Partial Anomalous Pulmonary Venous Return Showing Anomalous Venous Return to the Azygos Vein

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Partial anomalous pulmonary venous return (PAPVR) is a congenital heart disease with a reported incidence of autopsied case. The location of the anomalous pulmonary venous return is usually the right atrium, superior vena cava (SVC), and sometimes the brachiocephalic vein, inferior vena cava (IVC) or coronary venous sinus. Recently we experienced a rare case of PAPVR showing anomalous right total pulmonary venous return to the azygos vein. Furthermore, downward translocation of the right upper lobe bronchus was evident. This rare case is reported along with a review of the related literature.
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Case Report

A 57-year-old woman was found to have a chest roentgenogram abnormality in a mass examination in 1975, but she did not visit a hospital. On May 19, 1990, she visited our hospital because of a common cold. The chest roentgenogram abnormality was again pointed out, and the patient was admitted for detailed examination.

On admission, she was asymptomatic with a temperature of 36.4°C, pulse 78/min., and blood pressure 110/50 mmHg. On examination, a midsystolic murmur (LevineII/VI) was auscultated at the parasternal 2nd intercostal space. There was no abnormality of peripheral blood cell count biochemistry, arterial blood gas analysis or spirometry. An electrocardiogram was also normal.

A chest roentgenogram (Fig. 1) of posteroanterior view showed dilatation of the azygos vein and deformity of the right cardiac edge, and a lateral view showed a tumor shadow which was connected to a vessel behind the cardiac shadow.

From the findings of the lateral view, we initially suspected pulmonary arterio-venous fistula. Next we performed chest computed tomography (CT) and magnetic resonance imaging (MRI). Chest CT scan (Fig. 2) showed that a large pulmonary vein was connected to the azygos vein and that the dilated azygos arch was

connected to the SVC. These findings were more evident on chest MRI (Fig. 3) than on chest CT scan.

From these data, we suspected PAPVR showing anomalous right pulmonary venous return to the azygos vein. Therefore we performed cardiac echography and transesophageal cardiac echography, but no abnormal findings were obtained except for dilated SVC. Next we performed Radioisotope (RI) (^{99m}Tc -Human serum albumin) angiocardiology, which showed disappearance of RI flow in SVC probably due to increased blood flow in the azygos vein (Fig. 4). A lung time activity curve (Fig. 5) showed two peaks, suggesting a left-to-right shunt.

As the findings of RI angiocardiology also strongly suggested PAPVR showing anomalous venous return to the azygos vein, we performed cardiac catheterization (Table 1). O_2 saturation in the azygos vein was 97.4%, which is higher than normal. O_2 saturation in the upper SVC was 54.6% and that in the lower SVC, 93.1%. O_2 saturation step up in the SVC suggested left-to-right shunt through the azygos vein. The pulmonary-to-systemic blood flow ratio was 2.03, and the left-to-right shunt ratio was 53.1%. The venous phase of the pulmonary arterial angiogram (Fig. 6) showed that the whole right pulmonary vein was connected to the azygos vein and that blood was returning to the SVC. Retro-

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Fig. 1. Chest roentgenogram posteroanterior view showing dilatation of the azygos vein and lateral view showing a tumor shadow (arrow head) which was connected to a vessel behind the cardiac shadow.

Fig. 2. Chest computed tomography showing a large pulmonary vein (arrow) connected to the azygos vein and the dilated azygos arch connected to the SVC (arrow head).

Fig. 3. Chest magnetic resonance imaging showing a large vessel (arrow heads) is connected to the dilated azygos vein.

Fig. 4. RI cardioangiography showing disappearance of RI flow in SVC (arrow head) probably due to increased blood flow in the azygos vein.

Fig. 5. Lung time activity curve showing two peaks.

grade venography of the azygos vein (Fig. 7) showed dilatation of the central portion from the pulmonary venous connection of the azygos vein and normal peripheral azygos vein.

The findings of cardiac catheterization provided a definite diagnosis of PAPVR showing anomalous total right pulmonary venous return to the azygos vein.

As PAPVR has a tendency to be associated with pulmonary anomaly, we also performed bronchoscopy. The right upper lobe bronchus was at a lower position than normal. Bronchography (Fig. 8) showed branching of the right upper lobe bronchus from the truncus intermedius, in a lower position than normal. This case was associated with downward translocation of the right upper lobe bronchus.

Discussion

The incidence of PAPVR is 0.4–0.7% of all autopsy cases, PAPVR is not a rare disease, but the location of the anomalous pulmonary venous return is usually the right atrium, SVC or IVC. PAPVR showing anomalous pulmonary venous return to the azygos vein is very rare.

Table 1. Findings of Cardiac Catheterization

	O ₂ saturation (%)	Pressure (mmHg)
azygos vein	97.4	
upper SVC	54.6	
lower SVC	93.1	
upper RA	89.9	} (1)
middle RA	82.4	
lower RA	83.3	
upper IVC	78.2	
lower IVC	77.9	
inlet of RV	81.2	} 30/5*
outlet of RV	77.8	
apex of RV	80.8	
right PA	81.1	33/5 (17)
right PAWP		(4)
left PA	79.5	37/4 (17)
left PAWP		(4)
LV	97.8	129/4*
aorta	96.1	138/80 (110)

RA: right atrium, RV: right ventricle, PA: pulmonary artery, PAWP: pulmonary artery wedge pressure, LV: left ventricle, *: end-diastolic pressure, (): mean pressure

cardiac output: 5.22 l/min.

cardiac index: 4.02 l/min/m²

pulmonary-to-systemic blood flow ratio: 2.03 left-to-right shunt ratio: 53.1%

Fig. 6. Venous phase of the pulmonary arterial angiogram showing the whole right pulmonary vein connected to the azygos vein.

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Fig. 7. Retrograde venography of the azygos vein showing dilatation of the central portion of the azygos vein (arrow heads) from the pulmonary venous connection of the azygos vein and normal peripheral azygos vein (arrows).

Table 2. Case Reports of PAPVR with Anomalous Pulmonary Venous Return to Azygos Vein

No.	Author	Year	Age	Sex	Veins to azygos	Other anomalies
1	Guillabert	1859	55 yr.	M	Right middle lobe	Patent foramen ovale, right upper lobe veins to SVC
2	Theremin	1884	11 da.	F	Right lung	Agenesis of the left lung, one-lobed right lung
3	Theremin	1884	4 mo.	F	Right lung	Agenesis of the left lung, two-lobed right lung
4	Shepherd	1890	30 yr.	F	Right lung	Arterial anomalies
5	Hurwitz	1937	7 wk.	F	Left lung	Agenesis of the left lung, one-lobed right lung
6	Bordy	1942	56 yr.	F	Right upper lobe	Agenesis of the left lung, one-lobed right lung
7	Kjellberg ⁷⁾	1955	26 yr.	F	Two right upper lobe veins	Absent IVC, anomalous left brachiocephalic vein, pulmonary sequestration
8	Stecken	1963	18 yr.	F	One right lower lobe vein	Right upper lobe veins to IVC
9	Howard	1964	6 yr.	F	Right lung	Patent foramen ovale
10	Goldfarb	1966	53 yr.	F	Right upper lobe	Mitral stenosis
11	Steven	1979	8 yr.	F	Right lung	
12	Makino	1986	32 yr.	F	Right upper lobe, One right lower lobe vein	
13	Miyata	1988	53 yr.	M	Right upper lobe, One right lower lobe vein	
14	Our case	1991	57 yr.	F	Right lung	Downward translocation of right upper lobe bronchus

Fig. 8. Bronchography showing downward translocation of the right upper lobe bronchus.

A review of the world literature (1–11) revealed 13 case reports, and our case is the 14th (Table 2). Ninety percent of cases of PAPVR are associated with atrial septal defect (ASD), but the 13 reported cases of PAPVR showing anomalous pulmonary venous return to the azygos vein were not associated with ASD, as was the case in our patient. Five of the 13 reported cases were associated with pulmonary anomaly, including pulmonary hypoplasia and anomaly of the lung lobe. The present case was associated with downward translocation of the right upper lobe bronchus.

Regarding the treatment of PAPVR, surgical correction is usually indicated when the pulmonary-to-systemic blood flow ratio exceeds 1.5, because such PAPVR will often progress to pulmonary hypertension in spite of the lack of symptoms. Therefore surgical correction was indicated in the present case, but the patient did not consent in view of her asymptomatic condition. We are following her up at the outpatient clinic of our hospital.

This rare case of PAPVR showing anomalous venous return to the azygos vein is the 14th of its kind reported in the world. This case however, was also associated with downward translocation of the right upper lobe bronchus.

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