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WHEN COPD ISN'T COPD: A FOURTEEN-YEAR CASE STUDY OF ADULT-ONSET SWYER-JAMES-MACLEOD SYNDROME

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ABSTRACT

Swyer-James-MacLeod (SJML) syndrome, also known as unilateral hyperlucent lung syndrome, is a rare post-infectious pulmonary disorder characterized by emphysematous changes, unilateral pulmonary artery hypoplasia, and regional hypoperfusion [1, 2]. The hallmark radiographic feature - often referred to as the "hyperlucent lung" or "clear lung sign" - reflects air trapping with decreased vascular markings and serves as the signature diagnostic clue of this condition [3, 6]. SJML syndrome typically develops following childhood bronchiolitis or pneumonitis but may remain unrecognized until adulthood due to its nonspecific respiratory presentation [4, 7].

We report the case of a 41-year-old woman who has been under observation and treatment for the past 14 years. Despite chronic, non-productive cough and persistent unilateral pulmonary abnormalities, she was not initially diagnosed with SJML syndrome due to the absence of standardized diagnostic criteria. Radiological assessment, including high-resolution computed tomography (HRCT) and CT angiography, revealed a hyperlucent and volume-reduced left lung with bronchiectasic changes and pulmonary arterial hypoplasia, findings consistent with described syndrome. Pulmonary function tests demonstrated moderate airflow obstruction, while the patient's prolonged childhood exposure to tobacco smoke may have contributed to airway remodeling.

This long-term follow-up highlights the diagnostic importance of recognizing unilateral hyperlucency as the signature sign of SJML syndrome and underscores the need for multidisciplinary monitoring to optimize long-term respiratory outcomes [5–7].

KEYWORDS

Rare Pulmonary Disease, Hyperlucent Lung, Swyer-James-Macleod Syndrome, Adult Diagnosis

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Introduction

Swyer-James-MacLeod syndrome (SJMS) is a rare pulmonary condition characterized by unilateral (or occasionally lobar) hyperlucency of lung parenchyma, reduced vascularity, and air trapping, typically resulting from post-infectious obliterative bronchiolitis in early childhood [1, 2]. Although initially described in paediatric populations, the condition may remain undiagnosed until adulthood, where it may present as an incidental radiological finding or with nonspecific respiratory [3, 4].

Pathophysiologically, the syndrome is believed to result from a childhood insult (often viral or bacterial bronchiolitis or pneumonitis) that leads to inflammation and obliteration of small airways, followed by failure of normal alveolar development and distal vascular bed growth [5]. The resultant regional hypoperfusion and air-trapping manifest radiographically as a hyperlucent lung on chest radiograph and mosaic perfusion on CT imaging [6]. Clinically, patients may remain asymptomatic or eventually develop exertional dyspnoea, recurrent chest infections, haemoptysis, or complications such as bronchiectasis and pulmonary hypertension [3, 7].

Because the anatomical and functional complications may mimic more common conditions such as chronic obstructive pulmonary disease (COPD), asthma, or even pulmonary embolism, the diagnosis requires a high index of suspicion and radiologic-functional correlation [7]. In adults, recognition is often delayed and misdiagnosis may lead to inappropriate interventions [3, 4]. Moreover, the true incidence of SJMS remains uncertain; a radiographic study found a prevalence of approximately 0.01 % in more than 17 000 chest radiographs [6].

Management is largely conservative-centred on airway clearance, immunisation, and symptomatic bronchodilator or inhaled corticosteroid therapy in selected cases - while surgical resection is reserved for symptomatic patients with recurrent infections or severe localized disease [5, 4]. Despite its rarity, SJMS

should be considered in the differential diagnosis of unilateral hyperlucent lung, unexplained airflow obstruction, or recurrent infections, particularly when conventional etiologies are lacking.

Given the paucity of large series and the predominance of case reports and small retrospective analyses, further studies are warranted to elucidate long-term outcomes, optimal management strategies, and the interplay of co-morbidities in adult-onset SJMS [3, 7]. The present case report illustrates such an atypical presentation, emphasising the need for comprehensive diagnostic work-up and tailored follow-up in adult patients with isolated unilateral pulmonary abnormalities.

Case report

A 27-year-old woman of Eastern European descent presented to the pulmonology outpatient clinic with a chronic, non-productive cough persisting for several months. The patient was referred by her primary care physician following the detection of persistent fine inspiratory crackles on chest auscultation. Aside from a short course of antibiotics prescribed during her initial evaluation, she reported no history of chronic medication use and denied taking any over-the-counter or herbal remedies.

At presentation, the patient's vital signs were within normal limits: blood pressure 134/64 mmHg, heart rate 82 beats per minute, body temperature 36°C, and peripheral oxygen saturation (SpO_2) 97% on room air. She weighed 88 kilograms and measured 158 centimeters in height, corresponding to a body mass index (BMI) of 35 kg/m², consistent with class II obesity. The general physical examination was otherwise unremarkable.

On respiratory examination, fine, soft crackles were heard over the left lung field, particularly at the base, while the right lung was clear to auscultation. There were no wheezes, rhonchi, or signs of acute respiratory distress. Cardiac examination revealed normal heart sounds without murmurs or gallops, and there was no peripheral edema, cyanosis, or clubbing. The remainder of the systemic examination was unremarkable.

A chest radiograph (Figure 1) revealed an abnormally hyperlucent upper lobe of the left lung with subtle pleuroparenchymal adhesions at the left lung base and a rounded cardiac silhouette, though without radiographic evidence of cardiomegaly. The mediastinum appeared central, and there was no pleural effusion, mass lesion, or pneumothorax. Routine laboratory tests, including a complete blood count, C-reactive protein, and basic metabolic panel, were within normal limits.

The patient's past medical history was undisinguished, with no prior diagnoses of asthma, tuberculosis, or any chronic pulmonary disease. She denied any history of smoking or use of recreational substances. However, she reported long-term passive exposure to tobacco smoke from early childhood, as her mother was a heavy smoker who consumed multiple cigarettes daily in the patient's presence.

Her family history was notable for respiratory tuberculosis in her mother, confirmed in the past by bacteriological and histopathological evidence, and successfully treated. Her father had a history of arterial hypertension and obesity, but there was no known familial incidence of congenital pulmonary disorders, cystic fibrosis, or primary ciliary dyskinesia.

In view of the persistent auscultatory findings, radiologic abnormalities, and absence of smoking history, further evaluation was indicated. The patient underwent spirometry with bronchodilator reversibility testing and high-resolution computed tomography (HRCT) of the thorax.

The spirometric assessment demonstrated moderate airflow obstruction, characterized by a reduced forced expiratory volume in one second (FEV_1) and a decreased FEV_1/FVC ratio, with partial reversibility after inhaled bronchodilator administration. These results were consistent with moderate obstructive ventilatory impairment, corresponding to Stage III Chronic Obstructive Pulmonary Disease (COPD) according to the GOLD classification, although the diagnosis was considered atypical due to her young age and non-smoking status.

The HRCT scan (**Figures 2-6**) of the chest revealed hyperinflation and increased aeration of the left lung parenchyma, accompanied by reduction in overall left lung volume, areas of post-inflammatory scarring, bronchial wall thickening, and localized bronchiectasis. The imaging pattern raised suspicion for Swyer-James-MacLeod syndrome (also referred to as unilateral hyperlucent lung syndrome), likely of post-infectious etiology, possibly secondary to severe bronchiolitis in early life.

Given the atypical findings and the need to exclude vascular malformations or congenital hypoplasia, an angio-CT of the chest and genetic testing for alpha-1 antitrypsin (A1AT) deficiency were recommended.

While awaiting results, the patient was commenced on maintenance inhalation therapy, consisting of a long-acting bronchodilator (LABA), an inhaled corticosteroid (ICS), and a mucolytic agent for daily use. She received detailed counselling regarding proper inhaler technique, weight management, avoidance of respiratory irritants, and smoking cessation in her household environment to reduce second-hand smoke exposure.

At her follow-up visit, several months later, the patient reported a marked improvement in her overall condition, including reduced cough frequency and improved exercise tolerance. Repeat spirometry showed increased FEV_1 and FVC values, though the pattern of moderate obstruction persisted. On physical examination, fine crackles remained audible at the base of the left lung but were less pronounced. Oxygen saturation remained stable at rest.

The genetic analysis subsequently identified a heterozygous "Z" mutation in the SERPINA1 gene, corresponding to the PiMZ genotype, indicating a partial deficiency of alpha-1 antitrypsin. Although heterozygous carriers usually maintain near-normal enzyme activity, this genotype was recognized as an additional risk factor for early-onset pulmonary pathology, especially in the context of previous environmental smoke exposure and structural lung abnormalities.

The CT pulmonary angiography (**Figures 7-11**) confirmed hypoplasia of the left pulmonary artery and its segmental branches, resulting in decreased perfusion of the affected lung and supporting the diagnosis of unilateral hyperlucent lung. The contralateral (right) lung appeared structurally normal, with compensatory hyperexpansion.

At this point, approximately eighteen months had elapsed since the patient's initial presentation. Given her clinical improvement but persistence of moderate obstruction, the treatment regimen was modified: a short-acting bronchodilator (SABA) was added for as-needed symptom relief, and the daily mucolytic was discontinued due to lack of ongoing benefit. The patient was enrolled in a structured follow-up program involving regular spirometry and the six-minute walk test (6MWT) to assess functional capacity, oxygen desaturation during exertion, and the degree of activity-induced dyspnea.

Over subsequent years, the patient remained adherent to her prescribed inhaled therapy, demonstrating excellent compliance and significant reduction in disease exacerbations and respiratory infections, which had previously occurred several times per year. She reported progressive improvement in her quality of life, with only occasional mild cough and exertional breathlessness.

Serial spirometric assessments indicated a slow but steady decline in FEV_1, consistent with gradual progression of chronic airflow limitation typical of obstructive airway diseases. During exertion, transient oxygen desaturation episodes were noted, though resting oxygenation remained within normal limits. On physical examination, fine, isolated crackles persisted at the base of the left lung field, consistent with residual post-inflammatory parenchymal changes.

Repeat chest radiographs obtained over several years demonstrated a stable radiologic appearance, with no progression of hyperlucency, no new areas of fibrosis, and no signs of right heart enlargement or pulmonary hypertension. The patient denied recent hospitalizations or severe exacerbations and continued to perform daily activities without major limitation.

Methodology

This report is based on a retrospective, descriptive case study documenting the clinical presentation, diagnostic evaluation, and longitudinal management of a single patient with chronic respiratory symptoms. The methodology involved systematic data collection from the patient's medical records, clinical examinations, imaging studies, and functional assessments conducted at a tertiary pulmonary care center.

Study Design and Data Collection

Clinical data were obtained from routine outpatient visits over an 18-month diagnostic and treatment period, followed by long-term observation. Information included the patient's demographic characteristics, medical and family history, physical findings, vital parameters, and anthropometric measurements. Diagnostic data were derived from radiologic imaging (chest X-ray, HRCT, and CT pulmonary angiography), pulmonary function tests (spirometry with bronchodilator reversibility assessment), and genetic analysis for A1AT deficiency. Additional evaluations included serial spirometric follow-up and the six-minute walk test (6MWT) to assess exertional oxygen desaturation.

Ethical Considerations

Written informed consent was obtained from the patient for the use of anonymized clinical data and imaging materials.

Results

Over a 14-year observation period (2011-2025), the patient's clinical and functional parameters revealed a chronic, slowly progressive course consistent with Swyer-James-MacLeod syndrome.

Baseline Imaging (2011-2012):

The initial chest X-ray (**Figure 1**) and HRCT (**Figures 2-6**) revealed a hyperlucent left upper lobe, with markedly reduced vascular markings and mild bronchial wall thickening. The right lung demonstrated normal architecture. The characteristic "hyperlucent lobe" sign, reflecting hypoperfusion and air trapping, was considered the radiological signature of SJML syndrome. No parenchymal consolidations or interstitial fibrosis were present.

Angio-CT (**Figures 7-11**) of the chest demonstrated marked hypoplasia of the left pulmonary artery with reduced caliber and peripheral tapering of its segmental branches. There was asymmetric pulmonary perfusion, with a relative lack of vascular markings and decreased attenuation of the left lung parenchyma, consistent with regional hypoperfusion. The right pulmonary artery and its branches appeared normal in diameter and distribution. No intraluminal filling defects suggestive of thromboembolic disease were identified. These findings correlated with the previously observed hyperlucent left lung on X-ray and HRCT supporting the diagnosis.

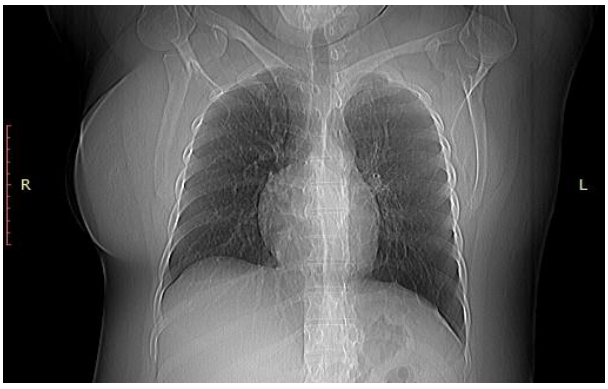

Fig. 1. *Chest X-ray (February 2011): Left-sided hyperlucency with reduced vascular markings and preserved right lung structure.*

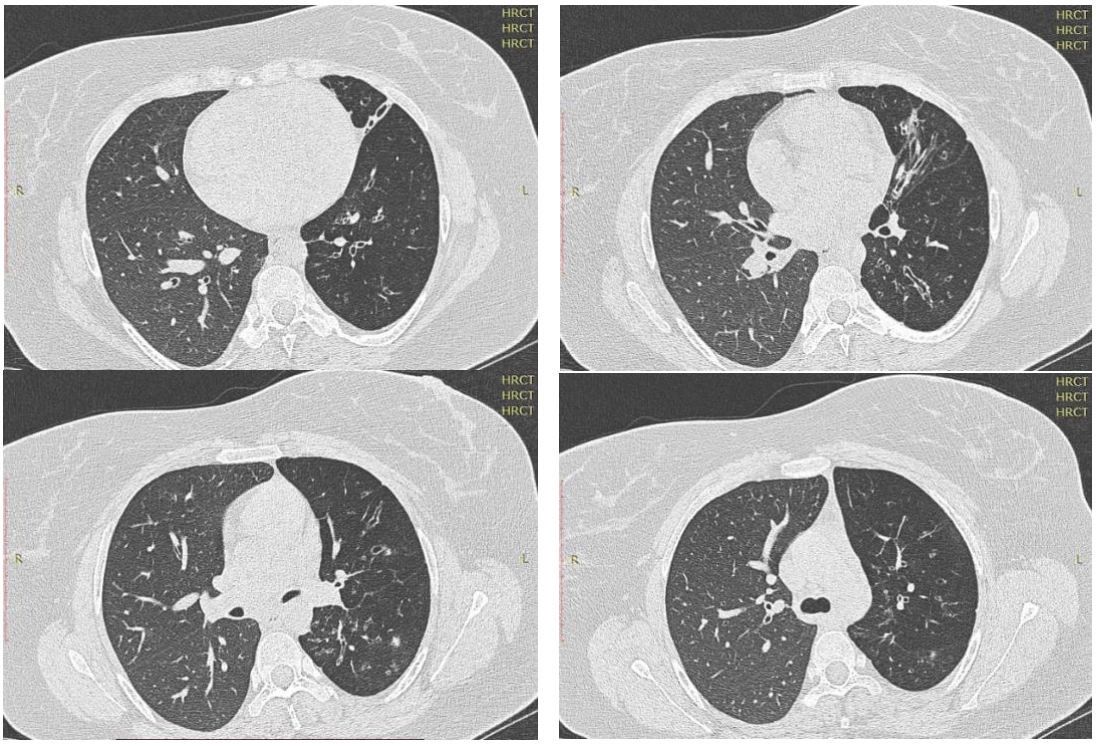

Fig. 2-6. *HRCT scans (2011): Sequential axial slices demonstrating decreased vascular density and air trapping in the left upper lobe, confirming unilateral hypoperfusion and bronchial wall thickening.*

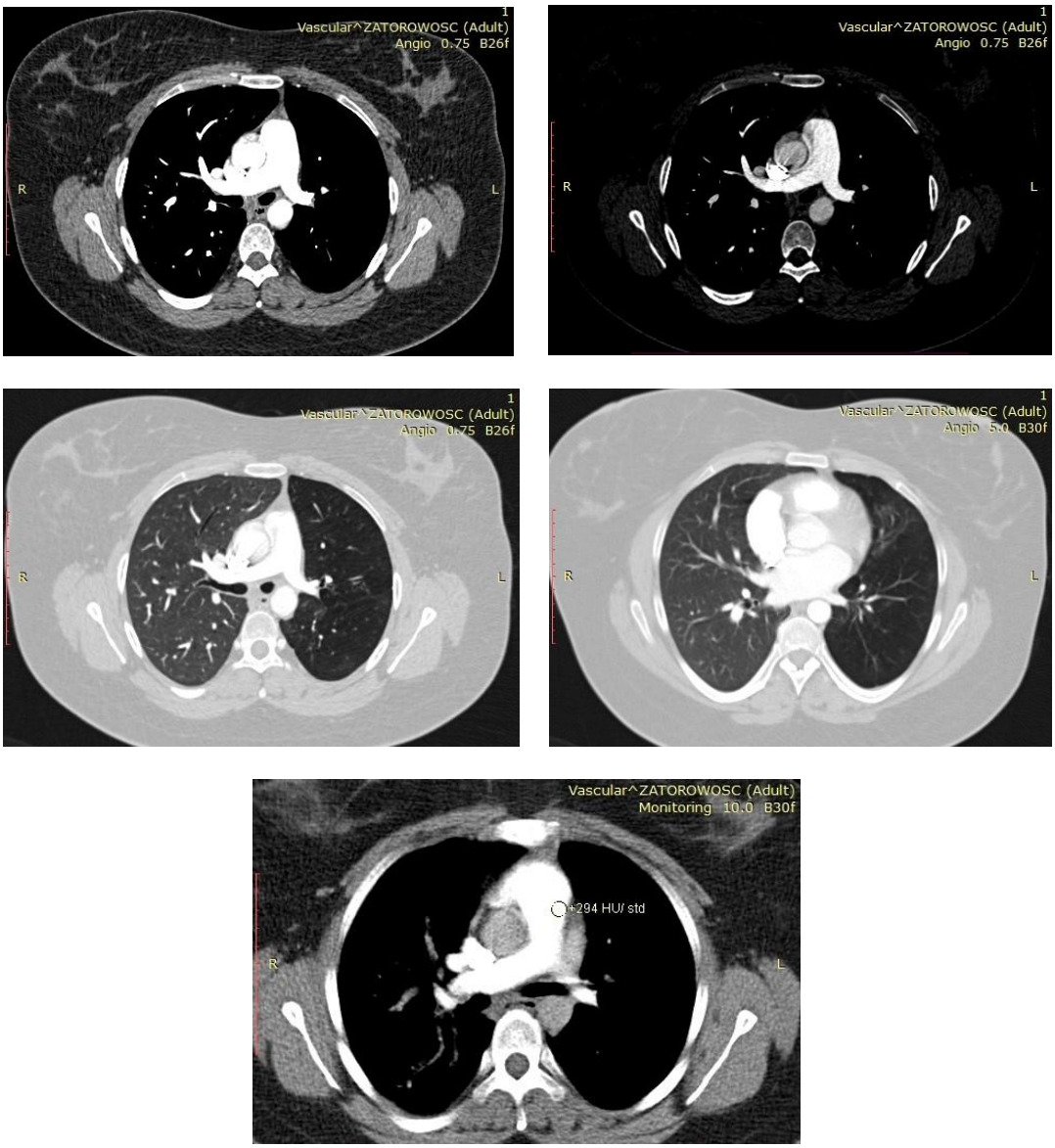

Fig. 7-11. *Angio-CT (2012) demonstrating hypoplasia of the left pulmonary artery and its peripheral branches, with markedly reduced perfusion and vascular attenuation within the left lung field. The right pulmonary artery and vascular structures appear normal.*

Functional Assessment (2011-2015):

Initial spirometry demonstrated moderate obstruction (FEV_1 improvement of 200 mL post-bronchodilator; FVC + 700 mL). Over the following four years, spirometric parameters remained relatively stable with mild fluctuations. Six-minute walk tests showed good exercise tolerance, though transient exercise-induced desaturation (4-6 percantage point) was noted.

Mid-term Follow-up (2016-2020):

During this period, obstruction worsened to a severe level but remained partially reversible in some tests. Despite mild-to-moderate dyspnea and occasional tachycardia, the patient preserved functional mobility. Imaging studies in 2017 and 2020 confirmed persistence of the hyperlucent left lung, consistent with stable unilateral air trapping and vascular hypoplasia, without new pathological lesions.

Recent Follow-up (2021-2025):

In later years, pulmonary parameters stabilized at a moderate-to-severe obstructive level, with no reversibility on bronchodilator testing. Repeated six-minute walk tests demonstrated stable distances, mild desaturation (6-9 percentage point), and absence of significant dyspnea. Radiographs in 2024 continued to

show a markedly hypovascular left upper lobe with preserved right lung perfusion, confirming a non-progressive pattern.

Throughout the observation period, the patient's BMI increased from 34.0 kg/m² to 41.7 kg/m², and oxygen saturation remained stable (94-97%). Blood pressure and pulse values were within normal limits during all follow-ups. Despite persistent obstructive patterns, the patient's overall clinical condition and exercise capacity remained satisfactory.

Year	BMI (kg/m²)	Spirometry Result	FEV_1/FVC Trend	Bronchodilator Reversibility	6MWT	Desaturation (percantage point)	Dyspnea / Tachycardia
2011	34.0	Moderate obstruction	FEV_1 ↑ 200mL, FVC ↑ 700mL	(+)	Baseline good	–	None
2012	38.0	Moderate obstruction, stable	Stable	(–)	Reduced distance	–6 pp.	Mild dyspnea
2013	40.0	Moderate obstruction	FEV_1 ↑ 50mL	(–)	Distance doubled	–9 pp.	Mild
2014	39.5	Moderate obstruction + mild restriction	Stable	(–)	Normal	None	None
2015	38.9	Moderate obstruction + moderate restriction	Stable	(–)	Good	–5 pp.	None
2016	41.7	Severe obstruction	FEV_1 ↓ 200mL, FVC ↓ 400mL	(–)	Good	–4 pp.	Moderate / (+)
2017	40.5	Severe obstruction (partly reversible)	Stable	(–)	Reduced by 60 m	–6 pp.	Mild / (+)
2018	38.8	Moderate obstruction	FEV_1 ↑ 70mL, FVC ↑ 100mL	(–)	Slightly reduced	–5 pp.	None / (+)
2019	40.5	Severe obstruction	FEV_1 ↑ 100mL, FVC ↑ 100mL	(–)	Very good	–6 pp.	Mild / (+)
2021	39.9	Moderate obstruction + low FVC	Stable	(–)	Good	–7 pp.	None / (+)
2022	41.7	Severe obstruction, reversible to moderate	FEV_1 ↓ 100mL, FVC ↓ 100mL	(+)	Slightly shortened	–7 pp.	None / (+)
2023	40.9	Severe obstruction, stable	Stable	(–)	Good	–7 pp.	None / (+)
2024	42.1	Severe obstruction, non-reversible	FEV_1 ↓ 100mL	(–)	Slightly shortened	–6 pp.	None / (+)
2025	41.7	Severe obstruction, stable	Stable	(–)	Distance ↑ 100m	–9 pp.	None / (–)

Discussion

This case of adult-onset Swyer-James-MacLeod syndrome underscores several clinically and diagnostically important features. First, the persistent hyperlucent left upper lobe - the so-called "clear lung sign" - emerged as the signature radiologic marker in this patient, reinforcing its diagnostic value in differentiating SJML syndrome from more common obstructive lung diseases. The imaging findings in this patient (**Figures 1-11**) demonstrate unilateral lung hypoperfusion and air trapping consistent with published descriptions of SJML pathophysiology (post-infectious obliterative bronchiolitis, vascular hypoplasia) [1,2,3,6]. The absence of progressive radiologic deterioration over 14 years suggests a relatively stable structural phenotype in this adult-onset presentation, though functional decline occurred gradually.

Second, the long-term spirometric and functional data highlight the behavioral and physiological trajectory of SJML syndrome in adulthood: moderate to severe airflow obstruction, variable reversibility, and exercise-induced desaturation, yet preserved exercise capacity. These findings resemble patterns reported in other adult SJML cases, although many of those lack such extended follow-up [4,7]. The presence of childhood exposure to second-hand tobacco smoke may have acted as an additional risk factor, perhaps accelerating small airway injury in conjunction with the congenital vascular abnormalities typical of SJML. This emphasizes the importance of thorough environmental history in adult patients with unexplained obstructive defects.

Third, from a management standpoint, the case reinforces the need for multidisciplinary monitoring – combining imaging, functional testing, and symptomatic evaluation over time. The stable imaging findings, moderate functional decline, and absence of frequent exacerbations suggest that early recognition and tailored management may modify the natural history of SJML. In particular, the clear lung sign may prompt early advanced imaging (e.g., HRCT, CT angiography) and targeted follow-up rather than assuming standard COPD etiology.

From a broader perspective, this case contributes to the evolving understanding of SJML in adult populations. Given the rarity of comprehensive longitudinal follow-up in SJML, this report addresses a gap in current literature. It invites further research into the determinants of functional decline, the role of additional risk modifiers (such as genetic predisposition or environmental exposures), and the optimal monitoring regimen for adult patients diagnosed with hyperlucent lung syndrome. It also raises a cautionary note: adult patients presenting with unilateral hyperlucency or atypical obstructive patterns may be misdiagnosed with COPD or asthma, underscoring the need for heightened diagnostic suspicion.

Finally, the implications of this case for practice extend beyond pulmonology: the use of signature radiologic markers like the clear lung sign, combined with long-term functional monitoring, may serve as a model for other rare pulmonary syndromes in adult medicine. It highlights how technological integration (advanced imaging, serial spirometry, exercise testing) aligns with the journal's focus on innovative applied diagnostics in social and clinical contexts.

Conclusions

SJML syndrome should be considered in the differential diagnosis of adult patients presenting with obstructive pulmonary changes in the absence of a clear etiologic factor. Despite advances in thoracic imaging and pulmonary diagnostics, there remains a limited number of reported cases and insufficient clinical data regarding this rare condition, which often leads to delayed recognition and suboptimal management.

Clinicians should remain vigilant and include SJML in the diagnostic workup, even among older patients, particularly when the clinical and functional findings suggest chronic obstructive pulmonary disease (COPD) but the overall presentation appears atypical. In such scenarios, hyperlucent lung syndrome may represent an underdiagnosed alternative, reminding us that not all cases of airway obstruction in adults necessarily correspond to classic COPD.

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