Anomalous pulmonary venous return with esophageal varices is quite rare and most of which have infradiaphragmatic pulmonary venous drainage. It may cause overflow of portal venous system complicating the portal hypertension and esophageal varices. They are mostly demonstrated by angiography or magnetic resonance angiography (MRA) studies, which are either invasive or time-consuming. We present a one-year-old girl with severe esophageal varices, which were incidentally identified during admission for bronchopneumonia. However, she had no clinical symptoms of portal hypertension or hematemesis before. We use multi-detector computed tomography (MDCT) and advanced 3D workstation to clearly demonstrate partial anomalous pulmonary venous return (PAPVR) with right pulmonary veins drain into esophageal vein, which causes severe esophageal varices.

Esophageal varices in children is rare and mostly resulted from portal vein thrombosis and secondary to liver cirrhosis even though liver cirrhosis is the most common etiology of esophageal varices in adult. Other causes such as mediastinal tumor, thymoma and chronic fibrosing mediastinitis have been rarely reported. However, some esophageal varices are still idiopathic which may be due to congenital weakness in venous channel of esophagus [1]. We presented a rare case of esophageal varices, which resulted from the isolated right partial anomalous pulmonary venous return in a child.

CASE PRESENTATION

A one-year-old female infant had progressive dyspnea for days and bronchopneumonia was suspected. She was admitted for further evaluation. She was born via cesarean section due to fetal distress with birth history of G1P1, GA: 39 wks and BBW: 1740g. She had cleft palate, single umbilical artery and skin dimpling over sacral area. She was also diagnosed to have (1) tracheal stenosis of right bronchus; (2) right tracheobronchus with luminal narrowing; (3) congenital heart disease with aortic interruption and patent ductus arteriosus status post correction; (4) left pulmonary artery stenosis status post balloon dilatation; (5) gastroesophageal reflux disease with delayed gastric emptying status post fundoplication and gastrostomy and (6) left kidney hypoplasia.

After admission, a few examinations were done. Bronchoscope was done and showed right tracheobronchus and right main bronchus narrowing. Gastro-esophagoscope incidentally disclosed severe esophageal varices, but the patient had no history.
of hematemesis and had normal blood hemoglobin level of 14 mg/dl. Due to unknown etiology of the esophageal varices, abdominal sonography and Doppler scans was arranged and showed no evidence of portal vein thrombosis with mean velocity of main portal trunk about 20-30 cm/s. Abdominal computed tomography revealed severe esophageal varices without space occupying lesion in the liver parenchyma and no evidence of portal hypertension. The liver biopsy revealed no liver cirrhosis. Angiography of thoracic and abdominal aorta was performed and revealed no vascular lesion at the territory of celiac artery and SMA. There were no varices from portal origin. Due to inconclusive abdominal CT scan and angiographic evaluation, chest CT was arranged. MDCT (Siemens Sensation 16) was performed with a 0.75mm collimation, 12mm per rotation table feed 9 (pitch of 1), 120KV and 66mAs. CT angiographic studies were performed with non-ionic contrast medium with iodine concentration of 320mg/mL (Iopamidol) administered at a dose of 2mL/kg. Studies were performed with contrast material injected by a power injector at a rate of 1mL/sec via right jugular central venous catheter. The bolus tracking ROI was placed on SVC with trigger value of 120 HU. Three-dimensional Maximum-Intensity Projection (MIP) reconstructions were created. The image revealed abnormal right upper pulmonary vein connect to right lower pulmonary vein through abnormal cluster of vessels in right pulmonary parenchyma and the right lower pulmonary veins finally enter lower third esophageal veins and cause engorged esophageal varices. The esophageal varices go upward and connected to left subclavian vein via tortuous collateral vessels including superior intercostal veins. The esophageal varices also extend downward and connect to the coronary vein and result in mild gastric varices. MIP shows abnormal vascular connections between right upper and lower pulmonary veins in right pulmonary parenchyma and severe esophageal varices (Fig. 1); and clearly defines the entry site of right lower pulmonary vein into esophageal veins (Fig. 2), and the connection between esophageal veins and left subclavian vein (Fig. 3). The findings such as right tracheobronchus, right main bronchus stenosis and dilatation of left pulmonary arteries were the same as echocardiography. After discussion with her family, regular follow up and future operation at childhood was suggested. After treatment of her current problems, she was discharged and regularly followed up at our hospital.

**DISCUSSION**

Esophageal veins normally drains into azygos and hemiazygos system. The azygos vein enters SVC from posterior aspect at level of T4 [2]. Sometimes, left superior intercostals vein receives the accessory hemiazygos vein and then drains into brachiocephalic vein.

Esophageal varices and dilated vein in subepithelial connective tissue are most commonly as a result of portal hypertension in adult. In portal hypertension, much portal blood flow reversely to collateral route through the coronary vein, across the esophagogastric hiatus, into the periesophageal plexus before reaching the azygos and hemiazygos systems, SVC and right atrium (RA). The periesophageal plexus communicates with veins in the submucosa of the esophagus and gastric cardia. Increased blood flows through these veins resulting in esophageal varices, also called uphill varices.

Other form of esophageal varices, downhill varices, happened while SVC obstruction due to central catheter related thrombosis, bronchogenic carcinoma, metastatic tumors involving mediastinum, lymphoma, substernal goiter, mediastinal radiation, and sclerosing mediastinitis. In this situation, blood flows downhill through the azygos-hemiazygos system, the periesophageal plexus, and into coronary veins before eventually entering the portal vein, inferior vena cava (IVC) then back to RA.

In children, esophageal varices are rare and mostly result from the extrahepatic etiology. Portal vein thrombosis and secondary biliary cirrhosis are the most common etiologies of esophageal varices. Anomalous pulmonary venous connections comprise varies developmental abnormalities in which blood is returned to the right atrium or its tributaries instead of the left atrium.

The prevalence of a partial anomalous pulmonary venous connection is 0.4%–0.7%. Classically, a partial anomalous pulmonary venous connection presents in childhood, more frequently on the right sided (90%) and is twice as common in males as in females [3, 4]. Eighty percent to 90% of patients have been noted to have associated with atrial septal defects [3, 5]. Isolated partial anomalous pulmonary venous return (without atrial septal defect) occurs in only 0.4% of cases in autopsy series of adult patients without known congenital heart disease [6, 7]. Esophageal varices with anomalous pulmonary venous return had been reported in the literature but quite rare [8, 9]. Most of them with infradia-
Figure 1. Maximum-intensity-projection image shows severe esophageal varices (arrow) and intra-parenchymal connection (triangle) of right superior pulmonary vein and right inferior pulmonary vein which finally drains into lower esophageal veins.

Figure 2. Maximum-intensity-projection image shows drainage of right inferior pulmonary vein into esophageal vein. (arrow)

Figure 3. Maximum-intensity-projection image shows severe esophageal varices and the venous drainage (arrow) into left subclavian vein (triangle) behind left subclavian artery. (star)
phragmatic pulmonary venous drainage may cause overflow of portal venous system complicating the portal hypertension and esophageal varices. They are mostly demonstrated by angiography or MRA studies which are invasive and time-consuming.

After the rapid development of MDCT technique, complex congenital cardiac lesions such as anomalous pulmonary venous return can be demonstrated easily and clearly. It is difficult to trace the whole course of the anomalous pulmonary venous return by using endocardiology, which is safe, non-invasive and inexpensive. Cardiac catheterization is invasive, expensive, time-consuming and sometimes can only depicts faint opacification of venous structure due to dilution of contrast medium and cardiac motion. MDCT has the advantages of high spatial resolution, non-invasiveness and short scanning time than MRI. MDCT also provides information about lung parenchyma, mediastinum and upper abdomen.

In this patient, we clearly demonstrate the course of anomalous pulmonary venous connection with esophageal vein and the long segment venous plexus from gastroesophageal junction upward to lower neck, which finally connects into the left subclavian vein.

We present this case to suggest PAPVR may contribute to uncommon etiology of esophageal varices and multi–detector row CT with volume rendering offer an unprecedented opportunity to study in vivo the anatomy of the systemic and pulmonary veins of the thorax. The complex vascular anatomy can be readily demonstrated by application of three-dimensional techniques.

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單獨食道靜脈曲張：罕見部分肺靜脈回流異常之表徵及電腦斷層影像之表現

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在臨床上，部分肺靜脈回流異常合併食道靜脈曲張頗罕見，且大多數都因為肺靜脈回流至橫隔膜下門脈，導致門脈高壓而產生。先前病例大多有賴於侵入性血管攝影或是核磁共振檢查來確診，我們報告一個罕見案例，此患者右側肺靜脈直接回流到食道靜脈導致食道靜脈曲張，臨床上並無門脈高壓之證據，並且利用多排電腦斷層以及立體（3D）重組，清楚顯示肺靜脈在肺臟之連接以及匯入食道靜脈之接點，導致嚴重食道靜脈曲張之狀況。