

Multiple Vascular Accidents Including Rupture of a Sinus of Valsalva Aneurysm, a Minor Ischemic Stroke and Intracranial Arterial Anomaly in a Patient with Systemic Congenital Abnormalities: A Case Report

Masataka Nakajima Arata Abe Yasuhiro Nishiyama Mina Harada-Abe
Akihito Kutsuna Yuya Goto Seiji Okubo Masahiro Mishina Ken-
ichiro Katsura Yasuo Katayama

Division of Neurology, Department of Internal Medicine, Nippon Medical School,
Tokyo, Japan

Key Words

Ischemic stroke · Hypoplasia of internal carotid artery · Low-lying carotid bifurcation ·
Rupture of the sinus of Valsalva · Intellectual disability

Abstract

A 39-year-old man with a history of rupture of a sinus of Valsalva aneurysm experienced an ischemic stroke. Although the patient presented left-sided hemiparesis for a week, no abnormal signals were indicated on diffusion-weighted imaging with repeated magnetic resonance scans. Carotid ultrasound and cerebral angiography were conducted, and they revealed hypoplasia of the left internal carotid artery with a low-lying carotid bifurcation at the level of the C6 vertebra. In addition, he was diagnosed with intellectual disabilities, evaluated by the Wechsler Adult Intelligence Scale-III, and congenital velopharyngeal insufficiency. We herein present the first report of a patient with cardio-cerebrovascular abnormalities, intellectual disabilities, and an otorhinolaryngological abnormality.

© 2013 S. Karger AG, Basel

Yasuhiro Nishiyama, MD, PhD
Division of Neurology, Department of Internal Medicine
Nippon Medical School
1-1-5 Sendagi, Bunkyo-ku, Tokyo 113-8602 (Japan))
E-Mail nomo16@nms.ac.jp

Introduction

Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA), which are rare congenital abnormalities, occur in less than 0.01% of the population [1, 2]. These are usually asymptomatic because of collateral circulation through the circle of Willis. The association between hypoplasia of the ICA and the occurrence of cerebral ischemia is unclear. This report describes a unique patient with a symptomatic ICA abnormality, some other congenital malformations that include a sinus of Valsalva aneurysm (SVA), and intellectual disabilities.

Case Report

A 39-year-old man was admitted to our hospital because of a sudden onset of hemiparesis on his left side with dysarthria. He had a history of a SVA rupture that was surgically treated at the age of 24 (fig. 1). During the surgery, there were no specific findings of cystic medial necrosis or crystalline lens subluxation due to the Marfan syndrome. In his infancy, there were no developmental abnormalities. In his school records, the subject had poor grades during childhood and adolescence. His family history over 3 generations was unremarkable in terms of neurological, psychological, and cardiac diseases. During the patient's examination, his body temperature was 37.0°C, blood pressure was 184/104 mm Hg, pulse was 83 beats per minute, respiratory rate was 20 breaths per minute, and oxygen saturation was 97% (while he was breathing ambient air). At the time of hospitalization, the patient presented with an acute stroke and the symptoms included left-sided weakness and dysarthria; these neurological symptoms lasted for a week after admission. We assessed the risk of juvenile ischemic stroke in a laboratory study, but high-risk factors that are linked to juvenile cerebral ischemia, such as collagen diseases and vasculitides, were not found. An electrocardiogram revealed a sinus rhythm within the normal range. Although we performed magnetic resonance imaging (MRI) with a 1.5-tesla unit on the first day and with a 3-tesla unit 2 weeks later, acute cerebral infarction was not detected (fig. 2a). Routine sonographic evaluation of the carotid arteries demonstrated a diffuse narrowing of the left ICA and a low-lying carotid bifurcation. Thereby, cerebral angiography was conducted, and it showed hypoplasia of the left ICA and a low-lying carotid bifurcation at the level of the C6 vertebra (fig. 3a). The A1 segment of the bilateral anterior cerebral arteries (ACAs) was united and formed an azygos type (fig. 3b). The common ACA duct and left posterior cerebral artery were mainly supplied by the right ICA. Aortography and 4 vessel studies (bilateral common carotid arteries and vertebral arteries) were conducted, and there was no embolic source, such as ulceration of the arterial surface or arterial dissection, or significant partial arterial stenosis. No right-to-left shunt was found in transesophageal echocardiography. ¹²³I iofetamine single-photon emission computed tomography brain imaging was performed, and no regional reductions or abnormal accumulations of tracer uptake into the brain were observed. The patient's intelligence quotient was evaluated with the Wechsler Adult Intelligence Scale-III [3]; his mental faculties were found to be at a significantly low level of functioning (table 1). Facial manifestations were unremarkable, but he showed symptoms of nasal speech and nasal air emission while talking. In the otorhinolaryngological examinations, velopharyngeal insufficiency was revealed by laryngoscopy. The patient was discharged after 3 weeks; MRI could not detect any significant abnormal findings such as infarction, hemorrhage, or malformation. We decided that the patient should continue with an antithrombotic therapy after assessing normal treatment for cerebral ischemia. The

patient is now taking warfarin; the target range for the prothrombin time/international normalized ratio is 2.0–3.0.

Discussion

The present case experienced 2 critical cardio-cerebrovascular diseases in his history of cerebral ischemic stroke and rupture of the SVA. He had also congenital cerebral arterial anomalies, including hypoplasia of the ICA, a low-lying carotid bifurcation, and an azygos ACA. In addition to the vascular abnormalities, he had intellectual disabilities. Patients with ICA hypoplasia generally have no neurological symptoms; the cerebral blood flow system can be aided by collateral blood flow such as that from the circle of Willis or anastomosis. The patient described herein has experienced a sudden onset of cerebral ischemia, but no evidence was indicated in the MRI examinations, blood examinations, cerebral angiography, or transesophageal echocardiography that was conducted to detect the embolic source of the thrombus. Therefore, the cause of his neurological symptoms was unclear. The exact mechanisms leading to agenesis or hypoplasia of the ICA remain unknown, but some investigators have suggested the occurrence of various mechanical disturbances during early development, such as pressure effects and excessive bending of the cephalic portion of the embryo to 1 side or the other. Azygos ACA, in which the 2 A1 segments of the ACA join to form a single trunk, is an uncommon variant that is seen in approximately 0.4–1% of the population [4]. SVA is also a rare congenital cardiac malformation. Rupture of the SVA is an uncommon disease that has been reported in 0.09% of the cases in one autopsy series. The incidence has been observed in 0.14–0.23% of Caucasians and in 0.46–3.5% of the Asian population [5–8]. In addition, acquired SVA occurs less frequently and is caused by conditions affecting the aortic wall, such as infection (syphilis, bacterial or fungal endocarditis, and tuberculosis), degenerative disease (atherosclerosis, connective tissue disorders, and cystic medial necrosis), or thoracic trauma [9, 10]. However, no evidence was found in our patient according to the pathological report of the aneurysm. An association between hypoplasia of the ICA and a low intelligence quotient has not been reported previously. There has been only 1 report of a patient with cerebral ischemia who had a hypoplastic ICA and a low-lying carotid bifurcation without other congenital vascular anomalies and intellectual disabilities [11]. In the present patient, we demonstrated that the patient experienced a cerebral ischemic attack with a hypoplastic ICA and a low-lying carotid bifurcation. Furthermore, he had also a past history of a SVA rupture, intellectual disabilities, and velopharyngeal insufficiency. Although these multiple manifestations might have been coincidental, this is the first report that describes a patient with cardio-cerebrovascular abnormalities along with intellectual disabilities. Further studies are required to characterize this constellation of findings in the patient.

References

- 1 Afifi AK, Godersky JC, Menezes A, Smoker WR, Bell WE, Jacoby CG: Cerebral hemiatrophy, hypoplasia of internal carotid artery, and intracranial aneurysm. A rare association occurring in an infant. *Arch Neurol* 1987;44:232–235.
- 2 Chen CJ, Chen ST, Hsieh FY, Wang LJ, Wong YC: Hypoplasia of the internal carotid artery with intercavernous anastomosis. *Neuroradiology* 1998;40:252–254.
- 3 Wechsler D: Wechsler Adult Intelligence Scale, ed 3. San Antonio, The Psychological Corporation, 1997.
- 4 Huh JS, Park SK, Shin JJ, Kim TH: Saccular aneurysm of the azygos anterior cerebral artery: three case reports. *J Korean Neurosurg Soc* 2007;42:342–345.

Nakajima et al.: Multiple Vascular Accidents Including Rupture of a Sinus of Valsalva Aneurysm, a Minor Ischemic Stroke and Intracranial Arterial Anomaly in a Patient with Systemic Congenital Abnormalities: A Case Report

- 5 Ott DA: Aneurysm of the sinus of valsalva. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2006;165–176.
- 6 Ring WS: Congenital heart surgery nomenclature and database project: aortic aneurysm, sinus of valsalva aneurysm, and aortic dissection. *Ann Thorac Surg* 2000;69:S147–S163.
- 7 Regueiro Abel M, Penas Lado M, López Ciudad V, Castro Beiras A: Sinus of Valsalva aneurysm as a cause of acute myocardial infarction. *Rev Esp Cardiol* 2002;55:77–79.
- 8 Vautrin E, Barone-Rochette G, Baguet JP: Rupture of right sinus of Valsalva into right atrium: ultrasound, magnetic resonance, angiography and surgical imaging. *Arch Cardiovasc Dis* 2008;101:501–502.
- 9 Batiste C, Bansal RC, Razzouk AJ: Echocardiographic features of an unruptured mycotic aneurysm of the right aortic sinus of Valsalva. *J Am Soc Echocardiogr* 2004;17:474–477.
- 10 Greiss I, Ugolini P, Joyal M, Boucharad D, Mercier LA: Ruptured aneurysm of the left sinus of Valsalva discovered 41 years after a decelerational injury. *J Am Soc Echocardiogr* 2004;17:906–909.
- 11 Dawson AG, Wilson A, Marskova J, Murray AD, Reid JM, Kuhan G: Hypoplastic internal carotid artery stenosis with a low-lying carotid bifurcation causing cerebral ischemia. *J Vasc Surg* 2012;56:1416–1418.

Table 1. Summary of the WAIS III assessment

Test	Raw score	Level of functioning
Verbal IQ	63	Extremely low
Performance IQ	61	Extremely low
Full scale IQ	59	Extremely low
Verbal comprehension	71	Borderline
Perceptual organization	63	Extremely low
Working memory	60	Extremely low
Processing speed	84	Considered low average

WAIS = Wechsler Adult Intelligence Scale; IQ = intelligence quotient.

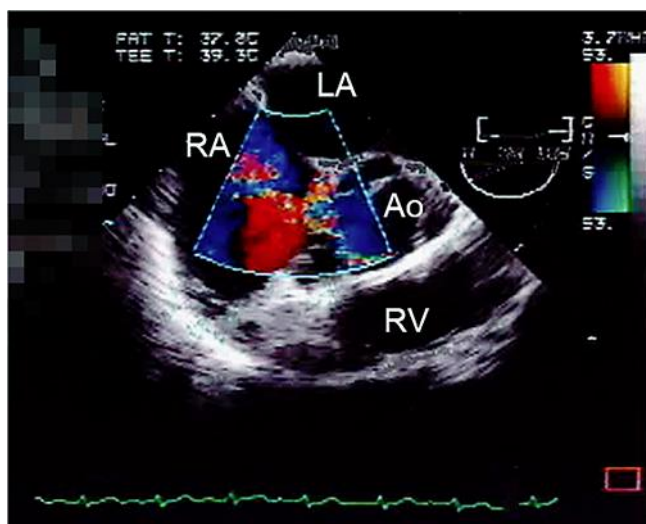


Fig. 1. Transesophageal echocardiography describing the right coronary sinus to the right ventricular shunt. This was performed before the surgery for the rupture of a congenital SVA.

Nakajima et al.: Multiple Vascular Accidents Including Rupture of a Sinus of Valsalva Aneurysm, a Minor Ischemic Stroke and Intracranial Arterial Anomaly in a Patient with Systemic Congenital Abnormalities: A Case Report

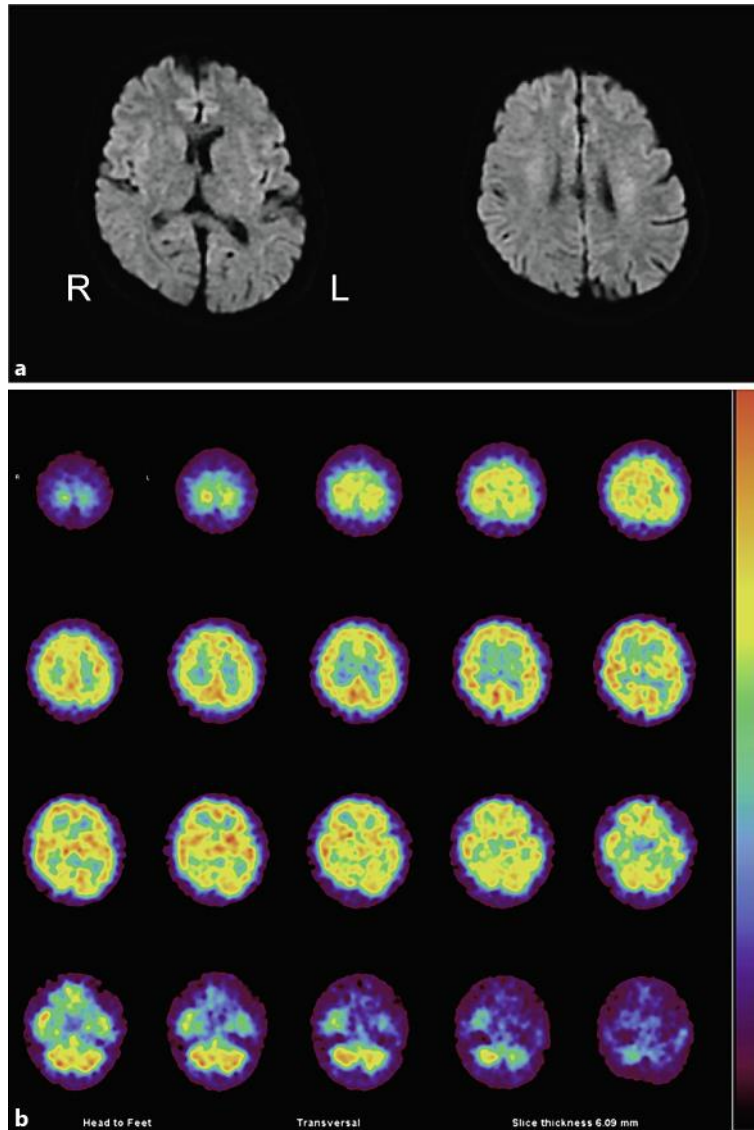


Fig. 2. MRI (a) did not detect a reasonable high-intensity area with diffusion-weighted imaging. In addition, ^{123}I iofetamine single-photon emission computed tomography brain imaging (b) did not detect any areas of abnormal blood flow.

Nakajima et al.: Multiple Vascular Accidents Including Rupture of a Sinus of Valsalva Aneurysm, a Minor Ischemic Stroke and Intracranial Arterial Anomaly in a Patient with Systemic Congenital Abnormalities: A Case Report

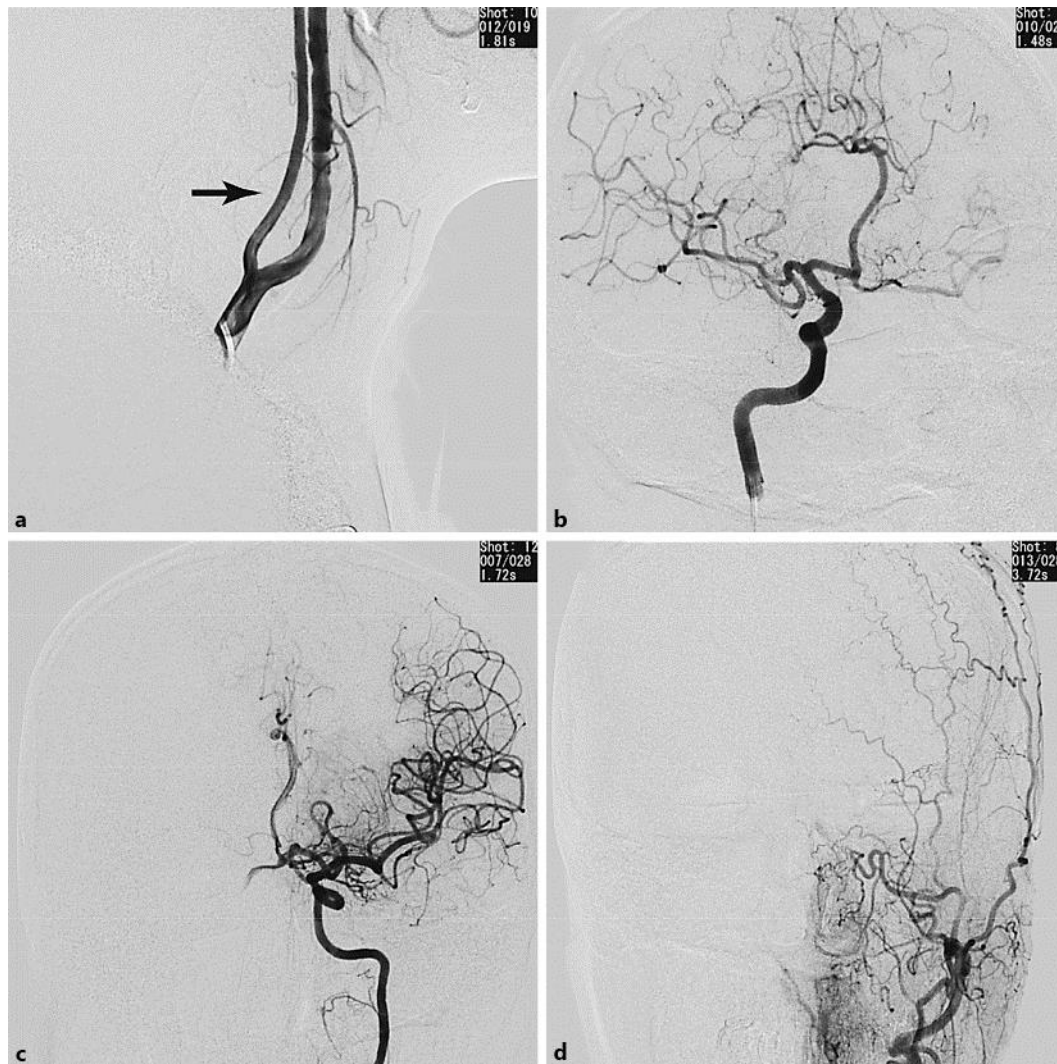


Fig. 3. Conventional cerebral angiography revealed that the left ICA (arrow) was hypoplastic and the carotid bifurcation was low-lying at the level of the C6 vertebra (a). The ACA was of the azygos type, and the left middle cerebral artery was visualized through the anterior communicating artery by the right conventional coronary angiography (b). The left ICA did not have stenosis (c). The left external carotid artery flow did not supply the ICA (d).