COVID-19-associated pneumonia in Swyer-James-MacLeod syndrome: A case report

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Abstract. Coronavirus disease 2019 (COVID-19) exerts differential effects on various individuals. The majority of infected individuals experience mild-to-moderate disease and usually recover, without requiring hospitalization. It has been reported that those who have underlying chronic diseases are more susceptible to infection and may thus develop significantly more serious illness. As a result, COVID-19 may aggravate pre-existing respiratory illnesses, such as interstitial lung disease, chronic obstructive pulmonary disease and asthma. Swyer-James-MacLeod syndrome is an uncommon clinical condition marked by post-infectious infantile bronchiolitis obliterans. Traditionally, the diagnosis is made in infancy following an investigation for reoccurring respiratory infections, although in rare cases, the diagnosis is made in adulthood. The present study describes the case of a 45-year-old patient with Swyer-James-MacLeod syndrome hospitalized due to COVID-19, which is the first one to be reported. To the best of our knowledge, there are currently no data available on the effects of COVID-19 in these individuals, stheir optimal therapy, or the impact of COVID-19 vaccination on their clinical course. Thus, it is hoped that the present study sheds some light into this condition.

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Introduction

On March 11, 2020, coronavirus disease 2019 (COVID-19) was recognized as a pandemic by the World Health Organization (WHO) (1). It has thus far affected >600 million individuals worldwide, with a total of 6.47 million related deaths (https://www.worldometers.info/coronavirus/). It has been highlighted that 75% of patients hospitalized for COVID-19 have at least one COVID-19-associated comorbidity. It has been reported that those with underlying chronic diseases are more prone to infection and to becoming seriously ill (2,3). Patients with COVID-19 who have a history of cardiovascular disease, malignancy, obesity, chronic respiratory disease, diabetes mellitus, or neurological disorders have the worst outcomes and are more susceptible to developing pneumonia or acute respiratory distress syndrome (2,3).

COVID-19 can affect the respiratory tract via a number of mechanisms and at varying degrees of severity, depending on the individual's immune system, age and comorbidities. Symptoms can vary from minor to severe, including cough, dyspnea and fever, as well as respiratory failure, shock and multi-organ system failure. As a result, prior respiratory disorders, such as interstitial lung disease, asthma and chronic obstructive pulmonary disease (COPD) may be exacerbated by COVID-19 (4).

Swyer-James-MacLeod syndrome is an uncommon clinical entity associated with post-infectious infantile bronchiolitis obliterans. It is characterized by pulmonary arterial hypoplasia and/or agenesis, resulting in the hypoperfusion of the lung parenchyma. Traditionally, the diagnosis is confirmed in infancy following an inquiry for recurring respiratory infec-

tions; however, in certain cases, patients with minimal or no bronchiectasis have few or no symptoms, and the syndrome may thus remain undetected until adulthood (5). Other symptoms include wheezing, breathlessness on exertion, decreased exercise tolerance and cough, which may be accompanied by hemoptysis and chest pain (6).

The most common finding associated with this condition on pulmonary function tests is airflow obstruction (7), while the presence of pulmonary hyperlucency on chest imaging is a hallmark of the syndrome (8). Swyer-James-MacLeod syndrome complications include recurrent infections, particularly in patients with bronchiectasis, lung abscess and pneumothorax (9,10). The cornerstone of treatment is conservative management. Surgery is only used in certain situations, such as when a patient has recurrent lung infections, does not respond to treatment, or has symptoms that are insufficiently handled by the most appropriate medical care. Pneumonectomy and lung volume reduction procedures, such as lobectomy and segmentectomy are surgical treatment options (6).

The present study describes the case of a 45-year-old patient with Swyer-James-MacLeod syndrome hospitalized due to COVID-19, which is the first one to be reported to date, at least to the best of our knowledge.

Case report

A 45-year-old male patient (non-smoker) presented to the department of Infectious Diseases-COVID-19 unit of Laiko General Hospital with complaints of fever, post-nasal drip, dry cough and dyspnea at rest over the past 12 days. He had a medical history of Swyer-James-MacLeod syndrome and gastroesophageal reflux. His medications included omeprazole and a combination of inhaled indacaterol/glycopyrronium bromide.

A clinical examination of this respiratory system revealed crackles on auscultation in all lung fields of the right lung and diminished lung sounds in all lung fields of the left lung. The evaluation of the other bodily systems did not reveal any abnormalities. His blood pressure was 135/80 mmHg, his heart rate was 120 beats per minute, his oxygen saturation in room air was 89%, and his body temperature was 38.5˚C. An electrocardiography revealed sinus tachycardia.

Arterial blood gas analysis revealed a partial pressure of oxygen (pO_2) of 56 mmHg, a partial pressure of carbon dioxide (pCO_2) of 28 mmHg, pH 7.51 and bicarbonate ($HCO3^-$) levels of 22.3 mmol/l in room air. A chest X-ray revealed patchy infiltrates in the right lung lower lobe and a hyperlucent left lung (Fig. 1).

The laboratory analyses included a complete blood cell count, serum biochemistry and coagulation tests. Notable laboratory findings included C-reactive protein levels of 133.17 mg/l (normal, <6 mg/l), ferritin levels of 710 ng/ml (normal range, 150-400 ng/ml) and d-dimer levels of 0.65 μg/ml (normal, <0.5 μg/ml). The procalcitonin test was negative (0.03 ng/ml; normal, <0.1 ng/ml).

The patient had a positive detection of severe acute respiratory syndrome 2 (SARS-CoV-2) nucleic acid in the examined nasopharyngeal sample using reverse transcription-polymerase chain reaction (RT-PCR). The patient was unvaccinated against SARS-CoV-2. He was hospitalized in the COVID-19 unit and was administered oxygen therapy with a Venturi mask supplying 35% oxygen, along with prophylactic subcutaneous enoxaparin at a daily dose of 40 mg, and intravenous dexamethasone at a daily dose of 6 mg, as well as remdesivir (200 mg on the first day, followed by a daily dose of 100 mg for the following 4 days) (11). The duration of therapy was 5 days for all agents.

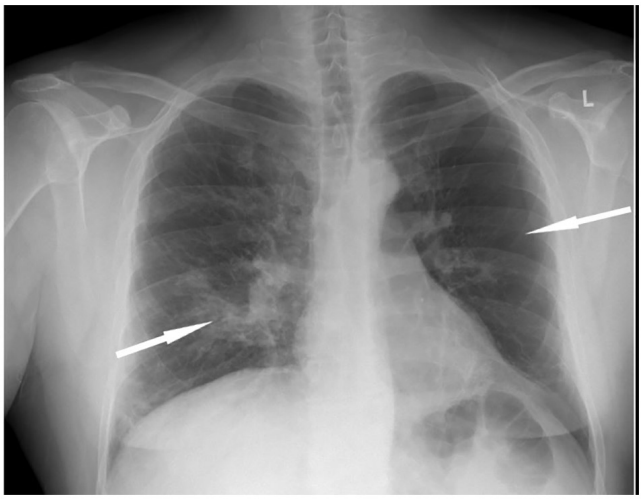

Figure 1. Chest X-ray illustrating patchy infiltrates in the right lung lower lobe (arrow) and a hyperlucent left lung (arrow).

The patient was also advised to continue receiving the combination of inhaled bronchodilators during his hospitalization. He underwent a computed tomography pulmonary angiogram, which revealed no pulmonary embolism, and a high-resolution computed tomography scan of the chest, which revealed lung infiltrates in all lung fields of the right lung, as well as a hyperlucent left lung with bronchiectasis and much fewer lung infiltrates (Fig. 2). Furthermore, a sputum culture was performed that did not reveal any microorganisms, and therefore the patient did not receive any antibiotics.

Following 3 days in the hospital, his fever had subsided and his oxygen levels had improved. He exhibited a gradual recovery and was discharged on the 6th day of hospitalization without the need for supplemental oxygen; a new arterial blood gas analysis revealed a pO_2 of 86 mmHg, a pCO_2 of 34 mmHg, pH 7.44 and $HCO3^-$ levels of 23.7 mmol/l in room air.

Discussion

To the best of our knowledge, this is the first reported case of COVID-19-associated pneumonia in a patient with Swyer-James-MacLeod syndrome. Since the first cases of COVID-19 were reported, COPD has been linked to a greater likelihood of a poor prognosis, as measured by the number of hospitalizations and mortality (12-14). Chronic inflammation (usually T2) and bronchial remodeling, both of which are common hallmarks of asthma, may enhance sensitivity to COVID-19. It has been debated whether asthma increases the chance of infection; however, new research suggests that the risk is limited to severe types of the disease (15). A recent study found that individuals with asthma had a higher risk of hospitalization due to COVID-19 than those without asthma, and that those who had two or more oral corticosteroid bursts in the preceding

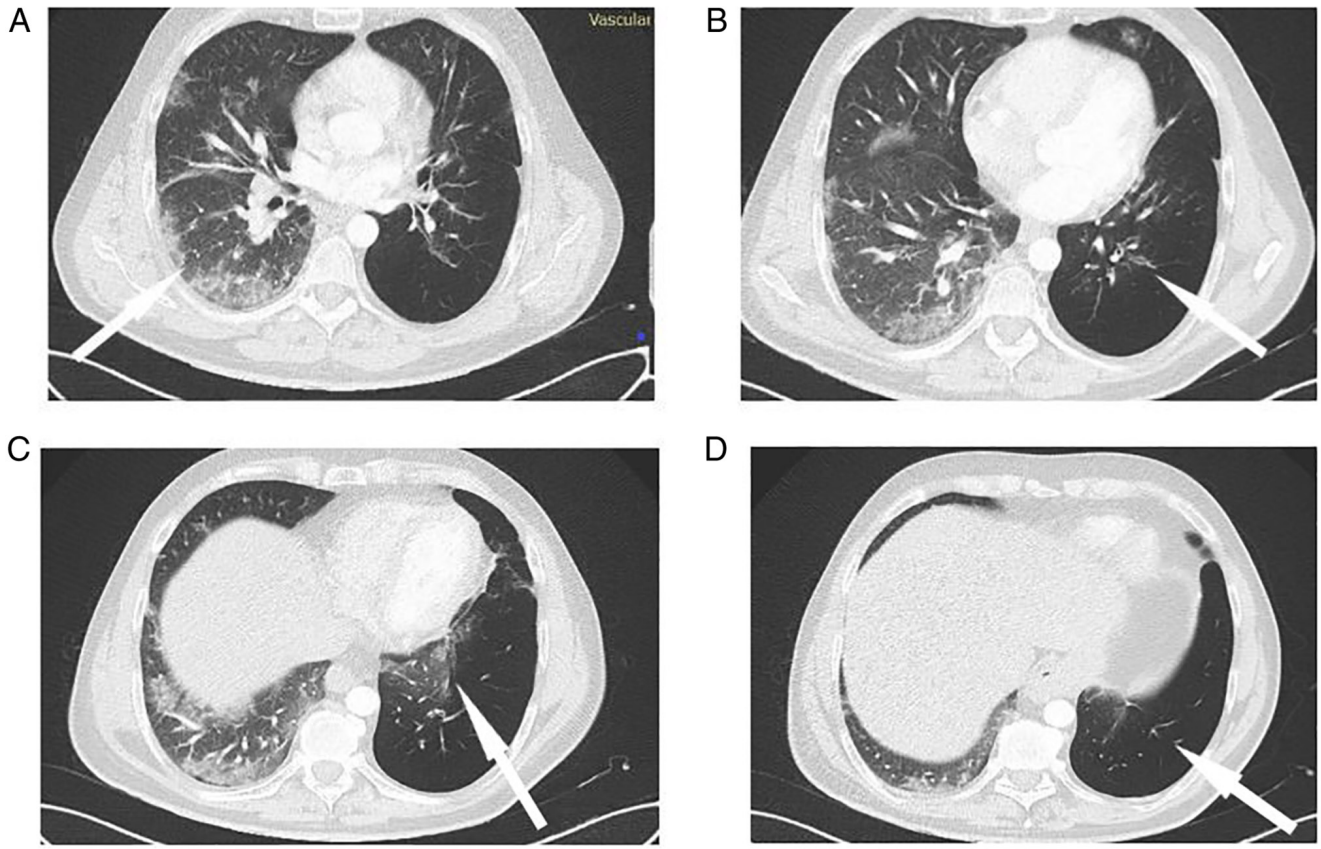

Figure 2. Chest computed tomography scan. (A) Arrow indicates infiltrates in the right lung. (B) Arrow indicates bronchiectasis in the left lung. (C) Arrow indicates infiltrates in the left lung. (D) Arrow indicates hypelucency of the left lung.

2 years had a higher chance of ICU admission or mortality, even accounting for vaccination status (16). However, there are currently no data available indicating the effects of COVID-19 in patients with Swyer-James-MacLeod syndrome, at least to the best of our knowledge.

As regards bronchiectasis, although the findings of studies on COVID-19 and bronchiectasis are conflicting (17-20), 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 even be more likely to require admission to an intensive care unit or to succumb to the disease (17-21). In the present study, the patient suffered from a syndrome characterized by an obstructive lung pattern and bronchiectasis, and required hospitalization and oxygen therapy; fortunately, he had a favorable outcome.

To the best of our knowledge, there are currently no data available on the use of bronchodilators in patients with Swyer-James-MacLeod syndrome and COVID-19-associated pneumonia. Patients with few or no bronchiectasis have minor symptoms or are asymptomatic and may remain undiagnosed until adulthood. Adult patients with this syndrome are frequently diagnosed following a chest X-ray for other reasons (6). Furthermore, there is currently no evidence that agents used for the treatment of COPD or its comorbidities adversely affect the prognosis of patients with COVID-19. Patients with COPD are strongly advised to continue their usual treatments, avoiding the use of nebulizers, particularly if they are not accompanied by a filter in the expiratory branch (5). Based on this fact, the patient described herein continued to receive his usual medication for this obstructive syndrome during his hospital stay.

Another interesting point is that the patient had a favorable outcome despite the fact that he was unvaccinated against COVID-19. It is well established that this is an additional risk factor for hospitalization and poor outcomes (22). Although there are no data suggesting that the efficacy of the vaccine in patients with chronic lung diseases, including Swyer-James-MacLeod syndrome, differs from that of the general population (5), it has been reported that airway immune responses to COVID-19 vaccination are the same in patients with COPD and healthy subjects (23).

In conclusion, the present study describes an interesting case of COVID-19-associated pneumonia in a patient with Swyer-James-MacLeod 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 data available on the effects of COVID-19 in these patients, their optimal management, or the role of COVID-19 vaccine in their clinical course.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' contributions

AB and VEG conceptualized the study. AB, VEG, PA, AG, PP and KT advised on patient care and medical treatment, and wrote and prepared the draft of the manuscript. PS, NT, GF, SC and DAS analyzed the data and provided critical revisions. AB and VEG confirm the authenticity of all the data. All authors contributed to manuscript revision and have read and approved the final version of the manuscript.

Ethics approval and consent to participate

Written informed was obtained from the patient described herein. A copy of the written consent is available for review by the editorial office of this journal on request.

Patient consent for publication

Written informed was obtained from the patient for publication of his data. A copy of the written consent is available for review by the editorial office of this journal on request.

Competing interests

The authors declare that they have no competing interests.

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