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COVID-19 Pneumonia in a Congenital Adrenal Hyperplasia; A Case Report

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1. Abstract

Congenital Adrenal Hyperplasia (CAH) is managed with long-term corticosteroid and is suggested to higher dose, based on the general sick day principles. I presented a 5-year-old CAH case by pneumonia managed in our pediatrics department. Considering stress doses of cortisone in management of CAH patients with COVID19 should be considered.

2. Key Clinical Message

Patients with CAH are at higher risk of rapid deterioration due to viral infection like COVID19 and their management should be done immediately by suitable strategy; considering stress dose of cortisone and routine etiology treatment.

3. Introduction

The prevalence of congenital adrenal hyperplasia is ranging between one in 10,000 to one in 67,000 live birth [1]. Studies have reported that individuals with adrenal insufficiency have an increased rate of respiratory infection-related deaths, possibly due to impaired immune function and they are more likely to develop a severe course of the disease as well [2, 3]. European Society for Pediatric Endocrinology (ESPE) recommends increasing the hydrocortisone dose, according to the general "sick day rules" in children with congenital adrenal hyperplasia. The suggested oral stress dose for an adult is 20 mg hydrocortisone every 6 hours, if they deteriorate more during acute coronavirus disease 2019 (COVID-19) infection, they advise immediate (self-)injection of 100 mg hydrocortisone intramuscularly, followed by intravenous infusion of 200 mg hydrocortisone per day, or until this can be established, administration of 50 mg hydrocortisone every 6 hours. Also, advise on doses for infants and children [3].

We present a case report of a child with a previous diagnosis of congenital adrenal hyperplasia (CAH) who was found to have SARS-CoV-2 infection.

4. Case Presentation

A 5-year-old child with CAH presented to the emergency department with fever and chills, abdominal pain, recurrent vomiting, cough and loss of appetite since 5 days ago. He was a term baby with a history of CAH (21- hydroxyprogesterone deficiency) from infancy and was operated for it and was on treatment with corticosteroid (Hydrocortisone 10mg ½ tablet t.i.d). He had two brothers with CAH who were about 21 years old and 23 years old with normal development. His parents reported symptoms of cough and a runny nose for a few days prior to this. At presentation, he had tenderness in right upper quadrant in abdominal examination. He had hypotension (BP: 75/pulse) and tachycardia (HR=130/min) and hypoxemia (85%) with tachypnea (RR=52/min). He had no symptoms of meningismus. He had no dermatological involvement. His eyes and throat were normal in exam. In right lung he had decrease in lung sound.

Pediatric endocrinology was consulted and according to his sepsis, stress dose of hydrocortisone (100 mg/m2) was started. Oxygen therapy was started and portable chest X-ray was done. It showed lower lobe pneumonia in right lung with little pleural effusion which was confirmed by chest sonography (consolidation was seen in lower and middle lobes of right lung) (Figure1). Antibiotic therapy for him was started by Ceftriaxone and Clindamycin. Vitamin C, famotidine and zinc gluconate was started. He had a rise in

creatinine (1.5 mg/dL) in his laboratory exam with high CRP (66 mg/dL) with no leukocytosis or lymphopenia. His other exam was in normal range. He received a normal saline bolus and was then placed on maintenance IV fluids. His echocardiography showed no pericardial effusion or low ejection fraction. According to immunosuppressive status of patient (under treatment with long-term corticosteroid) and pandemic of COVID19, PCR test for detection of SARS-CoV-2 was sent. After 48 hours his CBC showed lymphopenia (ALC: 720), rise in CRP (97 mg/dL) and SARS-CoV-2 was detected on nasopharyngeal swab. Blood culture, urinalysis, and urine culture were negative. In his abdominal sonography which was done for his recurrent vomiting, it showed two small stone in right kidney (the largest was 3.5 mm in size) and several lymphadenitis (the largest lymph node was 5cm in size). After 24 hour of treatment, his hypoxemia was cured and he did not dependent on oxygen. His tachypnea and respiratory distress was resolved. He received acetaminophen and ondansetron for abdominal pain and vomiting. According to hypotension, inotrope (Dopamine 5μg/Kg/min) was started and patient was transferred to PICU for better monitoring and isolation. His antibiotic was changed to Meropenem and Vancomycin. After 72 hours of admission, his inotrope was discontinued and his fever and vomiting was stopped. Over the course of his admission, he remained hemodynamically stable. He fed appropriately and did not require respiratory support. After 7 days of admission, his vital sign was stable and he had no complaint of vomiting, cough or abdominal pain and his lymphopenia was recurred. He was discharged by advice in quarantine for 2 weeks, oral antibiotic therapy and stress hydrocortisone dose was weaned down to his home regimen. He was followed on the day 14 after admission in clinic and had no complaint and his CXR shows improvement of pneumonia without effusion.

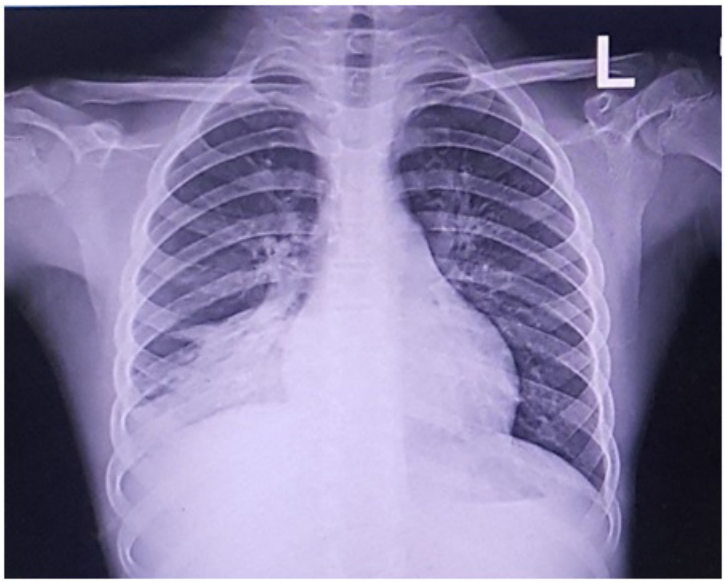

Figure1: Chest X-Ray of patient at admission

5. Discussion

My case suggests that SARS-CoV-2 in a child may demonstrate a fairly stable clinical course even in the presence of underlying adrenal insufficiency. In a longitudinal assessment of 156 patients with CAH, gastrointestinal and upper respiratory illnesses were the predominant reasons for stress dosing, adrenal crises, and hospital admissions [4].

Patients with adrenal insufficiency are at risk to develop a potentially life-threatening adrenal crisis if experiencing major stress, such as an acute illness. This requires administration of increased doses of glucocorticoid replacement to prevent and, if already in progress, treat the adrenal crisis [5, 6]. Adrenal crises are regularly observed in patients with PAI and SAI [7, 8] and contribute to the observed increased mortality in these patients.

There are limited cases diagnosed by COVID-19 and CAH. It can because of good compliance of patients and their parents, awareness of their families about stress dose of corticosteroids during illness phase and maybe their more cautions in performing prevention strategies. Subjects with immunocompromised status or with certain chronic diseases are included in the list of diseases requiring additional precautionary measures to reduce risk of COVID-19 and those patients with CAH known to have an element of immune deficiency because of defective action of neutrophils and natural killer cells , as well as having cortisol deficiency and on lifelong treatment, because of this factors, the risk of infection in that patient higher than the normal population by two to eight-fold [9].

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was also noted that stress dosing was more common in children compared to adults which could potentially be attributed to the fact that parents tend to be more vigilant about their children's presentation and need for management compared to adults treating themselves [4].

6. Conclusion

I here presented a known case of CAH with pneumonia treated classically and was improved clinically in follow up. Considering stress dose of cortisone in CAH cases should be considered beside routine etiology treatment.

7. Acknowledgement

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