

Pulmonary MR Angiography in Swyer-James Syndrome



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ABSTRACT

Aim: The aim of this study was to investigate the pulmonary magnetic resonance angiography (MRA) imaging findings in Swyer - James syndrome (SJS) as a alternative imaging modalities.

Methods: Thirteen patients had posteroanterior inspiratory/expiratory chest X-ray films and pulmonary MRA with SJS (6 males, 7 females) were retrospectively studied. Gradient-echo pulmonary MRA was performed in all patients with a 1.5 T MR unit. MRA imaging was performed in a single breath-hold during the injection of contrast media. Image analysis of the patients was performed. The results of pulmonary artery diameters and pulmonary peripheral vasculature were compared between normal sides versus the affected sides. Statistical analysis was performed with the Student *t* test.

Results: The smaller pulmonary artery on the affected side with poor peripheral vasculature was observed with gadolinium-enhanced gradient-echo MRA in all patients. There was a significant difference between the diameters of the pulmonary arteries of the normal versus the disease sides ($p < 0.0005$).

Conclusion: MRA successfully depicted the small pulmonary artery with diminished peripheral vasculature in patients with SJS. This study showed the pulmonary MRA imaging could be used as a alternative imaging modalities in the evaluation of patient with SJS.

Key words: Hyperlucent lung, MR, MR angiography.

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INTRODUCTION

Swyer-James syndrome (SJS) or Macleod syndrome, which is synonymous, was first described by Swyer and James 1953 (1). The following year, MacLeod (2) reported nine patients with unilateral pulmonary hyperlucency. SJS is a rare disease characterized by a hyperlucent lung or lobe, usually of normal or small size, associated with air trapping and diminutive pulmonary vascularization (3). SJS appears to be a consequence of bronchiolitis obliterans resulting from bacterial or viral (adenovirus) infection during childhood, which results in subsequent underdevelopment of the involved portion of the lung (4,5). This syndrome is usually diagnosed using chest radiography showing hyperlucency of one lung or lobe and a small hemithorax on the affected side. A lot of different imaging methods, including chest X-ray, computed tomography (CT), high resolution CT, ventilation and perfusion scan, conventional angiography or intravenous digital subtraction angiography, bronchoscopy and bronchography have been used for diagnostic evaluation in SJS (6-8). According to our knowledge, this is first study on the pulmonary magnetic resonance angiography (MRA) in SJS. The aim of this study was to show the pulmonary MRA imaging findings in SJS as a alternative imaging modalities.

MATERIALS and METHODS

Patient Population

From February 2001 to December 2007, 18 sequential patients who were diagnosed with SJS in our hospital were recruited to this retrospective study. These patients were examined by chest X-ray (all patients) and pulmonary MRA (13 patients) based on clinical assessment. However, 5 patients were excluded from the study due to they had not pulmonary MRA. Therefore, the study population consisted of 13 patients were 6 male and 7 female (age range: 17-70 years, mean: 38.46 years). All of the 13 patients had posteroanterior inspiratory/expiratory chest X-ray films and pulmonary MRA images. All patients had a history of childhood lung infection and complained from exertional dyspnea, chronic cough and expectoration. Diagnosis of SJS was based on history, clinical, chest X-ray and pulmonary MRA findings. Image analysis of the patients was performed.

MRI Study Protocol

Pulmonary MRA was performed in all patients with a MR unit (1.5 T, Siemens Magnetom Symphony Erlangen, Germany or 1.5 T, Picker Edge Cleveland, USA). A 20 ga intravenous cannula was inserted into a large ante-cubital vein. Patients were entered headfirst into the magnet. Patients were instructed to take several deep breaths before image acquisition. After scout images were obtained, a timing bolus of 2 mL of gadopentetate dimeglumine was injected via the cannula by manually to measure transit time by using a multiphase sagittal T1-weighted gradient-echo timing sequence. The scan delay time for the transit of the contrast material to the main pulmonary artery ranged from 10 to 15 seconds.

MRA was performed in a single breath-hold during the injection of contrast media. Gadopentetate dimeglumine (0.2 mmol/kg) was injected via the cannula by manually. For the contrast-enhanced MR imaging, coronal T1-weighted 3D fast multiplanar spoiled gradient-echo images were obtained with a 30°-45° flip angle, TE 2.8-3.2, TR 2-7 msec, a 35-40 cm field of view, a 144× 192 matrix, and 2.5-mm thick sections. Acquisition times were 20-25 seconds. Maximum intensity projections (MIP) images were obtained.

Image Analysis

The chest x-ray films and the MIP images of pulmonary MRA were independently and separately evaluated by two radiologists (MES, KO), and later decisions were made by consensus. Assessment was based on the following aspects: affected side, lung size, air trapping, pulmonary artery diameter, pulmonary peripheral vasculature. The diameters of the pulmonary arteries of the normal and the disease sides were measured on the MIPs images of the pulmonary MRA. The results of pulmonary artery diameters and pulmonary peripheral vasculature were compared between normal sides versus the affected sides. Statistical analysis was performed with the Student t test.

RESULTS

Table 1 summarizes the imaging techniques, chest radiographs and MRA findings of the patients, including the affected lung sides. Chest radiographs showed unilateral hyperlucency in the left lung in 10 of 13 patients and in the right lung in 3 cases. In all patients, due to marked air trapping little or no change was observed in volume between inspiration

Table 1. Chest x-ray and MR angiography findings of 12 patients with Swyer-James-MacLeod syndrome.

| Case | Age/sex | Chest radiography | | | MR angiography | |
|------|---------|-------------------|-----------|--------------|----------------|----------|
| | | Hp lung | Lung size | Air trapping | M/L/R PAD | Poor PPV |
| 1 | 31/F | L | Small | + | L- | + |
| 2 | 62/F | L | Small | + | L- | + |
| 3 | 70/F | L | Small | + | M+ L- + | |
| 4 | 44/F | L | Small | + | L- | + |
| 5 | 35/M | L | Small | + | L- | + |
| 6 | 40/F | L | Small | + | L- | + |
| 7 | 25/M | L | Small | + | L- | + |
| 8 | 40/M | R | Small | + | R- | + |
| 9 | 52/M | R | Small | + | R- | + |
| 10 | 18/M | R | Small | + | R- | + |
| 11 | 17/M | L | Small | + | L- | + |
| 12 | 31/F | L | Small | + | L- | + |
| 13 | 35/F | L | Small | + | L- | + |

Abbreviations: Hp; hyperlucent, M/L/R; Main/left/right, PAD; pulmonary artery diameter (+; increase, -; decrease), PPV; pulmonary peripheral vasculature, L; left, R; right.

(Figure 1a, 2a) and expiration (Figure 1b, 2b) in the affected lung. In all patients, the hyperlucent lung was reduced in size with decreased hilar shadow and pulmonary vascularity.

MRA showed a small pulmonary artery with a reduced number and size of branches with diminished peripheral vasculature in all patients at the affected side of the lung (Figure 1c, 2c). All patients were able to sustain adequate breath holding. MRA showed pulmonary hypoplasia in the all patients (in 10 patients on the left side and in 3 on the right side). The main pulmonary artery was found to be of normal size in

all patient except one case. The main pulmonary artery was found to be increased in size in case 3 with MRA.

MRA of pulmonary vasculature at the time of having maximal contrast enhancement showed pulmonary arteries in all patients. But, in 5 patients, the venous contamination were occurred as a imaging limitation.

The diameters of the pulmonary arteries of the normal and the diseased sides were measured on MIP images of pulmonary MRA. These measurements were demonstrated at Table 2. We founded the average diameters of the normal sides (21.19 ± 3.62 mm) and the

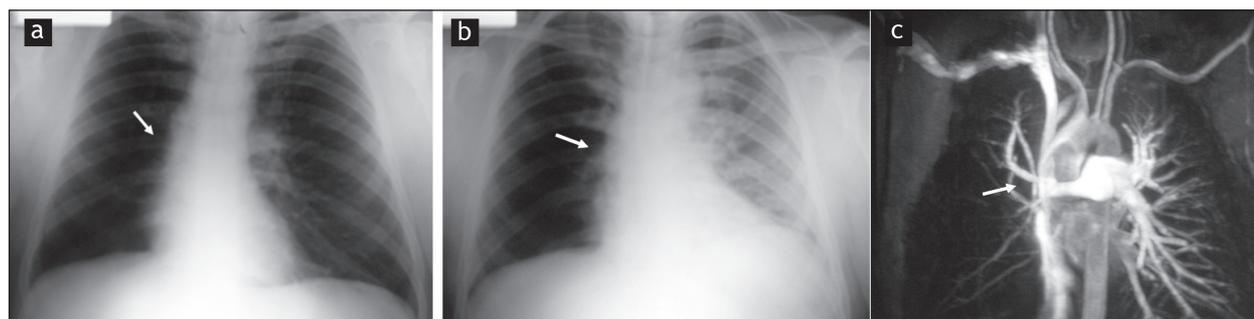


Figure 1. A 40-year-old man. Inspiratory (A) chest X-ray film reveals a small hyperlucent right lung with diminished vascular markings and small right hilum (arrow). Expiratory (B) chest X-ray film shows marked air trapping on the right lung and leftward mediastinal displacement (arrow). Pulmonary 3D MRA MIP image (C) shows a small right pulmonary artery with reduced size and number of branches with diminished peripheral vasculature in the right lung (arrow).

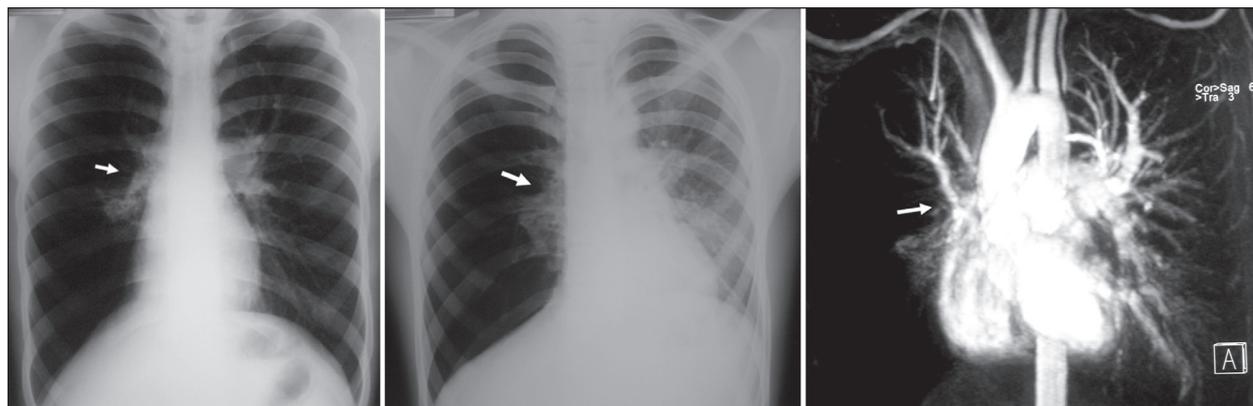


Figure 2. A 18 year old man. Inspiratory (A) and expiratory (B) chest X-ray films show a small hyperlucent right lung with decreased pulmonary markings and air trapping on the right lung and leftward mediastinal displacement (arrow). Pulmonary 3D MRA MIP image (C) reveals right pulmonary artery was small (arrow) and number of branches with diminished peripheral vasculature in the right lung with left pulmonary artery and size of branches were normal diameter.

Table 2. The measurements of diameters of pulmonary arteries at MR angiography of the patients with Swyer-James-MacLeod syndrome.

| Case | Affected side | Right pulmonary artery | Left pulmonary artery |
|------|---------------|------------------------|-----------------------|
| 1 | L | 19 mm | 8.5 mm |
| 2 | L | 31 mm | 11.5 mm |
| 3 | L | 22 mm | 17 mm |
| 4 | L | 19.5 mm | 13 mm |
| 5 | L | 18 mm | 8 mm |
| 6 | L | 21 mm | 15.5 mm |
| 7 | L | 20 mm | 8.5 mm |
| 8 | R | 10.5 mm | 24 mm |
| 9 | R | 16 mm | 24.5 mm |
| 10 | R | 7.5 mm | 21 mm |
| 11 | L | 18.5 mm | 12 mm |
| 12 | L | 19 mm | 8.5 mm |
| 13 | L | 18 mm | 8 mm |

Abbreviations: L; left, R; right.

affected sides (11.12 ± 3.36 mm). We compared the diameters of the pulmonary arteries of the normal and the diseased sides. A two-sample Student t test was used to evaluate the significance of differences of the diameters of the pulmonary arteries of the normal and the diseased sides. There was a statistically significant difference between the sizes of the normal and affected sides ($p < 0.005$).

DISCUSSION

SJS is characterized radiologically by a small- or normal-sized unilateral hyperlucent lung with decreased vascularity and air trapping on expiration. First thought to be of congenital origin, SJS is now considered a postinfectious form of bronchiolitis obliterans and has been observed to occur following pulmonary infection in childhood (6,7). Most patients present with recurrent pulmonary infections, wheezing, coughing, and expectoration; however, a few patients remain asymptomatic (8). Productive cough, dyspnea, occasional hemoptysis, and clinical manifestation of bronchiectasis also occur in SJS. The diagnosis of SJS is usually made on the basis of radiographic findings of hyperlucency of a lung or lobe, which results from air trapping (particularly during expiration) and decreased pulmonary vascular markings in the involved area (7,8).

MRA of the pulmonary vasculature is challenging due to respiratory motion effects, multidirectional course of the pulmonary vessels, cardiac and pulsation artifacts. Pulmonary contrast-enhanced breath-hold MR angiography has successfully addressed these considerations and has become a very promising technique for studying the pulmonary vasculature. Contrast material-enhanced MRA has become established as a powerful noninvasive tool for use in most vascular territories (9-12).

In the lungs, CT angiography is widely regarded as the technique of choice for the work-up of pulmonary vascular disease (13,14). There will remain, however, a subset of patients who, whether because of renal impairment or contrast agent sensitivity, are not candidates for CT angiography. There is, therefore, a role for MRA in the examination of the lungs, but the quality of modern CT angiographic examinations has set a high standard for alternative imaging techniques (15). Advances in MR technology using fast gradients and contrast agents have allowed MRA to make substantial advances. Gadolinium-enhanced MRA is a fast imaging technique that has been shown to accurately evaluate the major arteries such as the carotid system, aorta, pulmonary and renal arteries (16,17). It is possible to assess the entire pulmonary tree in a single breath-hold during the injection of contrast media (18). Three-dimensional contrast-enhanced MRA now offers several advantages that make pulmonary MRA possible. The gradient-echo MRA technique has an intrinsically short TE. This is sufficient to eliminate the susceptibility artifact. Breath-holding eliminates respiratory motion artifact (19).

The advantages of MRA over CT and digital subtraction angiography are lack of ionizing radiation (20). MRA needs no iodinated contrast medium, with its risk of hypersensitivity. Recent improvements in MRI techniques have substantially increased the potential of MRI for the evaluation of pulmonary circulation (21). Faster imaging techniques will allow shorter breathholds, which are necessary especially for patients with severe dyspnea (22). This will result in less severe motion artifacts. The potential use of blood pool agents holds promise with respect to better image quality (21).

In our study, MRA successfully and rapidly depicted the diameters of the small pulmonary arteries with a reduced number and size of branches with diminished peripheral vasculature in all patients. In MRA, both pulmonary arteries and veins are enhanced with the contrast agent, and thus the reviewer must be careful to distinguish them. The main disadvantages of MRA are the limited access to suitable MR technology and claustrophobia. But in this study, the MRA procedure could be tolerated by all patients. According to our knowledge, this study is first study on the pulmonary MR angiography in

SJS. The results of this study suggest strongly that pulmonary contrast-enhanced MRA in SJS is feasible, can depict the pulmonary vasculature in encouraging detail.

In conclusion, this study indicate that gadolinium-enhanced MRA is a fast and accurate technique for delineation of the main pulmonary artery and branches in patients with SJS and can be considered a noninvasive alternative to DSA. Pulmonary MRA may become competitive with the other imaging techniques such as CT angiography and DSA for diagnosing SJS, because of the advantages of lack of radiation and risk of iodinated contrast media. The small pulmonary artery with diminished peripheral vasculature can be visualized clearly. The pulmonary MRA technique might be considered to be a sensitive tool in patients with suspected SJS.

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