

C A S E R E P O R T

Point-of-care ultrasound for the diagnosis of Swyer-James-MacLeod syndrome in pediatric emergency setting: A case report

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Abstract. Swyer-James-MacLeod syndrome is a rare emphysematous disease characterized by decreased pulmonary vascularity and hyperinflation that are secondary to repeated childhood respiratory infections. We present a case of a 2-years old boy admitted to our pediatric emergency ward for dyspnea and fever with clinical and radiological suspicion of bronchopneumonia. The patient was monitored by using lung point-of-care ultrasound. These lung evaluations showed an increased representation of pulmonary A-lines and absence of B-lines as from hyperinflation. Although the clinical conditions improved, this unconventional imaging, not compatible with pneumonia, triggered us to perform further examinations. Thus, a chest CT scan was executed showing a reduction in parenchymal density of the left lung and hence definitely diagnosing Swyer-James-MacLeod syndrome. This report shows how the smart and skilled use of lung point-of-care ultrasound is able to detect potential “red flags” that could suggest the execution of further examinations, avoiding diagnostic delays and thus changing the patient's clinical history even in rare findings such as Swyer-James-MacLeod syndrome. (www.actabiomedica.it)

Key words: POCUS, children, ultrasound, lung, emphysema

Introduction

Swyer-James-MacLeod syndrome (SJMS) is an uncommon, emphysematous disease characterized by decreased pulmonary vascularity and hyperinflation that are secondary to repeated childhood respiratory infections and hyperactive airway disease (1). It may be limited to one lobe or one lung and it is mostly diagnosed in childhood (1). The main clinical features of this syndrome include: dyspnea, hemoptysis, reactive

upper airway disease and recurrent pulmonary infections. The etiology of this syndrome is various, as well as its long-term effects (2). Infections with adenovirus, measles, Bordetella pertussis, tuberculosis, mycoplasma pneumoniae, respiratory syncytial virus, and influenza A have been involved (3). Radiographic or computed tomography imaging is required in such patients for diagnosis. We present a case of SJMS in which the diagnostic suspicion was raised by the use of point-of-care ultrasound (POCUS) rather than chest x-ray.

Case Report

A 2-year-old boy presented to our Pediatric Emergency Department (PED) with dyspnea and fever of more than 24 hours' duration with suspicion of resistant bronchopneumonia. Twenty-five days before, the patient was diagnosed with pneumonia by his family pediatrician and was treated with dual antibiotic therapy with amoxicillin-clavulanate and clarithromycin. A chest x-ray showed an area of localized parenchymal consolidation in the left basal lung. Clinical recovery occurred after 5 days of therapy. The day before admission, the clinical scenario shifted when the patient started experiencing coughing and chest tightness. His clinical condition didn't improve even though a home-based salbutamol therapy was administered. Thus, he was conducted to our PED. Medical history was positive for episodes of wheezing. No allergies were reported. There was no history of travel or sick contacts. Immunizations were up to date. At the PED, his temperature was 38 °C; pulse, 140 beats per minute; respiratory rate, 50 breaths per minute; with arterial oxygen saturation of 94% on room air. Physical examination revealed signs of moderate respiratory distress. Nasal flaring with subcostal and intercostal retractions was noted. Upon auscultation of the lungs, poor air entry with bilateral expiratory wheezing was noted. Blood tests revealed an elevated white blood cell count and high levels of inflammatory markers, but no other relevant findings. A chest x-ray confirmed the previously diagnosed consolidation in the left basal lung. Therefore, the patient was admitted to our pediatric emergency ward with a diagnosis of left basal lung pneumonia. The patient didn't need oxygen or mechanical ventilation. A nasopharyngeal swab for respiratory viruses was performed, resulting in a positive test for rhinovirus. During hospitalization, the patient was treated with inhaled salbutamol, systemic steroids, and amoxicillin-clavulanate. Timely monitoring by using lung POCUS was applied. These lung evaluations showed an increased representation of pulmonary A-lines in the left posterior basal field and the absence of vertical artifacts and B-lines as from hyperinflation (Figure 1).

Even though the patient's clinical condition progressively improved, this unconventional imaging, not

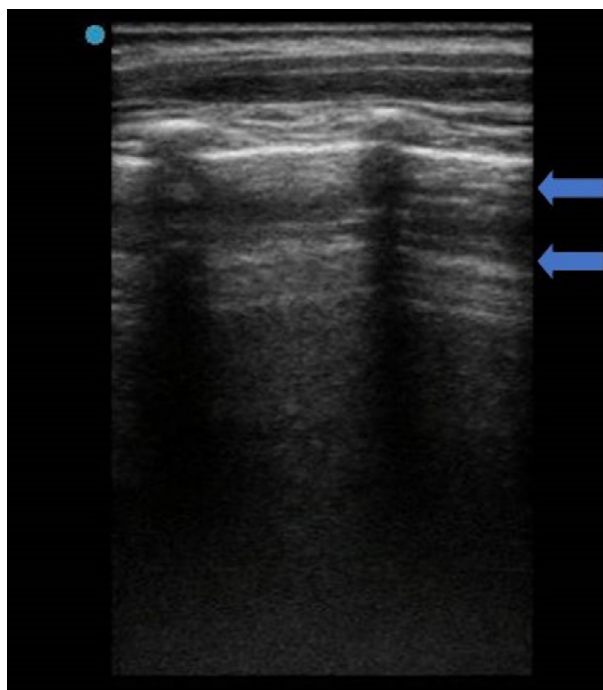


Figure 1. The ultrasound images were characterized by a pronounced pulmonary “A-line pattern” (arrows) with a total lack of vertical artifacts and B-lines.

compatible with viral superinfected pneumonia, triggered us to perform further examinations to confirm the diagnosis. So, a chest computed tomography (CT) scan was executed showing a reduction in parenchymal density of the left lung and a reduction in the caliber of the left pulmonary vessels as well (Figure 2).

This radiological picture was suggestive for sequelae of distal airways inflammation. There was also a moderate compensative overexpansion of the right lung. A diagnosis of Swyer-James-MacLeod syndrome was made. The patient was discharged in good clinical condition with scheduled follow-up care.

Discussion

Swyer-James-MacLeod syndrome is a rare finding of one-sided functional hypoplasia of pulmonary vessels and emphysema, with or without associated bronchiectasis (4). The syndrome was first described simultaneously in the 1950s by a respiratory physician,

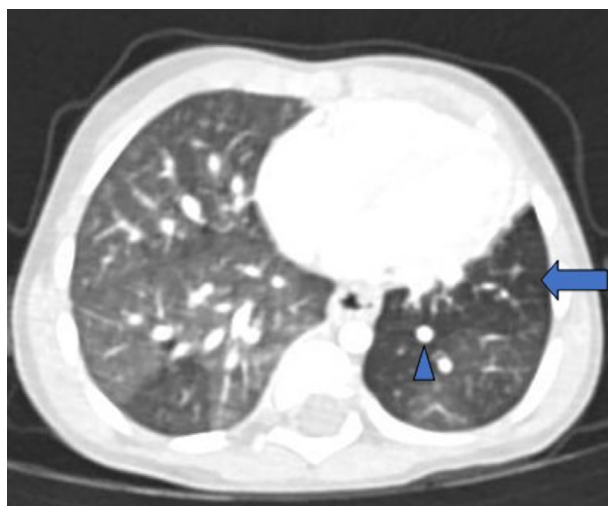


Figure 2. CT scan showing a decrease in left lung parenchymal density (arrow) and a reduction in caliber of left pulmonary vessels (triangle).

William Mathieson MacLeod in England (1954), by a physician, Paul Robert Swyer, and a radiologist, George James, in Canada (1953) (5). It is considered to be an acquired disorder secondary to viral bronchiolitis or pneumonia in childhood (6). The diagnostic criteria for this syndrome include one of the following three: a hyperlucent/hyperinflated lung on x-ray/chest CT scan, a diffusely decreased one-sided perfusion on chest CT scan, or a unilateral loss of perfusion on technetium Tc 99m lung scan (2). Usually, the first-line examination is a posteroanterior chest radiograph, which reveals reduced bronchovascular markings, a small hilar shadow, and hyperlucency of the involved lung segment or lobe (7). A chest CT scan is the subsequent investigation used to assess the extent and distribution of the disease (8). There are several differential diagnoses that should be mentioned when evaluating any patient with unilateral pulmonary hyperlucency including pneumothorax, asymmetric emphysema, congenital lobar emphysema, and pulmonary artery hypoplasia (2). Other differential diagnoses involve gastrointestinal herniation, bronchial compression, mastectomy and mediastinal fibrosis (9). Poland syndrome is another cause of a unilateral hyperlucent hemithorax, which is due to congenital one-sided lack of the pectoralis major and minor muscles, hypoplasia

of the breast and nipple and scarcity of subcutaneous tissue (10). Our report describes a diagnosis of SJMS in a pediatric emergency setting with the important contribution of POCUS. This report underlines that it is crucial to investigate all the possible causes of a certain clinical pattern, even the less frequent to ensure appropriate treatment. Since x-ray image analysis was not resolute, the diagnostic suspicion of SJMS was assumed upon the smart assessment of ultrasound pictures. POCUS is an extension of clinical examination in respiratory and emergency departments worldwide and holds high diagnostic performances for pleural and pulmonary diseases (11-12). POCUS offers the pediatrician a suitable tool to answer specific queries in clinical practice. Since the emergency setting requires a prompt diagnostic assessment (13), management of pain (14) and start of a therapeutic path, POCUS allows to narrow the differential diagnosis list and improve the security of routine procedures (15). It has replaced, in some ways, the classical stethoscope. It is performed and interpreted by a clinician in order to answer a focused question or achieve a specific procedural goal (16). Ultrasound is very suitable for pediatric patients because it constitutes an alternative approach to radiation exposure (17-18). On a lung ultrasound longitudinal scan, the pleural line is seen as a horizontal hyperechoic line between two nearby ribs that shows a posterior shadowing: this is the so-called “bat-wing” sign (19). A-lines are artefacts produced by the great difference in acoustic impedance at the interface between pleura and lung. They are hyperechoic horizontal lines, deeper than pleural lines, parallel and equidistant from one another: their presence generally excludes lung pathologies in the examined area in terms of effusion or consolidation (18). B-lines originate perpendicularly from the pleural surface in isolation and may be observed in a normal aerated lung. Nevertheless, as their number increases, commonly above 3 per interspace, suspicion for interstitial syndrome emerges (19). The ultrasound images in the report we described were characterized by pronounced pulmonary A-lines and absence of vertical artifacts and B-lines as from hyperinflation. This peculiar ultrasound pattern has been described in literature as “hypermirror effect” in association with emphysematous pathologies in adults (Figure 3) (11).

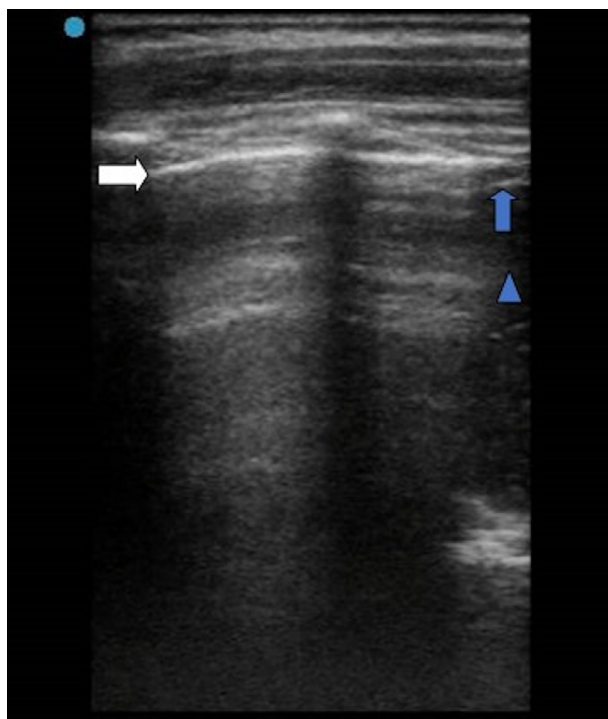


Figure 3. The “hypermirror effect” consists of an ultrasound finding regarding at least one third of a lung area characterized by a homogeneous pleural line (white arrow) and by the absolute lack of vertical artifacts. It is due to strong reverberations of ultrasounds on a very impeding interface generated by air. On the right side of the figure, the diaphragm, indicated by the blue arrow, defines the border to the abdomen, indicated by the triangle below.

As a matter of fact, in the presence of emphysema, the air content of the subpleural lung layers increases and, subsequently, the reflection coefficient at the pleural plane also boosts giving rise to more pronounced replicas and mirror effects (20). To the best of our knowledge, this is the first report describing a case of SJMS, whose diagnostic suspicion was assumed starting with the use of lung POCUS rather than the traditional chest x-ray. Although lung POCUS is widely expanding its role in air-trapping pathologies including emphysema (Figure 4), its diagnostic potential has to be fully defined with further large studies (11).

The traditional radiological methods (chest x-ray and CT scan) remain actually the gold standard to detect SJMS4 but lung POCUS may be a worthy tool to empower clinician’s capacity to move towards the appropriate diagnosis. Barriers to a widespread diffusion of POCUS among pediatricians include: a lack of a

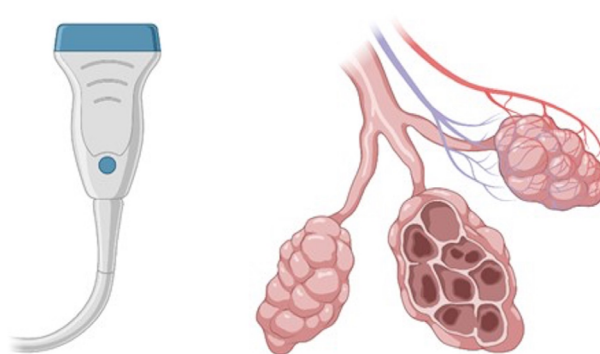


Figure 4. Lung POCUS is a valuable radiation-free tool to empower clinician’s diagnostic capacity to detect air-trapping due to enlargement of alveolar spaces such as in emphysematous pathologies (created with biorender.com).

structured recognized educational process since residency, a lack of image storage and archiving and less commonly a lack of ultrasound equipment (21-22).

In the majority of patients with Swyer-James-MacLeod syndrome, treatment consists of chest physiotherapy, low-dose inhaled corticosteroids, and inhaled bronchodilators even though, in some patients with relapsing infections, lobar or pulmonary surgery are considered (2).

Conclusion

This report shows how the smart and skilled use of lung POCUS is able to detect potential “red flags” that could suggest the execution of further examinations, avoiding diagnostic delays and thus changing the patient’s clinical history even in rare findings such as SJMS.

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Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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