Cystic Fibrosis with COVID-19 Disease

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Abstract

Based on experience with other viral respiratory illnesses, patients with cystic fibrosis were believed to have worse prognosis when infected with Coronavirus disease 2019. We report a case of cystic fibrosis in a young girl who developed coronavirus disease 2019 with short-term evolution of the disease and good recovery with no known major long term sequel.

Keywords: Cystic fibrosis, COVID-19, Oman, steroids, IL-6,

Introduction

Coronavirus disease 2019 (COVID-19) is a novel coronavirus that initially emerged in China in December 2019 but could spread widely all over the globe in a very short time [1]. The clinical manifestation of the disease varies from asymptomatic to severe respiratory failure requiring assisted ventilation and oxygenation measures [2]. Whereas some unfavorable prognostic factors were identified in the general population like male gender, age and uncontrolled comorbid diseases, very less is known about the effects of COVID-19 disease in cystic fibrosis (CF) cases [3].

Although CF patients always had increasing risk of complications from respiratory viral infections, no much is known if this is also applicable for COVID-19 disease [4].

Case Presentation

A 14 year old girl who is known case of CF with restrictive lung disease, exocrine pancreatic insufficiency and failure to thrive. She is known to have homozygous variant (c.1069 G>A, p.Ala357Thr(A357T)) of the disease and her respiratory cultures chronically grew pseudomonas. She presented to a tertiary care hospital in the Sultanate of Oman with four days history of increasing lethargy and fatigue with increase in her chronic productive cough associated with yellowish sputum occasionally. This was also associated with un documented fever and decreased oral intake. Up on presentation to the emergency room she was found to be hypoxemic with saturation of 60% (via pulse oximeter) in room air that picked up to 94% with 15 liters of oxygen via non rebreathing mask. Her respiratory rate was 32 breaths per minute. Her blood pressure was 90/52 mmHg and she was found to be dehydrated for which she was initiated on intravenous fluid. Her COVID 19 nasopharyngeal reverse transcriptase chain reaction (RT-PCR) came positive. Her presentation chest X-ray and admission and discharge laboratory investigations are listed in the table below. She was directly admitted into isolation ward and she was commenced on parenteral broad spectrum antibiotics as per sensitivity from her last sputum culture (piperacillin-tacobactam), dexamethasone (6mg intravenously as part of COVID-19 local management policy) and pharmacological deep venous thrombosis prophylaxis (enoxaparin 4000 IU subcutaneously once daily). Her clinical condition significantly improved in the following 3 days and she had no more increased cough, no fever and no significant shortness of breath. She was back to her baseline and she was able to maintain saturation of 95% and more in ambient air.

Table 1: Admission and discharge laboratory investigations

Laboratory investigation	Admission measures	Discharge measures	Reference range
Hemoglobin	9.7 g/dl	9.1 g/dl	11-14.5 g/dl
White cell count	17.7 x 10^9/L	7.2 x 10^9/L	2.4-9.5 x 10^9/L
Neutrophils	14.2 x10^9/L	5.9 x 10^9/L	1-4.8 x 10^9/L
C reactive protein	204 mg/L	89 mg/L	5-30 mg/L

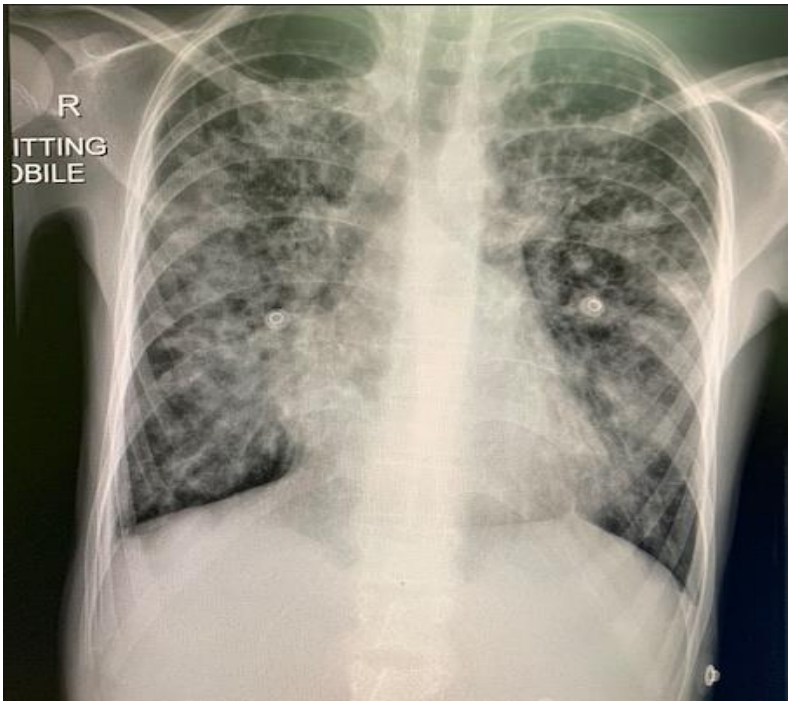

Figure 1: Chest X-ray on admission showing bilateral infiltrates involving more than 50% of the lung zones.

Discussion

CF is one of the fatal autosomal recessive inherited diseases that is estimated to affect 80,000 worldwide. It is caused by a mutation in the Cystic Fibrosis Transmembrane Regulatory (CFTR) gene [5]. To date, more than 2000 mutations have been identified with around 300 mutations known to cause the disease [6]. Although CF disease affects multiorgans including liver, intestine and pancreas, most of the CF morbidity and mortality are pulmonary in nature due to repeated respiratory inflammation and infection [7]. Although very limited data are available about the natural course of COVID-19 in CF patients, published evidence to date indicates lower incidence rates of COVID-19 disease among CF patients and no significant correlation was found between the two diseases [8]. At the initial stages of the pandemic, most specialists thought that this category of patients would have worse prognosis [3]. CF registry from 8 European countries described 40 patients with CF and confirmed COVID-19 disease did not show any unique risk factor for acquiring or severity of illness among this heterogeneous group [9]. Furthermore, another study from Spain showed lower mortality of CF patients when compared to general population [10]. On the contrary, in view of the global health crisis exerted by COVID-19 with the limitation of the ventilators and essential lifesaving resources, several health institutes had to put policies limiting their support to CF patients and other patients with chronic disabilities assuming that they will not have good prognosis if they are infected with COVID-19 disease [11].

Studies from different countries showed lower incidence of COVID-19 disease in CF patient (0.14%) in comparison to the general population (0.58%) [12]. Having CF patients being strictly and chronically adhering to the hand hygiene measures, masking and social distancing may have contributed to this significant low incidence [11,12].

The clinical manifestations of CF exacerbation attacks might overlap with active COVID-19 disease like cough, fever, increasing shortness of breath and change in the color or the consistency of the expectorated sputum like in our case. Differentiating CF exacerbation from COVID-19 disease may not always be straightforward and this reveals the importance of having low threshold for screening CF patients for COVID-19 [4].

Although the patient presented with severe COVID-19 disease (respiratory rate > 30 breaths per minute, oxygen saturation in room air < 93% and bilateral infiltrates in more than 50% of the lung fields in her chest X-ray) [13], she responded very well to the antibiotic and dexamethasone treatment and she was able to maintain saturation in room air after 3 days of treatment. She was discharged to continue home quarantine and she was back to her baseline. The full mechanism of mild COVID-19 disease course in general in the CF patients is not fully understood. It is thought that the disruption of interleukin-6 (IL-6) signaling in CF lungs that occur through increased serine protease release and subsequent cleavage of both membrane-bound and soluble IL-6 receptors may play an important role in having mild form of COVID-19 disease in CF patients. Moreover, most of the CF patients are on regular immunomodulatory medications (e.g. azithromycin) and DNase (mucolytic) that might also contribute to this relative protection [14].

Conclusion

CF patients are having low incidence and milder course of COVID-19 disease. It is very difficult to obtain the precise incidence of the disease in this category of the patients with chronic respiratory disease. The entire mechanism of having milder form of the COVID-19 disease among CF patients is not fully understood and it warrants closer evaluation and investigations of this

particular group of patients. Policies might need to be modified preventing any discrimination of the provided treatment and life saving measures for CF patients.

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