intravenous hydrocortisone 40 mg/m^2 (0.4 mL of hydrocortisone solution). The patient exhibited improved responsiveness within half an hour later, regaining full consciousness an hour later. Based on the rapid improvement in consciousness following hydrocortisone administration, we clinically diagnosed him with AC (1).

Twenty-four hours post-admission, the patient developed intermittent fever, which subsided within 60 h without the need for antibiotics. Intravenous hydrocortisone 40 mg/m^2 (0.4 mL of hydrocortisone solution) was administered every 8 h, totaling 120 mg/m^2 of hydrocortisone per day. Considering the patient's age and the presence of adrenal insufficiency of unknown cause, we maintained the intravenous administration of 120 mg/m^2 of hydrocortisone per day for 5 days to prevent the recurrence of AC (4). Throughout the clinical course, neither respiratory nor gastrointestinal symptoms were observed, and the peak level of CRP reached 1.7 mg/dL. The patient was discharged on the sixth day of hospitalization. On the day of discharge and the following day, hydrocortisone (36 mg/m^2/d) was administered. Subsequently, we advised the parents to administer 60 mg/m^2 of oral hydrocortisone daily from the departure day until the return day during travels. We determined the hydrocortisone dosage to be 60 mg/m^2/d according to the recommendations outlined in the latest clinical guidelines for 21-hydoxylase deficiency (5). Up to the age of 4 yr old, the patient did not experience AC during five different trips, including two visits to our hospital.

Ethical statement

This study complies with all the relevant national regulations and institutional policies, is in accordance with the tenets of the Helsinki Declaration, and was approved by the ethical committee at Keio University School of Medicine (20170130). Written consent was obtained from the patient's parents.

Discussion

In this article, we present the case of a young child with SOD complicated by intellectual disability and adrenocorticotropin deficiency, who experienced an AC during a trip.

We hypothesized that the long-distance trip, requiring prolonged periods in unfamiliar environments, such as vehicle travel and hotel stays, as a potential precipitating factor for AC in our patient. Consistently, White reported that approximately 10% of AC events occurred outside the home, including overseas locations (7.7%); during travel on planes, boats, or trains (1.5%); or on day trips (1.3%) (2). While fever on the day following admission in our case could be associated with a viral infection, we believe that infection was not the primary precipitating factor. This is substantiated by the absence of fever and the presence of a mild inflammatory response during the AC event. Our speculation is further supported by previous studies that revealing elevated CRP levels in the absence of obvious infection during AC events (6) and an increase in neutrophil count due to catecholamine secretion (7). Further studies are warranted to determine whether long-distance trip constitutes a significant precipitating factor for AC among children with primary or secondary adrenal insufficiency.

Due to his young age and intellectual disability, our patient was unable to articulate his symptoms, introducing a potential challenge in the early detection and timely treatment of AC. Consequently, we identify both young age and intellectual disability as predisposing factors for AC in our patient. Moreover, young children often experience abrupt fevers due to viral infections, which can serve as triggers for AC. Considering these factors, supplementing a stress dose of hydrocortisone during trips may prevent additional AC episodes in our patient. Given that all patients under daily hydrocortisone supplementation face a potential risk of developing AC during long-distance trips, it is imperative for physicians to educate patients or their parents about the risks of trip-associated AC. Recommending the administration of stress doses of hydrocortisone as needed while traveling becomes crucial. However, there is currently no evidence supporting the benefits of prophylactic stress dosing of hydrocortisone in preventing trip-associated AC.

In summary, we report a young child with SOD who developed AC during a trip. We consider that young children with intellectual disabilities and adrenocorticotropin hormone deficiency, such as SOD, are at high risk of trip-associated AC.

Conflict of interests: All authors declare no relevant financial relationships.

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