Case report

Portal-systemic encephalopathy after Fontan-type operation in patient with polysplenia syndrome

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Abstract

An 18-year-old patient, who had polysplenia and single ventricle, presented with altered mental status 9 years after a Fontan-type operation and pacemaker implantation. He underwent replacement of common atrioventricular valve and aortic valve plasty 1 year previously and has been placed on multiple medications including beta-blocker for his poor ventricular function. Blood chemistry revealed hyperammonemia of 2420 μg/l as a cause of this altered mental status disturbance. Superior mesenteric arteriography revealed large portal-systemic shunts in venous phase as a cause of hyperammonemia. To control blood ammonia level, we placed him on low protein diet, oral polymixin B, and lactulose instead of closing shunt with device. This case illustrates that portal-systemic shunt may result in hyperammonemia leading to altered mental status long after a Fontan-type operation.

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1. Introduction

Many cyanotic patients with heterotaxy and single ventricular physiology have been palliated by a Fontan-type operation. In these patients, multiple veno-venous collateral vessels frequently develop and can cause serious problems after a Fontan-type operation [1]. However, it is still uncommon to see portal-systemic encephalopathy [2] after a Fontan-type operation. We describe a patient who suffered from altered mental status due to hyperammonemia caused by a large portal-systemic shunt, probably associated with liver cirrhosis, 9 years after a Fontan-type operation.

2. Case report

An 18-year-old young man who had a Fontan-type operation for single ventricular physiology was sent to our hospital because of altered mental status after a short course of vomiting and anorexia.

His medical history includes four surgical and one catheter interventions. After birth, he had been relatively well with percutaneous arterial saturation of low 70% with significant pulmonary stenosis. At 5 years of age, he underwent cardiac catheterization that revealed dextrocardia, atrioventricular septal defect, double outlet right ventricle, L-malposition of the great arteries, pulmonary valve stenosis, left-sided inferior caval vein, and bilateral superior caval vein, suggesting asplenia and well developed pulmonary vasculature (pulmonary arterial index = 455 mm/M²) with low pulmonary vascular resistance of 1.2 Wood U M². He successfully underwent bilateral bidirectional Glenn procedure with uneventful postoperative course.

At 9 years of age, however, he showed significant cyanosis and underwent coil embolization ofazygous vein that increased percutaneous arterial saturation from 68 to 78%. At that time, he showed well developed pulmonary vasculature (pulmonary arterial index = 273 mm/M²) with low pulmonary artery pressure of 8 mmHg, and moderate degree of atrioventricular valve regurgitation. He successfully underwent total cavo-pulmonary connection using 20 mm extracardiac conduit with a boots shaped bottom incorporating left-sided inferior caval vein and isolated hepatic vein, plasticity of the common atrioventricular valve using edge to edge method creating double orifice and DuVega type annuloplasty, and received the diagnosis of polysplenia because of the morphology of bronchi and characteristics of the electrocardiogram. Postoperative course was complicated by sinus node dysfunction requiring pacemaker implantation within a month. Since then he had been doing relatively well in
functional class II of New York Heart Association until 16 years of age when he showed worsening common atrioventricular valve regurgitation.

At 17 years of age, he underwent replacement of common atrioventricular valve using 34 mm Saint-Jude medical valve and again redo aortic valve plasty by plicating each commissure. Postoperative cardiac catheterization revealed mean pulmonary arterial pressure of 16 mmHg and arterial oxygen saturation of 93% but significantly impaired cardiac function, with a ventricular ejection fraction of 33%. Despite multiple medications including oral diuretics and angiotensin converting enzyme inhibitor, he kept showing ventricular dysfunction and he has been placed on oral beta-blocker treatment since 6 months ago.

When he presented to us, he was drowsy and disoriented and blood chemistry revealed significantly elevated ammonia of 2420 μg/l (normal 120–660 μg/l) and total bile acid of 140 μmol/l (normal 0–10 μmol) with mildly elevated transaminases (Table 1). Hepatic ultrasound revealed mild surface nodularity with mildly dilated portal venous system. The scintigraphy using 99m technetium galactosyl human serum albumin showed significantly decreased liver function with receptor index of 0.81 [median (lower and upper quartiles) of healthy volunteers = 0.95 (0.94 and 0.96)] indicating liver cirrhosis [3]. Superior mesenteric and splenic angiography allowed us to make a diagnosis of polysplenia and showed major portal-systemic shunts in venous phase (Fig. 1A–D). Because he has been in chronic cardiac failure, simple closure of this shunt might induce portal vein hypertension or develop another shunt vessel. Therefore, we decided to place him on low protein diet, oral polymixin B, and lactulose to control plasma ammonia level. Since then he has not experienced any episodes of altered mental status so far.

3. Comments

Our patient illustrates that cyanotic patients with polysplenia and single ventricle may suffer from portal-systemic encephalopathy, possibly associated with latent liver cirrhosis, long after a Fontan-type operation. Though it is still unknown how many patients with cyanotic congenital heart diseases have these portal-systemic shunts, significant numbers of patients with congenital heart diseases, especially with polysplenia, are expected to have these portal-systemic shunts. Murray et al. [4] reported two patients who had heterotaxy and polysplenia with interrupted inferior caval vein complicated by extrahepatic portal-systemic shunts. They also searched literature and reported that, among 61 patients with congenital extrahepatic portal-systemic shunts, 31% had congenital heart diseases and 15% had polysplenia. However, they did not describe specific cardiac pathology that may associate with these portal-systemic shunts and did not mention the frequency of these shunts in patients with asplenia.

On the other hand, it is sometimes difficult to make a diagnosis of either polysplenia or asplenia just by looking at the combination of cardiovascular anomalies [5]. Though interrupted inferior caval vein may predispose these anomalous veno-venous shunts [6], not all patients with polysplenia present with interrupted inferior caval vein like...
our patient. Therefore, we must keep these shunts in mind in any patients with heterotaxy syndrome.

Also, it is not clear how often these patients with portal-systemic shunt present with encephalopathy due to hyperammonemia. Ikeda et al. [7] observed an interesting phenomenon of portal-systemic shunt in a patient with polysplenia and single ventricular physiology. Based on angiograms, they reported that the shunt flowed from systemic vein to portal vein before a Fontan-type operation, but it reversed after a Fontan operation. Though their patient did not show hyperammonemia, the patient is at risk to develop hyperammonemia depending on the portal venous pressure. In our patient, certainly latent liver cirrhosis might attribute this hyperammonemia by way of increasing portal vein pressure directing portal venous blood to inferior caval vein. Though there is still controversy about whether we should close these portal-systemic shunts before a Fontan-type operation, certainly we need to keep in mind that these portal-systemic shunts can cause altered mental status long after a Fontan-type operation.

References