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Case Report

Isolated unilateral proximal interruption of the pulmonary artery: findings of high-resolution computed tomography and three-dimensional volume rendering imaging of the pleura

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ABSTRACT

A unilateral proximal interruption of the pulmonary artery is a rare entity that is commonly associated with other congenital cardiovascular anomalies. However, less frequently, this condition may occur as an isolated finding, and some patients are completely asymptomatic. We report 2 cases of asymptomatic patients who had an isolated unilateral proximal interruption of the pulmonary artery. Herein, the radiological imaging findings are described with an emphasis on interlobular septal thickening of the affected lung demonstrated with high-resolution computed tomography. Three-dimensional volume rendering imaging clearly demonstrated reticular opacities on the surface of the affected side of the pleura.

Introduction

A unilateral proximal interruption of the pulmonary artery is an uncommon developmental anomaly. When it occurs in infancy, it is commonly associated with congenital cardiovascular defects. The most common associations include Tetralogy of Fallot and septal defects [1]. An interruption of a main pulmonary artery tends to involve the side contralateral to the aortic arch [1]. It is more commonly left-sided if it is associated with a congenital heart disease, but tends to occur on the right if it is an isolated abnormality. If it is an isolated phenomenon, patients are often asymptomatic for several

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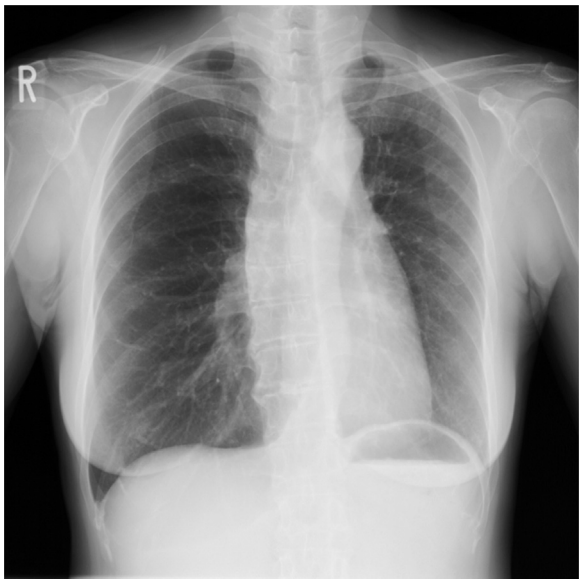

Fig. 1 – Chest radiograph posteroanterior (PA) view from case 1 shows a small hilum on the left. The anterior junction line is displaced to the left, and the reticulation is visible on the periphery of the left lung.

Fig. 2 – A three-dimensional (3D) volume rendering image from case 1 at the pulmonary arterial phase clearly demonstrates the absence of the proximal portion of the left pulmonary artery.

decades and present as adults with either an incidental recognition on an abnormal chest radiograph or hemoptysis [2]. We present 2 cases of proximal interruption of the pulmonary artery. Herein, the radiological imaging findings are described; moreover, interlobular septal thickening of the affected peripheral lung was demonstrated with high-resolution computed tomography (HRCT). Three-dimensional (3D) volume rendering imaging clearly demonstrated reticular opacities on the surface of the affected side of the pleura.

Case report 1

A 67-year-old woman with no significant past medical history was referred for evaluation of an abnormal chest radiograph obtained during an episode of an upper respiratory tract infection.

A chest radiograph (Fig. 1) demonstrated a small-volume left hemithorax and a small left hilum. The contralateral lung had a large volume and was herniated into the smaller hemithorax. Reticulation within the peripheral aspect of the left lung was observed.

Dynamic contrast-enhanced computed tomography (CT) demonstrated complete absence of the proximal portion of the left pulmonary artery at the pulmonary arterial phase. A 3D volume rendering image of the pulmonary arterial phase (Fig. 2) demonstrated proximal interruption of the left pulmonary artery. Enhanced CT indicated dilated, serpiginous abnormal arteries around the left hilum at the systemic arterial phase. Furthermore, we believed that the blood supply to the left pulmonary vasculature was provided by the systemic vessels, dilated bronchial arteries, left intrathoracic arteries, and left inferior phrenic arteries. No costal arteries appeared to be dilated. The distance between the main pulmonary artery and the peripheral left pulmonary artery was only approximately 1 mm. A 3D volume rendering image of the arterial phase (Fig. 3) showed the interrupted proximal portion of the left pulmonary artery, dilated collateral arteries, and peripheral left pulmonary artery.

HRCT on the lung window setting (Figs 4A and B) demonstrated smooth interlobular septal thickening in addition to peripheral, linear, and branching structures perpendicular to

Fig. 3 – A 3D volume rendering image from case 1 at the arterial phase reveals an interruption of the proximal portion of the left pulmonary artery, dilated abnormal arteries around left hilum, and the distal portion of the left pulmonary artery.

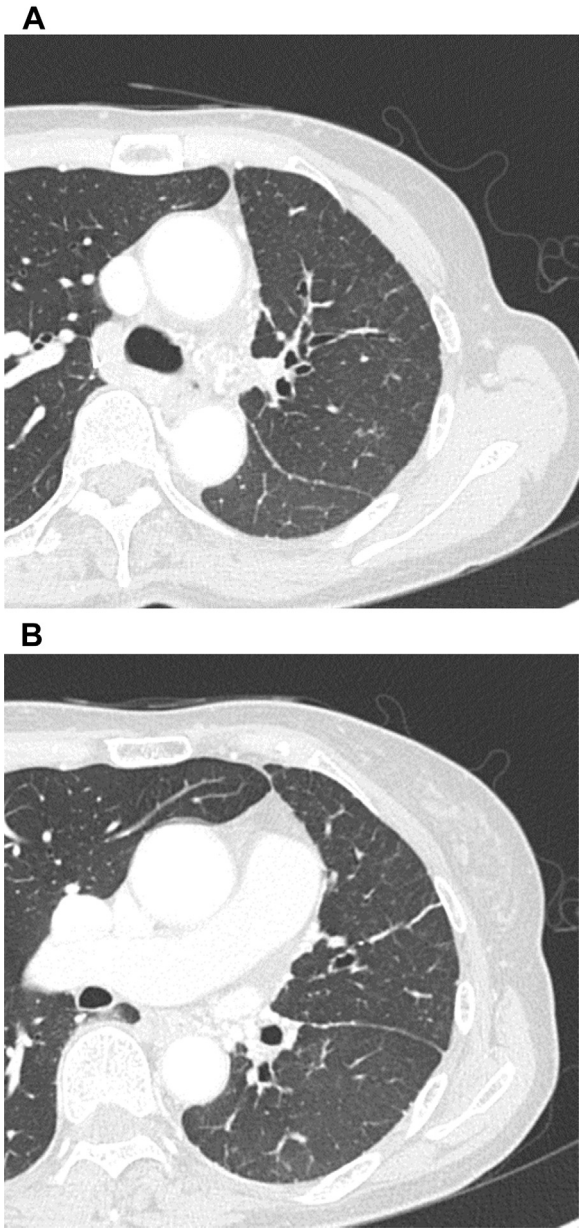

Fig. 4 – (A) HRCT from case 1 at the lower tracheal level demonstrates peripheral fine linear structures, which represent smooth interlobular septal thickening of the affected lung. (B) HRCT from case 1 at the lower bronchus level also demonstrates smooth interlobular septal thickening and peripheral linear structure perpendicular to the pleural surface.

the pleural surface, which were suspected dilated peripheral vasculature (Fig. 4B). A 3D volume rendering image of the pleura (Fig. 5) demonstrated reticular opacities on the surface of the affected side of the pleura. It was prominent, compared with those on the unaffected side. We thought that this finding was indicative of thickening of the peripheral interstitium on the affected side of the pleura.

Perfusion scintigraphy (Fig. 6) was performed with an intravenous administration of technetium-macroaggregated albumin (Tc-99m-MAA); and anteroposterior, left and right lateral, and

Fig. 5 – A 3D volume rendering image of the pleural surface from case 1 demonstrates loss of left lung volume and reticular opacities on the affected (left) side of the pleura. This is obvious when compared with the other side.

oblique projection planar images of the chest region were obtained. An absence of perfusion within the left lung was visualized. This finding confirmed that the blood supply to the left pulmonary vasculature was provided by the systemic vessels.

Fig. 6 – A perfusion scintigraphy from case 1 clearly reveals the absence of perfusion to the left lung.

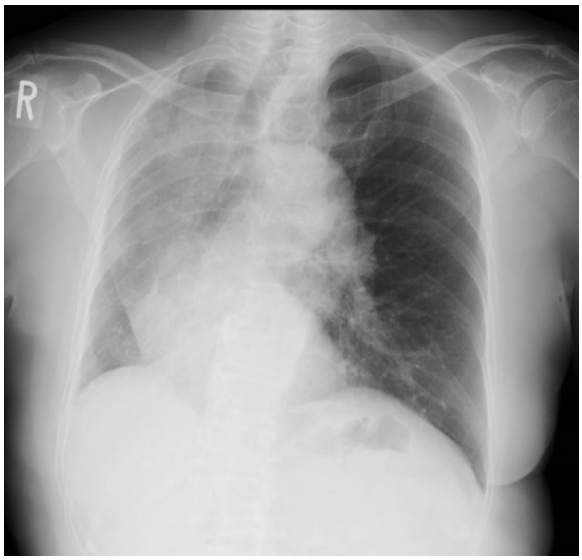

Fig. 7 – Chest radiograph PA view from case 2 demonstrates the loss of right lung volume with a mediastinal shift to the right and hyperlucent contralateral lung herniated across the midline.

An echocardiogram did not reveal any evidence of congenital cardiovascular defects or pulmonary hypertension.

Acquired causes of pulmonary obstruction (chronic thromboembolic occlusion or Takayasu arteritis) were ruled out clinically. The patient was diagnosed with an isolated unilateral proximal interruption of the left pulmonary artery.

Because of the patient's asymptomatic status at the time, no treatment was planned. However, she continued to visit us for follow-up examinations because this condition has potential risks including hemoptysis and pulmonary hypertension.

Case report 2

A 69-year-old woman presented with abnormal findings on a chest radiograph, which were detected during the examination of her cholecystitis. She did not present with any respiratory symptoms. A chest radiograph (Fig. 7) showed loss of right lung volume with a mediastinal shift to the right and hyperlucent contralateral lung herniated across the midline.

A 3D volume rendering image of the pulmonary arterial phase revealed the complete absence of the proximal portion of the right pulmonary artery in the pulmonary arterial phase (Fig. 8). An enhanced CT indicated dilated, serpiginous abnormal arteries around the trachea in the systemic arterial phase (Fig. 9).

Blood supply to the right pulmonary vasculature was considered to be provided by systemic vessels, mainly by dilated bronchial arteries. No costal arteries appeared to be dilated in this case.

HRCT on the lung window setting (Fig. 10) revealed interlobular septal thickening, and a 3D volume-rendered image of the pleura (Fig. 11) indicated prominent reticular opacities on the affected side of the pleura.

Fig. 8 – A 3D volume rendering image from case 2 at the pulmonary arterial phase indicates complete absence of the proximal portion of the right pulmonary artery at the pulmonary arterial phase.

An echocardiogram did not reveal any evidence of congenital cardiovascular defects or pulmonary hypertensions.

Acquired causes of pulmonary obstruction (chronic thromboembolic occlusion or Takayasu arteritis) were ruled out clinically. The patient was diagnosed with an isolated unilateral proximal interruption of the left pulmonary artery.

Since the patient was asymptomatic, no treatment was planned. At the time of this report, she was being treated for

Fig. 9 – An enhanced CT image from case 2 at the tracheal level demonstrates dilated, serpiginous abnormal arteries around the trachea at the systemic arterial phase. These are considered to be dilated bronchial arteries. The loss of volume on the affected side is prominent.

Fig. 10 – HRCT from case 2 at the main bronchus level demonstrates loss of volume of the right lung and smooth interlobular septal thickening of the affected lung is prominent.

the lumbar pain, which is not considered to be related to her condition of the pulmonary artery.

Discussion

Here, we presented 2 asymptomatic cases of patients who had an isolated unilateral proximal interruption of the pulmonary artery and reported on the radiological findings. Frantzel reported the first case of a unilateral proximal interruption of the pulmonary artery in 1868 [2]. The main pulmonary arteries are derived from the proximal aspects of the sixth aortic arch during the intrauterine development. If the proximal portion of the main pulmonary artery fails to appear during embryologic development, this condition develops [3].

Fig. 11 – A right anterior oblique view of 3D volume rendering of the pleural surface from case 2. Note the reticular opacities of the pleura on the affected side and the prominent loss of volume of the affected lung.

Because some patients with a unilateral proximal interruption of the pulmonary artery can remain asymptomatic for a long period, the actual prevalence is difficult to establish [4]. Bouros et al [2] derived the most accurate estimation of its prevalence; among 600,000 participants, only 3 patients were found to have an isolated unilateral proximal interruption of the pulmonary artery. In most cases, the aortic arch is on the contralateral side of the affected pulmonary artery. Ten Harkel et al [4] reported that out of 107 cases, only 3 cases had their aortic arch on the same side as that of the affected pulmonary artery. We presented 2 asymptomatic adult patients who had an isolated unilateral proximal interruption of the pulmonary artery. Furthermore, the aortic arch and the affected pulmonary artery were on the same side in one of our cases (case 1), which is extremely rare [4].

Some authors have previously reported CT findings of a serrated pleural thickening and fine linear structures at the periphery of the lung in cases with a unilateral proximal interruption of the pulmonary artery [1,3,5–8]. These findings were thought to represent transpleural vessels supplying the peripheral lung. Some cases presented CT images of peripheral fine linear opacities on lung window settings with enlarged intercostal arteries on mediastinal window settings [1,5,6,8].

In contrast, Ryu et al [9] reported cases with a proximal interruption of the right pulmonary artery that had reticular opacities on HRCT images. The authors speculated that pleural thickening and adhesion could disrupt the pulmonary lymphatic drainage and caused smooth interlobular septal thickening.

In our cases, HRCT demonstrated interlobular septal thickening of the affected lung, as reported by Ryu et al [9]. In addition, peripheral, linear, and branching structures perpendicular to the pleural surface were also revealed by HRCT. These were suspected to be dilated peripheral vasculature (Fig. 4B). This opacity corresponded to the interlobular septal thickening, which contains peripheral veins and lymphatics in healthy individuals [10,11]. Moreover, 3D volume rendering imaging of the pleura (Figs. 5 and 11) demonstrated reticular opacities on the surface of the affected side of the pleura, which were prominent when compared with the other side. We thought that the reticular opacities on the surface of the affected lung represented the thickening of the interstitium. The dilated collateral vessels developed mainly around a left hilum, and prominent dilatation of the intercostal arteries was not detected in our cases.

The peripheral interstitium extends over the surface of the lung beneath the visceral pleura and envelopes the lung in a fibrous sac, from which the connective tissue septa penetrates the lung parenchyma. The pulmonary veins and lymphatics are contained within the connective tissue of the interlobular septa [10,11]. Smooth interlobular septal thickening is usually observed in association with venous, lymphatic, or infiltrative diseases [10,11].

In our cases, HRCT (Figs. 4 and 10) revealed smooth interlobular septal thickening, and 3D volume rendering imaging of the pleural surface (Figs. 5 and 11) clearly demonstrated the reticular opacities that were considered to indicate thickening of the peripheral interstitium on the affected side. These findings revealed that thickening of the interstitium and interlobular septa may occur under the hemodynamic condition of an isolated unilateral proximal interruption of the pulmonary artery.

There is no doubt that the dynamic changes of the pulmonary flow are the primary causes for peripheral lung opacities on CT; however, there are several potential etiologies. These findings could represent the transpleural vessels directly supplying the peripheral lung as previously reported [1,3,5–8], or peripheral interstitium and interlobular septa may be the pathway of the collateral vessels. They might also be attributed to the dilatation of peripheral pulmonary veins because of increased volume and pressure from the systemic flow, which supplies the peripheral lung. Alternatively, these findings could represent disruption of pulmonary lymphatic drainage caused by pleural thickening and adhesion, as reported by Ryu et al [9].

Further examinations are needed to investigate the etiology of the HRCT findings associated with this condition.

In conclusion, we described 2 cases of an asymptomatic isolated unilateral proximal interruption of the pulmonary artery. Dynamic CT was useful for noninvasive evaluation of the vessels, and perfusion scintigraphy confirmed the hemodynamics of the pulmonary arteries. We reported the HRCT findings of unilateral proximal interruption of the pulmonary artery with a focus on the smooth interlobular septal thickening. 3D volume rendering imaging of the pleura revealed reticular opacities on the pleural surface of the affected side, which was considered to be indicative of the thickening of the interstitium. Proximal interruption of the pulmonary artery should be included in the differential diagnosis of diseases causing interlobular septal thickening.

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