CASE REPORT

Late presentation of TAPVC with multiple cerebral abscesses

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SUMMARY

We report the case of a 24-year-old man, presenting with fever, headache, vomiting and seizures, subsequently diagnosed with cyanotic congenital heart disease. Evaluation revealed non-obstructive supracardiac total anomalous pulmonary venous connection (TAPVC) as the underlying disorder. Surprisingly, the patient denied any past cardiac symptoms. Presentation in adulthood is infrequent for TAPVC, and primary manifestation with cerebral abscesses is still more unusual.

BACKGROUND

Total anomalous pulmonary venous connection (TAPVC) is a rare cyanotic congenital heart disease characterised by drainage of all four pulmonary veins into the right atrium through anomalous connections. Based on the pattern of communication between the pulmonary veins and the right atrium, TAPVC can be further subclassified as supracardiac, cardiac, infracardiac and mixed. Although cyanotic congenital heart disease is a known risk factor for cerebral abscess, primary presentation with this complication is limited to individual case reports. The reason for this paucity is unclear since the basic mechanism of abscess formation, that is, haematogenous dissemination of circulating bacteria bypassing pulmonary filtration through the shunt lesion, is operative in TAPVC.

This case report is important because it highlights an atypical and heretofore unreported presentation of TAPVC. Although rare, underlying congenital heart disease should be carefully ruled out in all patients with multiple cerebral abscesses.

CASE PRESENTATION

A 24-year-old man presented with high-grade fever, severe headache and recurrent vomiting over the past 3 days. He had also suffered one episode of generalised tonic-clonic seizure. Since the past 1 day, he had developed sudden onset weakness of the left side of his body. His medical history was uneventful. He denied any history of substance abuse. Careful questioning failed to elicit any past cardiac symptoms. He was born of a non-consanguineous marriage, and the antenatal period was without incident.

At admission, the patient was haemodynamically stable, conscious and oriented. General physical examination revealed mild cyanosis, and no clubbing. Neurological examination confirmed the presence of flaccid hemiparesis on the left. Plantar reflex was extensor on the left side with mildly

exaggerated deep tendon reflexes. Sensory examination was normal. Cardiovascular examination demonstrated a right ventricular apical impulse with a sustained left parasternal heave. A grade 3/6 ejection systolic murmur was audible at the left second intercostal space.

INVESTIGATIONS

Routine laboratory investigations showed normal blood glucose, serum electrolytes and renal and liver function tests. Complete blood counts and erythrocyte sedimentation rate were also normal. Arterial saturation at presentation was 84% on room air. Electrocardiography showed right axis deviation, right atrial enlargement and right ventricular hypertrophy. Chest X-ray (figure 1) revealed pathognomonic snowman appearance typical of supracardiac TAPVC. Transthoracic echocardiography was corroboratory, showing TAPVC of supracardiac type with all four pulmonary veins joining to form a vertical vein draining into the superior vena cava (figures 2 and 3). Echocardiography also confirmed situs solitus, levocardia with atrioventricular/ventriculoarterial concordance and a left-sided aortic arch (figure 2). A large ostium secundum atrial septal defect measuring 20 mm was seen with right to left shunting. The interventricular septum was intact.

Figure 1 Chest X-ray (posteroanterior view) showing pathognomonic snowman or figure-of-eight appearance of supracardiac total anomalous pulmonary venous connection. The lower part of the snowman appearance is formed by the heart and the upper part is formed by the dilated vertical vein (VV), innominate vein (IV) and the superior venacava (SVC) as denoted.

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Figure 2 (A) Subxiphoid short axis view at the level of diaphragm shows IVC to the right (blue) and aorta to the left (red) of the spine. (B) Subcostal sagittal view showing IVC draining into the right atrium. (C) Parasternal long axis view showing the pulmonary venous confluence posterior to the left atrium. RA, right atrium; RV, right ventricle; LV, left ventricle; IVC, inferior vena cava; Ao, aorta; Sp, spine; Li, liver; PVC, pulmonary venous confluence.

Moderate pulmonary arterial hypertension was noted with mild tricuspid regurgitation. The right atrium and right ventricle were dilated.

Contrast-enhanced cranial CT showed multiple ring enhancing lesions in both cerebral hemispheres (figure 4).

Stereotactic aspiration from the lesions yielded thick pus. Aerobic and anaerobic cultures of the aspirate were persistently sterile.

DIFFERENTIAL DIAGNOSIS

Supracardiac TAPVC complicated by multiple cerebral abscesses producing hemiparesis.

TREATMENT

Empirical parenteral antibiotic therapy was initiated with ceftriaxone (2 g intravenous q12 h) and metronidazole (500 mg intravenous q6 h) and continued for 6 weeks. One week after initiation of antibiotic therapy, the patient became progressively drowsy. Neurological examination showed bilateral extensor plantar response; motor examination was unchanged. Repeat

cranial CT showed significant increase in the hydrocephalus with markedly asymmetric dilation of the right lateral ventricular system. Midline shift with subfalcine herniation to the left was also noted (figure 5). Neurosurgical consultation was taken and emergency external ventricular drainage was performed. Following the procedure, there was progressive improvement in sensorium, although neurodeficits persisted. Ventriculoperitoneal shunt was then performed.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful, and the patient was fully conscious and oriented. Neurodeficits persisted however, with only minimal improvement in power. Physiotherapy was initiated once motor tone improved. Surgical correction of TAPVC was advised, but refused by the patient on financial grounds. He was subsequently discharged and kept on follow-up. On follow-up, he showed steady improvement in motor power with complete recovery of all deficits 1 year after the index illness.

DISCUSSION

Neurological complications constitute a large component of morbidity associated with cyanotic congenital heart disease, and can arise from a variety of mechanisms ranging from the acute global hypoxia of cyanotic spells to focal ischaemia due to arterial thrombosis, paradoxical embolisation and cardioembolisation.¹ Cerebral abscesses comprise a distinct cause for neurodeficits in cyanotic congenital heart disease, and 25–46% of all cases of cerebral abscess can be attributed to underlying congenital cardiac disease.² Proposed pathogenic processes for abscess formation include chronic hypoxia predisposing to anaerobic infection, sluggish microcirculation and passage of infected blood across right to left shunts.¹

TAPVC is an uncommon form of complex cyanotic congenital heart disease and comprises 2% of all congenital heart diseases.³ It is characterised by anomalous drainage of all four pulmonary veins into the right atrium. Blood flow to the left atrium then occurs across an atrial septal defect (ASD). Thus TAPVC is associated with obligatory right-to-left shunting of mixed oxygenated and deoxygenated blood. Associated pulmonary venous obstruction or a restrictive ASD can produce early onset pulmonary hypertension and rapid clinical deterioration. This course is often seen with infracardiac TAPVC³ and less commonly with the supracardiac form.

The pathophysiology of TAPVC demands the presence of substantial right-to-left shunting for survival into adulthood; the

Figure 3 Transthoracic echocardiography demonstrating an echo-free space posterior to LA (star) representing the pulmonary venous confluence and a large ostium secundum atrial septal defect (arrow) shunting right to left (A, four-chamber view). Torrential flow in SVC is noted draining into RA (B, four-chamber view). (C) The ascending vertical pulmonary venous confluence drainage into the IV in a suprasternal view. RA, right atrium; LA, left atrium; RV, right ventricle; IV, innominate vein; VV, vertical vein; LV, left ventricle; SVC, superior vena cava.

Figure 4 Contrast-enhanced cranial CT (axial section, A and B), (sagittal section, C) showing multiple well-defined hypodense lesions of varying sizes with thin uniform ring-like peripheral enhancement and variable perilesional oedema. Lesions are noted at the grey white junction, diffusely scattered in both cerebral hemispheres. The largest lesion is seen in the right parietal lobe surrounded by perilesional oedema.

Figure 5 Contrast-enhanced cranial CT showing progression of hydrocephalus with markedly asymmetric dilation of the right lateral ventricular system (sagittal section, A; coronal section, B). Midline shift with subfalcine herniation to the left is also noted.

presence of such shunting implies increased risk for development of cerebral abscesses. Although a case series of patients with cyanotic congenital heart disease and cerebral abscess could not identify any cases of TAPVC² and other larger studies focusing on TAPVC did not report cerebral abscess as a complication,^{3–5} it is likely that such cases have gone unreported in the past. The impact of dilution of bacteremic deoxygenated blood from the vena cava with oxygenated blood filtered from pathogens in passage through the pulmonary circulation could partially explain the apparently lower incidence of cerebral abscess among patients with TAPVC; there is however, no available data to support this hypothesis. Presence of an ASD is in itself a risk factor for development of cerebral abscess; bidirectional shunting is important however, so that cerebral abscesses are rarely seen in patients with isolated ASD prior to Eisenmengerisation.⁶ In this instance, presence of a TAPVC facilitated right-to-left shunting as well as chronic hypoxia—important prerequisites in the pathogenesis of cerebral abscess.

The delayed presentation in adulthood observed in our patient is an unusual and instructive feature of this case report. TAPVC typically becomes symptomatic in early infancy and untreated, carries a grim prognosis with a mortality rate approaching 80% by the age of one.⁷ Survival into adulthood

with unrepaired TAPVC is infrequent and usually manifests with dyspnoea, atypical chest pain and atrial arrhythmias.^{8–9} Nevertheless, presentation at an advanced age is possible with TAPVC, and this disorder should be included among the differential diagnoses of young adults with cyanotic heart disease.

In summary, our case demonstrates two distinct and extreme aspects of TAPVC—primary presentation with a severe neurological complication, and asymptomatic survival into adulthood without corrective surgery.

Learning points

- ▶ Cerebral abscess is a potentially devastating complication of any cyanotic congenital heart disease including total anomalous pulmonary venous connection (TAPVC).
- ▶ Survival into adulthood with minimal or no symptoms is possible with TAPVC.
- ▶ Congenital heart disease should be excluded in all patients presenting with cerebral abscess regardless of age and cardiac symptomatology.

Contributors GV and KN were involved in data collection, writing, review of literature. GV and RKS were involved in the clinical care, concept, review and editing of the manuscript. KN was involved in acquisition of echo images, annotation and review as well as editing the manuscript.

Competing interests None.

Patient consent Obtained.

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