CASE REPORT

Beating the odds: a rare case of supracardiac total anomalous pulmonary venous return (TAPVR) in an adult patient

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This case report had won second place in poster presentation during the 21st National Conference on Medical and Health Sciences (17–18 October 2016) held in School of Dental Sciences, Health Campus, Universiti Sains Malaysia, Kelantan, Malaysia.

Accepted 9 January 2018

SUMMARY

Total anomalous pulmonary venous return (TAPVR) is a rare congenital heart defect, and patients are usually symptomatic at a very young age. Survival to adulthood without surgical correction is extremely rare. We report a 33-year-old woman with a heart murmur and a history of a successful pregnancy. Echocardiogram revealed a large atrial septal defect with suspicious pulmonary vein anomaly. Chest radiograph demonstrated classical 'snowman' configuration. Cardiac catheterisation was consistent with anomalous pulmonary venous drainage. Cardiac CT confirmed supracardiac TAPVR, whereby all the pulmonary veins drain into the anomalous vein and finally to the superior vena cava. She remained asymptomatic and underwent a successful surgical repair.

BACKGROUND

The incidence of total anomalous pulmonary venous return (TAPVR) is 0.6 to 1.2 per 10000 live births. This accounts for only 0.7% to 1.5% of children with congenital heart disease.¹ Patient survival into adulthood is less than 7%, and it usually occurs in supracardiac type without pulmonary venous obstruction.^{2,3} This case report describes radiological findings of an adult patient with supracardiac TAPVR. To the best of our knowledge, this is the first case report in Malaysia.

CASE PRESENTATION

A 33-year-old woman was incidentally found to have a heart murmur during the regular maternal check-up in mid-2015 at the nearby district hospital in Kelantan, Malaysia. She also claimed to have been diagnosed with a heart murmur during her first pregnancy in June 2008. After having a successful pregnancy

and delivery with no complication, she defaulted the follow-up appointment. However, following a miscarriage of her second pregnancy in June 2015, she was referred to our hospital for further cardiac assessment. During the visit, she was pink and was not in respiratory distress. There was no digital clubbing, and the peripheral capillary refill was less than 2s. The peripheral capillary oxygen saturation was 96%–98% under room air with a blood pressure of 108/61 mm Hg. Cardiovascular examination revealed a heart rate of 80 bpm with regular rhythm. Normal heart sounds with the presence of ejection systolic murmur grade 3/6 were audible at the left upper sternal border with evidence of displaced apex beat.

INVESTIGATIONS

Chest radiograph revealed smooth widening of the superior mediastinum resembling a classic 'snowman' configuration. The cardiothoracic ratio was increased at 0.58 with no evidence of cardiac failure (figure 1A). Serial radiographs showed no significant changes (images not shown).

Transoesophageal and transthoracic echocardiograms performed in November 2015 showed dilated right atrium (RA) and right ventricle (RV), large atrial septal defect (ASD) with bidirectional shunt, mainly from right to left. An ascending vein connecting to the dilated superior vena cava (SVC) were identified (images not provided). These structures likely form the respective convex left and right superior mediastinal border on the chest radiograph thus raising a high suspicion of pulmonary vein (PV) anomaly (figure 1B). Turbulence flow was noted in the main pulmonary artery with a pressure gradient (PG) of 71 mm Hg (4.2 m/s) suggestive of pulmonary stenosis.

Cardiac catheterisation was subsequently performed which revealed common drainage of the right upper, right lower and some inferiorly positioned left PV into a collecting vein. This vein ascended and drained into the left innominate vein and finally to the SVC (figure 2A–D). The inability to demonstrate the superiorly directed left PV has raised the possibility of a partial anomalous pulmonary venous drainage. Haemodynamic data obtained as follows: aortic pressure 113/65 (85) mm Hg, main pulmonary artery pressure 39/9 (22) mm Hg, RV pressure 63/3 mm Hg. The elevated RV pressure with a pressure gradient of 24 mm Hg was also suggestive of pulmonary stenosis. Oxygen

Figure 1 (A, B) Chest radiograph: classic snowman appearance of supracardiac TAPVR. Note that the mediastinum is widened in relation to cardiac heart border. TAPVR, total anomalous pulmonary venous return.

To cite: Talib N, Sayuti KA, Abdullah MS, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-221074

Figure 2 (A–D) Cardiac catheter angiogram from femoral vein approach: contrast study shows right upper, right lower and left PVs drain into the collecting vein, join to form an anomalous vertical vein into left innominate vein and SVC. Lt, left; PV, pulmonary vein; Rt, right; SVC, superior vena cava.

saturation from arterial blood gas was 92.6% under room air. The calculated pulmonary vascular resistance (PVR) was 1.7 wood unit.

A cardiac CT scan was requested to further delineate the anomalous PV and to look for any residual pulmonary venous drainage to the left atrium (LA) (figure 3A,B, figure 4). The CT scan confirmed supracardiac TAPVR, whereby there were one right superior PV (from upper and middle lung lobes), one right inferior PV, one left inferior PV and four superior PVs draining separately into a collecting vein (figure 5). The collecting vein was located above the LA and ascended anterior to the left pulmonary artery (LPA) to form a dilated vertical vein. This vein drained into a dilated left innominate vein and eventually into the SVC. There was no evidence of pulmonary venous obstruction. Other CT findings include situs solitus, ASD, cardiomegaly

Figure 3 (A, B) Contrast-enhanced CT cardiac with maximum intensity projection in coronal view: all the right PV and left lower PV drain into a collecting vein. Later followed by left upper PV to form an anomalous dilated vertical vein which eventually drains into left innominate vein and SVC. LT, left; PV, pulmonary vein; RA, right atrium; RT, right; SVC, superior vena cava; IVC, inferior vena cava.

with the dilated RV forming the left lateral heart border. The coronary artery origin and the tracheobronchial tree anatomy were normal.

TREATMENT

The patient was referred to our national cardiothoracic centre where she underwent a successful surgical correction in June 2017. Surgical findings confirmed the diagnosis of supracardiac TAPVR with dilated innominate vein, SVC and the right heart chambers. The pulmonary artery was also larger than the aorta and tense to palpation. The vertical vein measured 20 to 25 mm and the ASD was large measuring 30 mm×20 mm in diameter. Anastomosis between the LA and the confluence of PVs was performed. CardioCel patch was used to repair the atrial septum and partition the atrial chambers.

OUTCOME AND FOLLOW-UP

Patient was discharged well on the sixth postoperative day.

DISCUSSION

TAPVR describes abnormal communication between all of the PV and the right side of the heart instead of the LA. It results from an embryological developmental failure of the PV which causes persistent patency of primitive systemic veins. This condition leads to the mixing of oxygenated and deoxygenated blood in the circulations. TAPVR is a symptomatic congenital cardiac disease and it is commonly associated with other congenital anomaly such as heterotaxy syndrome. It is classified into four types which are supracardiac, cardiac, infracardiac and mixed pattern. The most common type is the supracardiac TAPVR that accounts for 50% of all TAPVR while the rarest type is the mixed pattern. The anomalous vein may become obstructed or non-obstructed depending on its course. At neonatal age, patients commonly present with cyanosis and respiratory distress requiring early surgical intervention. The goal of surgery is to re-establish a direct pathway between the PV and the LA, as well as removing any pulmonary venous obstruction.

The unusual presentation of our patient is she has been living with a heart murmur but remained asymptomatic until her third decade of life without the need of medication. There are several factors which contribute to the long survival in supracardiac TAPVR, for example, absence of concomitant congenital heart disease (eg, isomerism), a large ASD, absence of the pulmonary venous obstruction, low pulmonary arterial resistance and mild/moderate tricuspid valve regurgitation.^{3,4} A large ASD prevents pulmonary overcirculation and maintains a satisfactory cardiac output through right to left shunt.⁵ This also contributes to the amount of mixing of the oxygenated blood which explains the lack of cyanosis in our patient. The unobstructed pulmonary venous circulation as seen in the CT scan of this patient is also a factor for late diagnosis. Pulmonary venous obstruction usually occurs when the vertical vein is located posterior to the LPA and trapped between the dilated LPA and left bronchus. Narrowing at the entrance to the innominate vein may also occur.⁶

The oldest reported patient who had a successful TAPVR repair was at the age of 57 years.⁷ The oldest patient with unrepaired TAPVR was reported to survive until the age of 62 years.⁸ In 1994, another TAPVR patient with unnoticed heart murmur was reported to have a successful pregnancy at 29 years old.⁹ This report described pregnancy and successful delivery are possible if the pulmonary pressure and vascular resistance are not markedly elevated.

Figure 4 Contrast-enhanced CT cardiac with maximum intensity projection in four-chamber view. Large ASD with dilated RA and RV noted. Note that the left heart border bordered by the dilated RV (red arrow). ASD, atrial septal defect; LA, left atrium; RA, right atrium; RV, right ventricle.

The classical 'snowman' configuration on the chest radiograph is usually seen in supracardiac TAPVR. Additional finding like pulmonary venous congestion is only appreciated when there is significant pulmonary venous obstruction. However, the radiographic appearance may be non-specific in some patients and should not be used as the main tool for making the diagnosis of TAPVR. The radiograph may also lead to the diagnosis of a mediastinal mass for the untrained eyes. Nonetheless, the recognition of an abnormal chest radiograph combined with heart

Figure 5 Three-dimensional volume rendering image in coronal view. LT, left; IJV, internal jugular vein; PV, pulmonary vein; RT, right; SVC, superior vena cava.

murmur should prompt a good medical practitioner for a second opinion or referral.

Other than echocardiogram which is highly operator-dependent, cardiac catheterisation is the gold standard imaging modality to diagnose TAPVR. However, the availability of other imaging modalities such as CT and MRI, which are non-invasive but equally accurate in making the diagnosis, has made cardiac catheterisation less needed. Cardiac catheterisation is reserved for cases which require haemodynamic information and for patients who are suitable for palliative catheter procedures.¹

Both cardiac CT and MRI provide accurate anatomical delineation of pulmonary venous pathways and their connections. Intravenous contrast will optimise the visualisation of the vessels. The advantage of CT scan over MRI is superior spatial resolution that enables accurate evaluation of the airways, lung parenchyma and other associated structural anomalies which is useful in preoperative planning and postoperative assessment.

Despite having a rare type of congenital heart disease, our patient has been living a normal life without limitation to her daily activities. However, the recent miscarriage has raised some concerns. Her systemic circulation which is dependent on the right to left shunt across the ASD was likely to be impaired especially in the third trimester during which the resting cardiac output of normal individuals usually increases by 50%–100%. This could have resulted in reduced foetal perfusion which potentially leads to intrauterine growth restriction, premature delivery and miscarriage. Although her baseline oxygen saturation was normal, it is likely that this has also dropped during the pregnancy, thus further increased the risk of pregnancy complications. In spite of the late diagnosis of TAPVR with a miscarriage, surgical repair is still an option in adulthood and a few case reports have suggested favourable outcome.^{5,7}

Learning points

- ▶ The key factors favouring supracardiac total anomalous pulmonary venous return (TAPVR) survival into adulthood are the presence of large atrial septal defect and unobstructed pulmonary venous circulation.
- ▶ Patients with asymptomatic congenital heart disease, abnormal physical examination and chest radiograph warrant referral to the paediatric cardiologists.
- ▶ Despite classic appearance in some diseases, radiograph alone is non-diagnostic in detecting many congenital cardiac defects.
- ▶ Multiple imaging modalities are extremely useful in making a diagnosis of a complex congenital heart disease like TAPVR since they can provide anatomical and functional information.
- ▶ Sophisticated imaging software combined with a well-trained cardiac imaging team will help to improve the accuracy of diagnosis and thus leads to the best management strategy for the patients.

Acknowledgements The authors' appreciation goes to colleagues and supporting staffs. Their contributions are sincerely appreciated and gratefully acknowledged.

Contributors The completion of this undertaking could not have been possible without the participation and assistance of all the authors. NT and KAS planned, conducted, reported and reconstructed the images and confirmed the correct diagnosis. MRMZ had provided important information from the physician's perspective including the findings on angiography study.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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