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Covid –PCD: A Case Report

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Abstract **Case Report**

Coronavirus disease 2019 (COVID-19) is a public health emergency of international concern. The global population lacks immunity to COVID-19 and is generally susceptible. It is reported that at least 2% of patients with COVID-19 have underlying lung disease, which is related to severe clinical manifestations and higher mortality compared with those without chronic lung diseases [1]. The existence of underlying conditions, especially chronic respiratory diseases, with long-term drug treatment, may affect the progress, treatment and prognosis of COVID-19. Here, we report the case of a 50 year old female who is a known case of Kartagener's syndrome and developed infective exacerbation of bronchiectasis complicating severe COVID-19 pneumonia with hypercapneoic respiratory failure which responded to anti-pseudomonal antibiotic coverage and non-invasive positive pressure ventilation therapy. The case in the current study indicated that SARS-CoV-2 infection precipitated bronchiectasis exacerbation in a patient of primary ciliary dyskinesia as limited data is available regarding this association and there is concern about its poor outcome.

Keywords: Primary Ciliary Dyskinesia, Kartagener's Syndrome, bronchiectasis, Severe COVID-19 pneumonia, Hypercapnoeic respiratory failure.

INTRODUCTION

COVID-19 pneumonia may turn out be a severe infection with high rate of mortality particularly in the elderly and the immunocompromised. Patients with chronic lung diseases are thought to be at an increased risk. The disease course and optimal treatment strategies might differ depending upon the underlying disease. Primary ciliary dyskinesia (PCD) is a multisystem, genetic disease which affects approximately 1 in 10,000 people and leads to chronic upper and lower airway disease, laterality defects, including congenital heart disease, and other health problems [2-5]. People with PCD *because of their abnormal ciliary function do not clear bacteria and other airborne particles from their upper and lower airways, which leads to chronic respiratory infections.* We know little about how the circulating novel strain of coronavirus affects people with PCD. Herein we report a case of COVID-19 pneumonia in a 50 year old female with Kartagener's syndrome who presented with type 2 respiratory failure.

CASE PRESENTATION

A 50 year old female who is a known case of kartageners syndrome presented to our ER with one week history of fever, cough with mucopurulent expectoration and breathlessness which had worsened over the last 3 days. She was diagnosed as Kartageners syndrome 10 years back and had recurrent history of lower respiratory tract infections requiring hospitalisations. Her follow up visits were infrequent and she was not compliant with her medications. She had not been vaccinated against SARS-CoV-2. On examination she was conscious, oriented, febrile and undernourished with grade 3 clubbing of all digits. Her blood pressure was 110/80 mmHg, heart rate of 112 beats/minute and respiratory rate was 26 breaths/minute. Her oxygen saturation was 84% while breathing ambient air and improved to 95% on 15l/min NRBM. On auscultation she had bilateral coarse crepitations and scattered rhonchi with squeaks. A nasopharyngeal swab taken tested positive for SARS-CoV-2 by RT–PCR. Chest radiograph showed bilateral patchy opacities in the middle and lower zones with reticular markings and cystic shadows and dextrocardia.

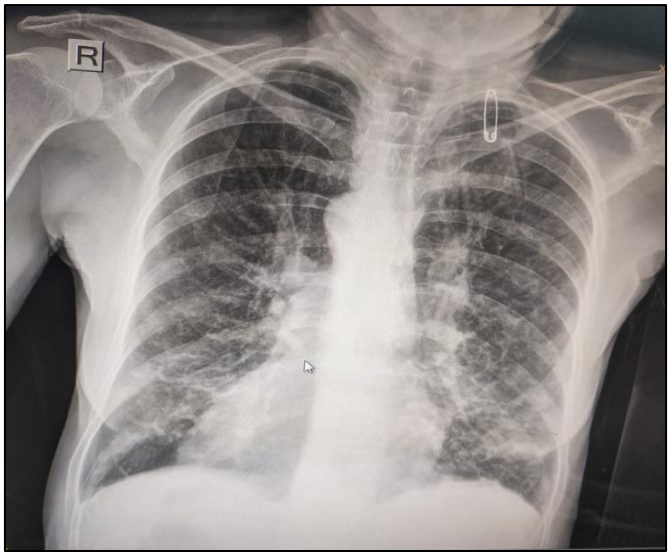

Fig. 1: Chest X-ray shows bilateral cystic changes and patchy infiltrates predominantly in the lower zones with dextrocardia

A plain HRCT of chest demonstrated peripheral patchy ground glass opacities mainly affecting bilateral lower lobes superimposed on cystic bronchiectatic changes with dextrocardia and situs inversus suggestive of COVID-19 pneumonia with a CT Severity Score of 12/25.

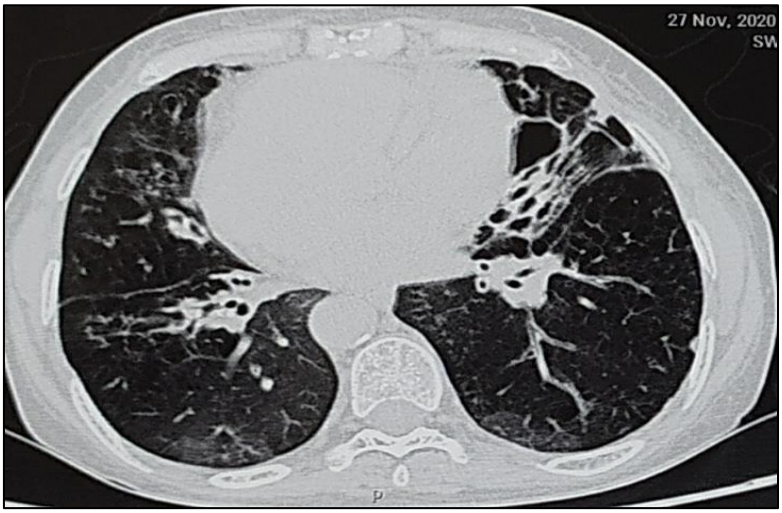

Fig. 2: Computed Tomography of chest showing bilateral cystic bronchiectasis with peripheral ground glass opacities

Her total leucocyte counts on admission were 17000/cumm with 90% neutrophilia, elevated CRP 326.85and d- dimer of 1.24µg/ml (normal < 0.5µg/ml). Her ABG on admission was suggestive of hypercapnic respiratory failure with normal lactates.

Table 1: Arterial blood gas analysis of the patient at the time of admission

Parameters	Value	Normal Range
Ph	7.26	7.35-7.45
PaO_2 (mmHg)	52.6	80-100
$PaCO_2$ (mmHg)	76.8	35-45
HCO_3^- (mmol/L)	32.6	22-26

PaO_2: partial pressure of oxygen in arterial blood; $PaCO_2$: partial pressure of carbon dioxide in arterial blood; HCO_3^-: Bicarbonate ion

A 2D echocardiography done revealed moderate pulmonary hypertension with PASP of 55 mmHg. She was shifted to ICU and started on nebulization, intravenous antibiotics with antipsuedomonal coverage, intravenous dexamethasone, mucolytics, inhaled bronchodilators and subcutaneous enoxaparin. She was initiated on non-invasive positive pressure ventilation using vented face mask with chest physiotherapy. She also received five doses of the antiviral remdesivir for features of covid pneumonia on

HRCT of chest. Her sputum culture grew pseudomonas aeruginosa and antibiotics were upgraded as per the sensitivity report. She gradually started improving with declining counts and FiO_2 requirement and improved ABG parameters with NIPPV support. She had 14 days of ICU stay before she was transferred to ward where overnight BIPAP support was given and pulmonary rehabilitation was introduced. She was discharged after 19 days of hospital stay on domiciliary oxygen therapy and overnight home bipap support with 14- day course of oral ciprofloxacin antibiotics and advised to take influenza, pnuemococcal and covid-19 vaccination.

DISCUSSION

Disorders of ciliary motility may be congenital or acquired. Congenital disorders are labeled as PCDs. Nearly 50% of PCD patients has situs inversus. Such cases of PCD with situs inversus are known as Kartagener's syndrome [6]. PCD is a phenotypically and genetically heterogeneous condition wherein the primary defect is in the ultrastructure or function of cilia [5]. Such defects are identified in approximately 90% of PCD patients and involve the outer dynein arms, inner dynein arms, or both.

Kartagener's syndrome (KS) is a rare autosomal recessive genetic disorder which was first described by Siewert in 1904; however, Kartagener recognized the clinical syndrome in 1933. The syndrome includes the clinical trial of chronic sinusitis, bronchiectasis, and situs inversus [7, 8]. Camner *et al.*, first suggested ciliary dyskinesia as the cause of KS in 1975. In 1977, Eliasson *et al*., first coined the term "immotile cilia syndrome" for KS to categorize infertility with chronic sinopulmonary infections [9, 10].

Normal ciliary function is critical for respiratory host defense and motility of sperm, and ensures proper visceral orientation during embryogenesis. In KS, the gene mutation at *DNAI1* and *DNAH5* leads to impaired ciliary motility, which predisposes to recurrent sinopulmonary infections, infertility, and errors with left–right body orientation [11].

Our patient presented with COVID -19 pneumonia complicated by bronchiectasis exacerbation in a known case of Kartagener's syndrome. The usual presentation in COVID-19 pneumonia is fever, dyspnea and acute respiratory symptoms which can lead to refractory pulmonary failure. It is common among patients with COVID-19 pneumonia to develop hypoxemic respiratory failure due to acute respiratory distress syndrome (ARDS), a life-threatening form of respiratory failure. Here, our patient presented with a picture of COVID-19 pneumonia with hypercapnoiec respiratory failure which responded to non-invasive positive pressure ventilation therapy (NIPPV). COVID-19 patients have a relatively low rate of secondary bacterial infection. These infections are commonly observed in the critically ill patients .The patient in the current study had superimposed bacterial infection of the respiratory tract. Clinicians should be aware that SARS-CoV-2 infection is probably a precipitating factor of bronchiectasis exacerbation while bronchiectasis is a risk factor for greater severity of SARA-CoV-2 infection. One may hypothesize that the novel corona virus triggers acute bronchial inflammmation and exacerbation of the chronic disease of airways. As regards bronchiectasis, although the findings of studies on COVID-19 and bronchiectasis are conflicting [12-15], it should be noted that when making comparisons in patients with COVID-19 with or without bronchiectasis, the former are more likely to experience the severe manifestations of the infection, may require oxygen therapy or hospitalization and may be even more likely to require admission to an ICU or to succumb to the disease. In the present case, the patient suffered from a syndrome characterized by an obstructive lung pattern and bronchiectasis, and required ICU admission with oxygen therapy and non-invasive ventilation, fortunately she had a favorable outcome. Another interesting fact was that the patient had a favorable outcome despite the fact that she was unvaccinated against COVID-19. It is well established that this is an additional risk factor for hospitalization and poor outcomes [16].

CONCLUSION

In conclusion, the present study describes an interesting case of COVID-19 associated pneumonia in a patient with Kartagener's syndrome. Patients with this syndrome are vulnerable to COVID-19 as are patients with other chronic lung diseases, requiring oxygen therapy and hospitalization. However to the best of our knowledge there are currently no sufficient data available on the effects of COVID-19 in these patients, their optimal management, or the role COVID-19 vaccine in their clinical course.

We do not know how SARS-CoV-2 affects people with PCD and whether severity of PCD, age and other patient characteristics influence COVID-19 disease course. PCD registries and research databases are not designed for a longitudinal collection of patient-reported information on symptoms and health behaviour in real-time [17]. However, such data are needed to comprehensively understand the impact of SARS-CoV-2 on people with PCD. Patient-reported data are also essential to understand how public health measures that aim to reduce the spread of COVID-19 are adopted by people with PCD, and how the pandemic affects their life and medical care.

Standard treatment for sinopulmonary problems in people with KS includes chest physiotherapy, mucolytics and antibiotics. A long-term low-dose prophylactic antibiotic is required in those with frequent exacerbation of bronchiectasis (≥3

times/year). Influenza and pneumococcal vaccination should be routinely advised. Vaccinations against COVID-19 can prevent the spread of the disease and can protect against severe disease in case of infection. People with primary ciliary dyskinesia (PCD) may be at higher risk of severe COVID-19 disease and are therefore strongly recommended to get vaccinated.

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