Persistent Primitive Hypoglossal Artery with Atrial Septal Defect

Key words: persistent primitive hypoglossal artery, atrial septal defect, congenital disorder, embryology

Introduction

Persistent primitive hypoglossal artery (PPHA) is a rare anomaly of brain arteries, and one of the remnants of embryonal circulation. Atrial septal defect (ASD) is also a remnant of embryonal circulation. We present a patient with PPHA and ASD, and propose new insights.

Case Report

A 66-year-old man was referred to our department for investigation prior to cardiac surgery on ASD. He had no complaints. On neurological examination, there were no abnormal symptoms. ASD was reconfirmed by ultrasonic cardio-graphy and cardiac catheterization. Cardiac catheterization showed that the pulmonary-to-systemic flow ratio (Qp/Qs) was 2.49 and surgery was recommended.

Because magnetic resonance angiography (MRA) showed an abnormal artery, catheter angiography was performed. Angiography revealed the abnormal artery left right ICA to be a large extracranial branch and the basilar trunk originated from the artery. CT revealed the right anterior condyloid foramen to be enlarged (Fig. 1). Therefore, the abnormal artery was thought to be PPHA.

Cardiac surgery was successfully performed.

Discussion

Diagnosis of PPHA is based on the following criteria described by Lie and revised by Brismar (1).

- The PPHA leaves the internal carotid artery as a large extracranial branch.
- It passes through the anterior condyloid foramen.
- The basilar trunk originates from the PPHA.

In a review of the literature, 47.5% of cases of PPHA seemed to have some association with clinical features such as subarachnoid haemorrhage, cerebral aneurysms and arteriovenous malformation (2).

When the embryo is 4 mm in length, the following anastomotic vessels are between the primitive ICA and longitudinal neural artery: the primitive trigeminal artery (PTA), primitive hypoglossal artery (PHA) and primitive otic artery (POA). When the embryo is 5 to 6 mm long, the carotid basilar anastomosis allows these vessels to disappear. The PTA involutes by the 12 mm stage, the PHA by the 8 mm stage and the POA by the 5 to 6 mm stage.

If the embryo has inappropriate development of the vertebral artery or inappropriate fusion between the vertebral artery and basilar artery, circulation of the posterior brain is supplied mainly by persistent anastomotic vessels. Therefore, the PPHA must be of benefit to such an embryo.

ASD is also one of the remnants of embryonal circulation.

Figure 1. Catheter angiography showed right PPHA (arrow). CT showed that the right anterior condyloid foramen was enlarged.
ASD is due to excessive absorption of the septum primum or inappropriate growth of the septum secundum. During the 5 to 6 mm stages the septum primum begins to grow. Before the septum primum reaches the atrio-ventricular cushions, a perforation appears in part of the septum. At the same time when the embryo is 9 mm long, the septum secundum begins to grow.

If the posterior circulation of the brain decreases in the embryo, it may be rational to increase the stream of blood passing from the right atrium to the left atrium because brain circulation is dependent on this stream. So inappropriate growth of the atrial septum may be of benefit to such an embryo.

Only one case of coexisting PPHA and ASD has previously been reported and this case also had congenital intrahepatic shunts (3). Because both PPHA and ASD are rare, the relationship between PPHA and ASD remains unclear. As indicated above, the coexistence of PPHA and ASD may be remnants of a rational embryonic circulation in an embryo with insufficient vertebral circulation.

However, cases of ASD and other persistent primitive arteries such as persistent PTA have not been reported. It may be because the time that the septum grows is relatively distant from the times that primitive arteries other than the PHA regress.

This case report is important because such a case is rare and the above discussion, although controversial, is quite unique. This case may serve as a basis for the study of important embryological processes.

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