The clinical presentation and collateral pathway development of congenital absence of the internal carotid artery

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ABSTRACT

Objective: The objective of this study was to investigate the clinical presentation, risks, and collateral pathway development of the congenital absence of the internal carotid artery (ICA).

Methods: Sixty-four patients (10 new patients and 54 patients from the relevant literature) were studied. Data on demographic, clinical, and radiologic features were collected, followed by an analysis of the risks associated with ICA agenesis.

Results: There were 31 male and 33 female patients whose ages ranged from 5 months to 75 years, with a mean age of 31.1 years. The range of clinical symptoms recorded included transient ischemic attack (17 patients), subarachnoid hemorrhage (12 patients), developmental delay (13 patients), asymptomatic (8 patients), and other symptoms (15 patients). All 64 patients presented with absence of unilateral or bilateral ICAs, as measured by cervical computed tomography angiography or magnetic resonance angiography. The carotid canal was absent in all patients on computed tomography of the base of the skull, and abnormal development of collateral circulation pathways was observed. Five patients presented with basilar artery dilation on angiography. Aneurysms were observed in the angiography results from 16 patients. Ten patients presented with variations in the ophthalmic artery origin (the ophthalmic artery originated from the ipsilateral middle meningeal artery in six patients and from the ipsilateral middle cerebral artery in four patients).

Conclusions: From analysis of our 10 cases of ICA agenesis and our review of the relevant literature, we conclude that young patients with ICA agenesis may present with developmental delay, subarachnoid hemorrhage, or other developmental abnormalities, whereas older patients most commonly present with transient neurologic events. Complications of carotid agenesis are related to specific anatomic subtypes and the resulting collateral circulation development. (J Vasc Surg 2018;■:1-8.)

Keywords: Congenital; Internal carotid artery; Agenesis; Computed tomography angiography; Risks

Congenital absence of the internal carotid artery (ICA) is a rare anomaly.[1] The carotid canal, which is located in the temporal bone, is an important anatomic structure of the base of the skull. The carotid canal transmits the ICA, the sympathetic nerve plexus, and the internal carotid venous plexus, a venous network around the ICA connected with the cavernous sinus. Because of the close relationship between the carotid canal and the ICA during embryonic development, congenital absence of the ICA is often associated with carotid canal atresia and ICA-related structural abnormalities, such as variations in the origin of the ophthalmic artery, pituitary hypoplasia, sympathetic nerve dysplasia, and abnormal development of collateral circulation pathways. The clinical manifestations of this disease range from asymptomatic status to transient ischemic attack (TIA), subarachnoid hemorrhage (SAH), hypopituitarism, and developmental malformations in multiple organs. There are no summary reports of the relationships of embryonic development, radiologic findings, and clinical presentation in patients with ICA agenesis. The purpose of this study was to report 10 new cases of the absence of the ICA; to review the literature; to analyze the demographic features, clinical manifestations, and radiologic findings of these cases; and to assess the risks associated with ICA agenesis (eg, ischemia and aneurysms caused by variations in the origins of the ophthalmic artery or abnormal development of collateral circulation pathways).

METHODS

Patients. The study was approved by the Institutional Review Board at the hospital, and no consent was

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required of the patients because of the retrospective nature of the study. The medical records and radiologic data of 10 patients with congenital absence of the ICA, who were registered between 2002 and 2016, were reviewed retrospectively; these included 4 male patients and 6 female patients with ages ranging from 16 to 67 years, with a mean age of 42.3 years. The diagnosis of congenital absence of the ICA was based on carotid canal atresia on computed tomography (CT) of the base of the skull and nonvisualization of the atretic side of the ICA in angiographic findings.

Radiologic studies. All patients underwent CT angiography (CTA) using a spiral CT scanner at our institution. The CTA scanning parameters were 0.625-mm slice thickness, pitch ≤1, and 22- × 22-cm field of view. Four patients underwent conventional magnetic resonance imaging (MRI). The scanning equipment used in this study was a 1.5T superconductive MRI imaging system. The MRI scanning parameters were 3-mm slice thickness and 0- to 0.5-mm interval. The scanning position included an axial fast spin-echo T2-weighted sequence (3000-3500 ms/120-130 ms [repetition time/echo time]); sagittal fast spin-echo T1-weighted sequence (500-600 ms/10-15 ms [repetition time/echo time]); echo train length, 11 to 27; number of excitations, 2 to 4; 256 × 256 matrix; and 24 × 24-cm field of view.

Our goals in radiologic evaluation were to assess the carotid canal atresia on the CT scan of the skull base and to assess the type of collateral development based on the classification proposed by Lie[2] (Table I). Collateral circulation pathways in Lie types include ICA, external carotid artery, anterior cerebral artery (ACA), middle cerebral artery (MCA), anterior communicating artery (ACOM), and posterior communicating artery (PCOM). We assessed the diameter of the basilar artery on CTA scans using the method reported in the study by Smoker et al.[3] When the diameter of the basilar artery is >4.5 mm, patients are diagnosed with basilar artery dilation. We assessed the origin of the ophthalmic artery on the side of the affected ICA and the contralateral ICA using CTA scans. We measured the height of the pituitary gland in the sagittal fast spin-echo T1-weighted sequence. Patients with a pituitary gland height <3 mm, a posterior pituitary gland (bright spot) that disappeared on T1-weighted MRI, or pituitary stalk interruption were diagnosed with pituitary hypoplasia, according to the criteria reported in the study by Tsunoda et al.[4] CTA and MRI were used to assess the presence of other abnormalities in the brain or at the base of the skull.

RESULTS

Demographic, clinical, and radiologic findings from the 10 patients are summarized in Table II.

Clinical manifestations. Six patients presented with paroxysmal headache, vertigo, or general weakness due to TIA, and one patient had a history of SAH. One patient was asymptomatic. The remaining three patients were admitted for other causes. One patient had a recurrent history of otitis media, one patient presented with a sudden headache with vision loss with left parietal and occipital lobe infarctions detected by CTA, and one patient presented with a 2-week history of seizures.

ARTICLE HIGHLIGHTS

- **Type of Research:** Retrospective cohort study and review of the literature
- **Take Home Message:** In 64 patients with congenital absence of the internal carotid artery complications depended on age, anatomic subtypes, and collateral circulation.
- **Recommendation:** Data suggest that agenesis of the internal carotid artery is rare. In young patients, it is manifested by developmental delay or subarachnoid hemorrhage; in older patients, it usually is manifested with transient ischemic attack.

Radiologic findings. Four patients presented with right-sided ICA agenesis, whereas six patients presented with left-sided agenesis. One of the six patients also had right-sided ICA hypoplasia.

Nine patients with ICA agenesis were identified as type A (Fig 1). Collateral circulation to the MCA territory ipsilateral to the ICA agenesis was accomplished through the PCOM, and collateral circulation to the ACA territory ipsilateral to the ICA agenesis was accomplished through the ACOM. Another patient was identified as type E (Fig 2). Left-sided ICA agenesis with a contralateral hypoplastic ICA and bilateral MCAs were supplied by enlarged PCOMs. Five patients with ICA agenesis were diagnosed with basilar artery dilation. All patients showed variations in the origin of the ophthalmic artery on CTA. An atherosclerotic contralateral ICA was found in one patient. A dilated and tortuous contralateral ICA was diagnosed in one patient. Ophthalmic arteries originating from the ipsilateral middle meningeal artery were observed in six patients. The ophthalmic arteries of the remaining four patients originated from the ipsilateral MCA.

Aneurysms were observed in four patients, one with a clinoid segment of the ICA (A type) and three with PCOM aneurysms (E type in one patient, A type in two patients). Three of the four aneurysms presented as basilar artery dilations. The clinical manifestations of four patients with aneurysms included seizures (one patient), headache with general weakness (two patients), and ischemic events (one patient). Four patients underwent routine cranial MRI scanning. The posterior pituitary gland, pituitary stalk, and height of the pituitary gland were normal in these patients.