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Late-onset total anomalous pulmonary venous connection in a 70-year-old woman

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SUMMARY

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital cardiac anomaly. There are a few reports of untreated TAPVC diagnosed in patients older than 60 years. Herein, we report the successful surgical treatment of TAPVC in a 70-year-old woman. A 70-year-old woman with TAPVC presented with symptoms of acute heart failure. We closed an atrial septal defect and performed tricuspid annuloplasty and commissurotomy of the pulmonary valve. Postoperative CT showed no residual shunt, and the pulmonary veins drained into the left atrium. She had an uneventful postoperative course. This report describes the case of the oldest known patient who underwent surgical treatment for TAPVC. Surviving into adulthood with little or no symptoms is uncommon in patients with TAPVC, and cases of late-onset TAPVC, such as our case, are rare. Nevertheless, close vigilance is necessary to prevent misdiagnosis in patients with this clinical presentation.

BACKGROUND

Total anomalous pulmonary venous connection (TAPVC) is a rare congenital cardiac anomaly of the pulmonary veins characterised by abnormal connections of the pulmonary veins to the systemic venous circulation instead of the left atrium. TAPVC develops in 1 of 7809 persons, with an estimated 504 cases annually in the USA.¹ Patients with TAPVC generally have an unfavourable prognosis, and only ~20% of patients survive the first year of life without intervention.² There are few case reports of untreated TAPVC diagnosed after the age of 60 years.^{3 4} Herein, we report the successful surgical treatment of intracardiac TAPVC in a 70-year-old woman. To the best of our knowledge, she is the oldest patient who has undergone TAPVC treatment to date.

CASE PRESENTATION

A 70-year-old woman presenting with palpitations, exertional dyspnoea and mild oedema of the lower extremities was admitted to our hospital for evaluation. Her medical history revealed that she had a soft pulmonary mid-systolic flow murmur (in the second and third left intercostal spaces) during childhood but had not presented at a hospital since then. All other parameters were non-significant. She had not experienced any episodes of chest discomfort, palpitations or exertional dyspnoea until

recently. She was diagnosed as having acute heart failure. Her serum B-type natriuretic peptide (BNP) level was 1333 pg/mL and arterial saturation at presentation was 90% in room air.

INVESTIGATIONS

Transthoracic and transoesophageal echocardiography, cardiac catheterisation and CT during admission revealed the following: (1) a large atrial septal defect (ASD) with a diameter of 2.9 cm and shunt flow from the left to the right atrium ($Q_p/Q_s=5.99$); (2) all four pulmonary veins joined to form an anomalous common pulmonary vein that drained into the right atrium; (3) the coronary sinus drained into the dilated right atrium (77.8×94.8 mm); (4) severe tricuspid regurgitation; (5) moderate regurgitation and stenosis of the pulmonary valve; (6) pulmonary hypertension (PH) (systolic pulmonary artery pressure: 51 mm Hg, mean pulmonary artery pressure: 29 mm Hg, pulmonary artery wedge pressure: 24 mm Hg)⁵ and (7) low pulmonary artery resistance (4.0 Wood units) (figure 1A–C). We made a diagnosis of PH in the TAPVC with a large ASD.

TREATMENT

The patient underwent surgery to correct these anomalies. After establishing cardiopulmonary bypass (CPB), the ascending aorta was cross-clamped, and cardiac arrest was achieved through antegrade cardioplegia. The right atrium was opened with the usual oblique incision. We found a large ASD (figure 2A) and the opening of an anomalous common pulmonary vein in the right atrium. The opening of the coronary sinus was far from that of the pulmonary vein. A fine fibromuscular septum separated the pulmonary vein confluence from the left atrium. The size of the left atrium was reduced, and no pulmonary vein

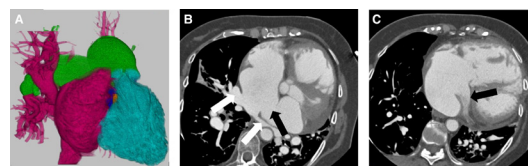


Figure 1 Preoperative CT findings: (A) All four pulmonary veins are seen draining into the right atrium (red), as demonstrated by CT. (B) A large atrial septal defect with a diameter of 2.9 cm (indicated by a black arrow) and bilateral pulmonary veins joining the right atrium (indicated by white arrows). (C) Coronary sinus opening seen in the dilated right atrium (black arrow).



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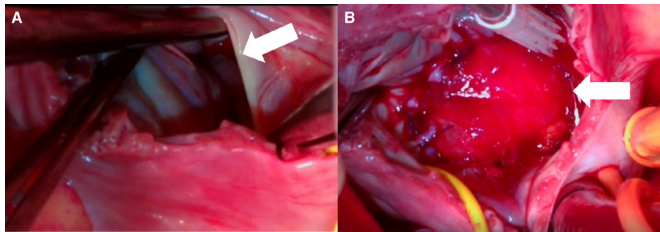


Figure 2 Intraoperative findings: (A) A large atrial septal defect with a diameter of 2.9 cm (white arrow). (B) The fibromuscular septum and a part of the atrial septum were removed and patched with autologous pericardium around the openings of the pulmonary veins to eliminate potential distortion or narrowing of the pulmonary veins (white arrow).

was draining into it. The fibromuscular septum and a part of the atrial septum were removed and patched with autologous pericardium around the openings of the pulmonary veins to eliminate potential distortion or narrowing of the pulmonary veins (figure 2B). Careful observation of the pulmonary valve revealed a small amount of calcification in the annulus and a minor fusion in the commissure; the calcifications were removed and commissurotomy performed. Tricuspid annuloplasty was performed using a 31 mm Edwards MC3 ring (Edwards Lifesciences, Irvine, California, USA). Postoperative transthoracic echocardiography revealed no shunt flow from the left to the right atrium, with improvement in pulmonary and tricuspid regurgitations. Aortic cross-clamp, CPB and operation times were 80, 128 and 213 min, respectively. No residual shunt was observed; all four pulmonary veins appeared to be draining into the left atrium, and the coronary sinus opened into the right atrium, as observed on CT (figure 3A–C). PH and postoperative BNP improved (systolic pulmonary artery pressure: 34 mm Hg, mean pulmonary artery pressure: 21 mm Hg, pulmonary artery wedge pressure: 12 mm Hg, BNP level: 308 pg/mL). We did not perform any further invasive pulmonary haemodynamic measurements.

OUTCOME AND FOLLOW-UP

The patient had an uneventful postoperative course and was discharged home on postoperative day 10. She was energetic during her follow-up visit 1 month after discharge. The patient gave written informed consent for the publishing of her case.

DISCUSSION

TAPVC is a rare congenital anomaly that may cause severe PH and congestive heart failure. Fetal echocardiography is technically challenging but helpful in the early diagnosis of congenital cardiac anomalies. While most patients with TAPVC have no



Figure 3 Postoperative CT findings: (A) Postoperative CT image showing all four pulmonary veins draining into the left atrium (blue). (B) No residual shunt from the atrial septal defect is seen in the postoperative image owing to the autologous pericardial patch (black arrow). (C) Coronary sinus opening seen in the right atrium (black arrow).

symptoms at birth, about 50% develop symptoms during the first month of life. Without surgical repair, most of them die within the first year of life. There have been several case reports of adults with untreated TAPVC but only a few reports of cases diagnosed after the age of 60 years.^{3,4}

TAPVC is classified into four types: supracardiac, cardiac, infracardiac and mixed connection.⁶ In general, patients with supracardiac or cardiac connection survive longer than those with infracardiac or mixed connection.⁷ The major factors that determine the survival of these patients are the size of the ASD and pulmonary arterial pressure.³ Our patient had a large ASD, which was thought to be the reason for her long survival.

We decided to perform a surgical repair for our patient to improve the symptoms of heart failure caused by the large right-to-left shunt, severe regurgitation of the tricuspid valve, moderate regurgitation and stenosis of the pulmonary valve. Postoperative tricuspid insufficiency and cardiac arrhythmias may have negative long-term effects on patients with TAPVC.⁸ In summary, surviving into adulthood with little or no symptoms is uncommon among patients with TAPVC, and cases of symptoms developing late in life are sporadic, such as in our patient. Since the use of fetal echocardiography was not widespread in the past, we may encounter more cases of late diagnosis or undiagnosed TAPVC in the future, among elderly patients presenting

Patient's perspective

Around the spring of 2020, I started to feel palpitations and suffocation when climbing up stairs. Gradually, I noted swelling of both my lower legs and started gaining weight. When I visited the hospital, I was diagnosed with acute heart failure. In May, I learnt that the cause was TAPVC. I was very shocked. I heard from my mother that I had a heart murmur when I was a child, but I was not taken to the hospital because no major symptoms were noted. I became a member of a society, got a job, and experienced childbirth. After that, I went to the hospital because of a cold, but I was fine. I was started on medicines for controlling heart failure for 3 months, but I decided to undergo because my breathlessness did not improve. Since I had no other symptoms, I was surprised that the cause was a heart abnormality that was pointed out back when I was a child. The doctor also said that he had never encountered any patient with late-onset symptoms of TAPVC, who remained without symptoms for as long as I have. The doctor was surprised. The symptoms of heart failure that I had before, have disappeared now. I am living well. I'm glad I had the surgery.

Learning points

- ▶ The survival of patients with total anomalous pulmonary venous connection (TAPVC) depends on the size of the atrial septal defect and pulmonary arterial pressure.
- ▶ Surviving into adulthood with little or no symptoms is uncommon in patients with TAPVC, and in rare cases, the patients develop symptoms late in life, as noted in this report.
- ▶ Since fetal echocardiography was not commonly performed in the past, we may encounter more cases of late diagnosis or undiagnosed TAPVC in the future among elderly patients presenting with heart failure.

with heart failure. Nevertheless, close vigilance is necessary to prevent misdiagnosis when patients present with clinical features similar to those in our patient.

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