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Basal Encephalocele Associated With Hypoplasia of the Internal Carotid Artery

—Case Report—

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Abstract

A 22-year-old woman presented with a basal encephalocele associated with hypoplasia of the internal carotid artery (ICA) manifesting as a 6-year history of decreased vision in the right eye. She underwent encephalo-arterio-synangiosis under a diagnosis of hypoplasia of the ICA at age 6 years. Magnetic resonance imaging showed the encephalocele medial to the right temporal lobe. Frontotemporal craniotomy was performed for resection of the encephalocele and repair of the defect of the skull base. The pathogeneses of these developmental anomalies were probably related to developmental failure of the embryonic primordium during the 4th and 10th weeks.

Key words: basal encephalocele, hypoplasia of the internal carotid artery, transsphenoidal type

Introduction

Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are rare congenital anomalies with incidences of less than 0.01%.[4,10,16] Basal encephalocele is an even rarer congenital anomaly with an estimated incidence of 0.003% (1 in every 35,000 live births).[15] We describe a case of basal encephalocele associated with hypoplasia of the ICA, and discuss the relationship between these developmental anomalies.

Case Report

A 22-year-old woman presented with a 6-year history of decreased vision in the right eye. She had suffered intraventricular hemorrhage at age 6 years, and the diagnosis was hypoplasia of the ICA. Right common carotid angiography had demonstrated occlusion in the C2 portion of the ICA. The circulation to the distal area was maintained through a collateral arterial network. The cause of the hemorrhage was considered to be rupture of the collateral vessels. She underwent encephalo-arterio-synangiosis. She suffered meningitis and stayed in hospital for 2 weeks at age 21 years. Magnetic resonance (MR) imaging showed an encephalocele with 2 cm diameter medial to the right temporal lobe (Fig. 1 *left*). She was successfully treated with antibiotics, and followed up in the outpatient clinic of our hospital.

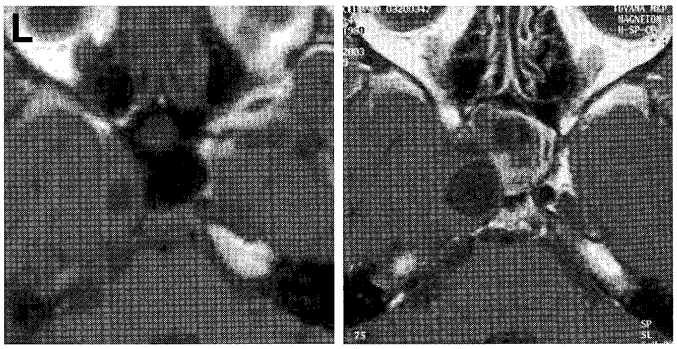

Fig. 1 T_1-weighted magnetic resonance images with contrast medium showing an encephalocele in the right middle fossa (*left*) and enlargement of the encephalocele (*right*).

On admission, ophthalmological examination showed visual impairment and temporal hemianopsia in the right eye. The right optic disc was atrophic. Sclerodermia in the right forehead was also recognized. Carotid angiography showed hypoplasia of the right ICA and abnormal collateral chan-

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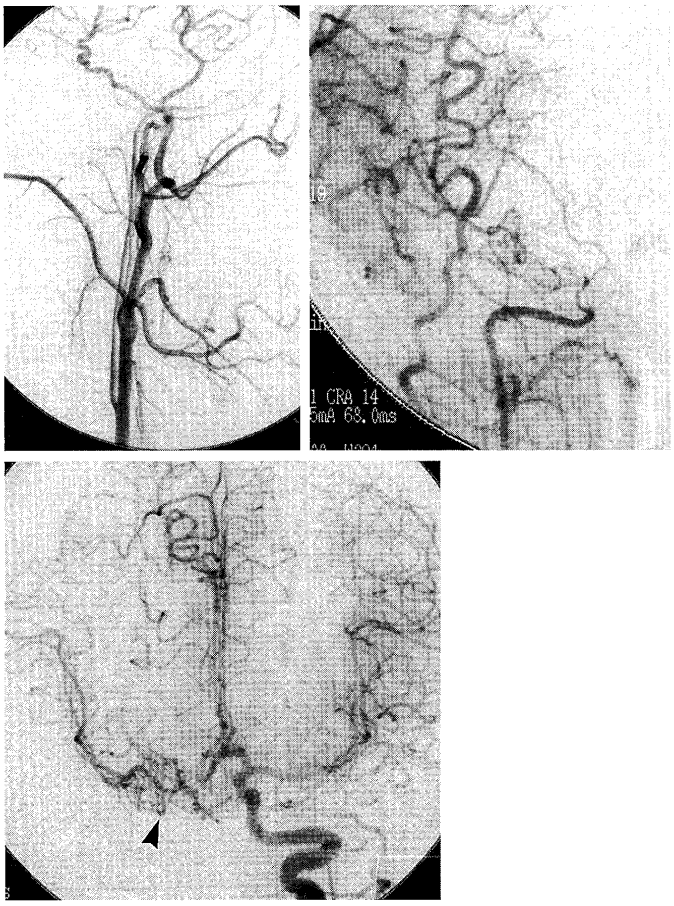

Fig. 2 Right carotid angiograms, lateral view (*upper left*), showing narrowing of the cervical internal carotid artery, and anterior view (*upper right*), showing absence of the intracranial internal carotid artery. Left carotid angiogram, anterior view (*lower*), showing abnormal collateral channels around the circle of Willis (*arrowhead*).

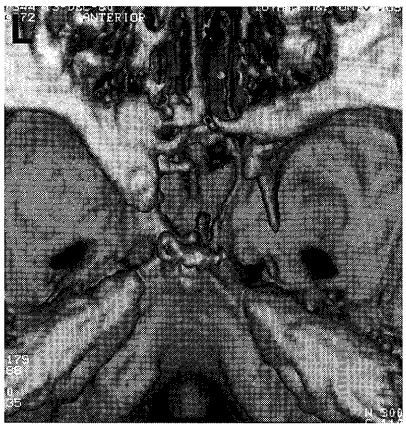

Fig. 3 Three-dimensional computed tomography scan of the skull base.

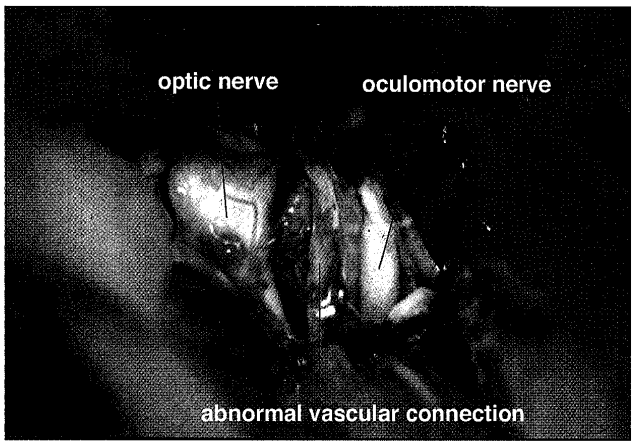

Fig. 4 Intraoperative photograph showing the encephalocele protruding into the sphenoid sinus and displacing the right optic nerve superiorly, and the middle cerebral artery and third cranial nerve. Abnormal vascular connection to the right middle cerebral artery (*arrowhead* in Fig. 2 *lower*) is confirmed.

nels of the circle of Willis (Fig. 2). Three-dimensional computed tomography (3D CT) showed absence of the bony carotid canal and the anterior clinoid process, a defect in the floor of the pituitary fossa, and protrusion of abnormal bone in the middle cranial fossa ipsilateral to the hypoplasia of the ICA (Fig. 3). MR imaging showed enlargement of the encephalocele with cystic component (Fig. 1 *right*).

Transsphenoidal resection of the encephalocele was first performed, but complete removal of the encephalocele and repair of the defects of the wall of the sphenoid sinus and the skull base were not possible. Therefore, frontotemporal craniotomy was performed for removal of the encephalocele and repair of the defect of the skull base (Fig. 4). Defects of the right anterior clinoid process and the wall of the sphenoid sinus were recognized. Absence of the intracranial ICA and the abnormal vascular connection to the right middle cerebral artery were confirmed. The encephalocele protruded to the sphenoid sinus through the bone defect and displaced the right optic nerve superiorly. The encephalocele was resected and the resulting defect was repaired with dura substitute. The postoperative course was uneventful. Her visual impairment remained unchanged.

Discussion

Basal encephalocele can be classified into transethmoidal, sphenoethmoidal, transsphenoidal and

frontosphenoidal based on the location of the defect in the skull base.[17] The present case was the transsphenoidal type, which is the least common, representing 5% of basal encephaloceles.[15] Cerebrospinal fluid rhinorrhea and recurrent meningitis are clinical characteristics of the transsphenoidal type, as seen in the present case.[9]

The various theories on the pathogenesis of the basal encephaloceles can be summarized as follows: Failure of the ethmoid plate to close around the olfactory nerves; localized increased intraventricular pressure; failure of neuroectodermal separation at the anterior neuropore from the surface ectoderm, preventing the subsequent development of the mesodermal elements; persistence of the craniopharyngeal canal; and developmental failure of ossification centers in the sphenoid bone.[9,15,17,19] Teratogenic factors are generally accepted to affect the normal development of the sphenoid bone that continues from the 5th week to the 10th week of gestation.[13,18] Chondrification of the sphenoid is first noted in the 5th week of gestation. The 7-week embryo has a completed cartilaginous sphenoid body originating from a basisphenoid center, and extends caudally to meet the basiocciput and rostrally to form the dorsum sellae. Continued rostral extension of the sphenoid surrounds the hypophysial canal and terminates at the nasal septum. The 10-week embryo shows closure of the hypophysial canal.

Three types of intracranial vascular anomalies are known to be associated with hypoplasia or aplasia of the ICA: Aneurysms related to the circle of Willis; arterial anomalies with abnormal collateral channels around the circle of Willis; and dilated vascular channels.[11] Associated anomalies with hypoplasia or aplasia of the ICA also include cerebral hemiatrophy,[1] Klippel-Trenaunay syndrome,[5] cardiac anomalies such as ventriculoseptal defect,[2,6] arachnoid cyst,[8,10] neurofibromatosis,[3] hemangioma in the tongue,[12] and nasopharyngeal angiofibroma.[10]

The present case of basal encephalocele was ipsilateral to the hypoplasia of the ICA. Aplasia of the ICA depends on abnormal regression of the first and third aortic arches during the formative stage of the vasculature during the 4th and 5th weeks of embryogenesis.[7,14] Normal development of the chondrocranium of the skull base starts from the 5th week and continues to the 10th week. In the present case, 3D CT demonstrated absence of the bony carotid canal and the anterior clinoid process, a defect in the floor of the pituitary fossa, and protrusion of abnormal bone in the middle cranial fossa. These multiple anomalies in the skull base and the hypoplasia of the ICA probably resulted from the developmental failure of the embryonic primordium during the 4th and 10th weeks. The pathogenesis of the basal encephalocele in the present case most likely involved the congenital defect of the skull base.

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