

noiditis after SAH was not exclusively associated with posterior circulation aneurysms. Spinal arachnoiditis after an MCA aneurysm was a complication in our patient 1.

Shunting of the syrinx to the subarachnoid, pleural, or peritoneal space is still widely accepted as the treatment of choice for patients with syringomyelia caused by arachnoiditis.<sup>5</sup> Shunting of the syrinx to the subarachnoid or peritoneal cavity was associated with a recurrence rate of 60%, whereas microsurgical dissection of the arachnoid scar and decompression of the subarachnoid space had a recurrence rate of 33%, with a mean follow-up period of 28 months.<sup>14</sup> Successful long-term management of the syrinx was associated with basal or focal spinal arachnoid scarring, no history of spinal meningitis, microsurgical dissection of the arachnoid scar and decompression of the subarachnoid space.<sup>14</sup> Cacciola et al.<sup>16</sup> reported 20 patients with syringomyelia refractory to CSF flow restoration who underwent a procedure for placement of a syringopleural shunt. Follow-up MRI revealed syrinx collapse in 17 patients and a marked shrinkage in two. They recommended that a syringopleural shunt should be the syrinx diversion procedure of choice.

#### 4. Conclusions

Two of 198 patients had syringomyelia following aneurysmal SAH. Thus, the rate of occurrence of syringomyelia was 1.0% following aneurysmal SAH. Spinal arachnoiditis and spinal syringomyelia are important chronic complications after SAH.

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## Acute infarction in a patient with agenesis of the internal carotid artery and transcavernous anastomosis

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#### ABSTRACT

A 70-year-old man presented with left-sided weakness and dysarthria. His brain MRI and magnetic resonance angiography demonstrated acute infarctions in the bilateral middle cerebral artery territories in a setting of agenesis of the right internal carotid artery (ICA) with transcavernous anastomosis, a rare developmental anomaly. In this setting, atherosclerotic disease in the unilateral common carotid artery or ICA can induce acute infarctions in the bilateral anterior circulation.

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## 1. Introduction

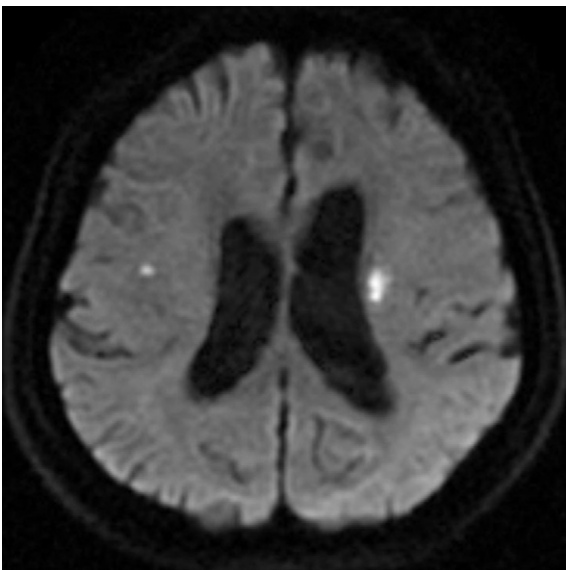
Agenesis, aplasia, and hypoplasia of the internal carotid artery (ICA) are rare congenital anomalies with a 0.01% of incidence.<sup>1</sup> Patients were mostly asymptomatic because of sufficient collateral circulation from the contralateral ICA or vertebrobasilar system via the circle of Willis. Much less commonly, the collateral circulation is supplied by a trans cavernous anastomotic vessel between the ICA. Only 17 patients with this anastomosis have been reported. We report a symptomatic patient with unilateral ICA agenesis or aplasia with trans cavernous anastomosis.

## 2. Case report

A 70-year-old man presented with left-sided weakness and dysarthria. He had a long history of hypertension and diabetes mellitus. A brain MRI was performed. Diffusion weighted imaging revealed multiple acute infarctions in both the middle cerebral artery (MCA) territories (Fig. 1). The infarction pattern was suggestive of embolic phenomena. On three-dimensional time-of-flight (3D-TOF) magnetic resonance angiography (MRA), an aberrant vessel was seen to arise from the right cavernous ICA (Fig. 2A) and supplied the left MCA without a left ICA. The A1 segment of the left anterior cerebral artery (ACA) could not be seen (Fig. 2B) and both ACA originated from the right ICA. On 3D-TOF MRA, the left common carotid artery (CCA) had a smaller diameter compared to its opposite and continued as a single vessel with the normal course of the external carotid artery (Fig. 2C). These findings thus demonstrated congenital absence of the unilateral ICA with trans cavernous anastomosis. Absence of the left carotid canal on a non-enhanced CT scan confirmed agenesis of the left ICA (Fig. 3). On 3D-TOF MRA, atherosclerotic change with mild stenosis was noted in the petrous portion of the right ICA, trans cavernous anastomotic vessel, and proximal portion of the right cervical ICA (Fig. 2B). Transesophageal echocardiography and holter monitoring failed to show an occult cardioembolic source.

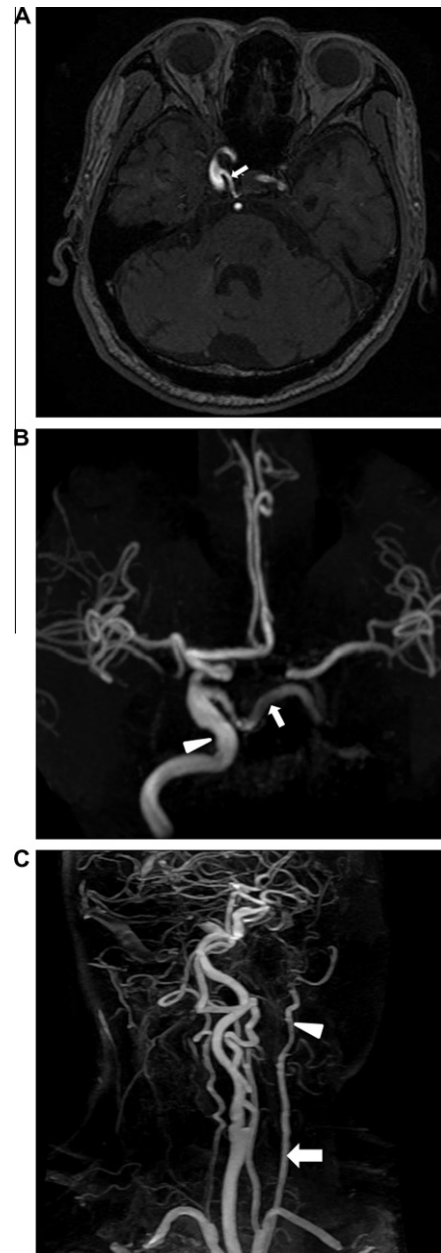
## 3. Discussion

The term “absence” can be referred to as agenesis or aplasia. Lie defined agenesis as a complete failure of development of an organ.<sup>2</sup>



**Fig. 1.** Axial diffusion weighted imaging showing acute infarctions in both middle cerebral artery territories.

The term “agenesis” can be used when the carotid canal is absent at the skull base. “Aplasia” was defined as a lack of organ development but existence of its precursor. “Hypoplasia” denotes incomplete development of an organ. Thus, hypoplasia of the ICA means that the vessel appears normal, but diminished in size. In our patient, the trans cavernous anastomosis arising from the right ICA confirmed a congenital absence of the left ICA. And absence of the carotid canal on the skull base differentiated agenesis from the aplasia.



**Fig. 2.** Three-dimensional time of flight (3D-TOF) magnetic resonance angiography (MRA): (A) axial source image showing an aberrant vessel (arrow) arising from the right cavernous internal carotid artery (ICA); and (B) the circle of Willis (showing only the anterior circulation) confirming absence of the left cavernous ICA – the anastomotic vessel (arrow) originating from the right cavernous ICA leads to a normally developed left middle cerebral artery – both anterior cerebral arteries originate from the right ICA; the left A1 is not seen. Atherosclerotic change with mild stenosis of the right petrous ICA (arrowhead) and trans cavernous anastomotic vessel are noted. (C) Oblique view of a carotid MRA showing a narrow left common carotid artery (arrow) and a normal external carotid artery (arrowhead) however, the ICA is not visualized.



**Fig. 3.** Axial non-enhanced CT scan of the skull base showing an absence of the left carotid canal and a normally developed right carotid canal (arrow).

Lie described six patterns of collateral circulation in agenesis, aplasia and hypoplasia.<sup>2</sup> Among them, unilateral ICA agenesis is classified into type A, type B and type D. In type A, the ipsilateral ACA is supplied through the anterior communicating artery (ACOM) and the ipsilateral MCA through a hypertrophied posterior communicating artery. In type B, the ipsilateral ACA and MCA are supplied through the patent ACOM. In type D, unilateral ICA agenesis is associated with transcavernous vessels connecting both cavernous segments of the ICA, similar to our patient. Type A is the most common and type D is the least common.

According to Padgett's analysis of the embryo, the ICA originates from the terminal segments of the dorsal aorta and the third aortic arch at the 3-mm embryonic stage, and the circle of Willis develops during the 7-mm to 24-mm stage of embryonic development.<sup>3</sup> Cali et al. documented that the pattern of collateral circulation in an absence of the ICA depends on the stage at which the disruption occurred.<sup>4</sup> Transcavernous anastomoses would dominate if the disruptions occurred before full development of the circle of Willis, and collateral flow via the circle of Willis would dominate, if the

disruption occurred after the 24-mm stage of embryonic development.<sup>4</sup>

Congenital absence of the ICA with transcavernous anastomoses has been reported only 17 times to our knowledge. Most patients had no symptoms: five patients had an intracranial aneurysm.<sup>5</sup> Altered hemodynamic force increases the incidence of intracranial aneurysms above the naturally occurring 2% to 4%.<sup>1</sup> Another three patients had cerebral infarctions<sup>6–8</sup>, but these infarcted territories and patterns were poorly described radiologically.

In Lie's original type D illustration, the transcavernous vessel supplied the ipsilateral MCA and ACA.<sup>2</sup> However, the most frequent occurrence, which included our patient, was hypoplasia or aplasia of the ipsilateral A1 segment of the ACA. Therefore, most of the ipsilateral MCA territory is supplied from the transcavernous anastomosis only without an alternative flow – the entire anterior circulation depends on a single carotid artery. Thus, atherosclerotic disease in the unilateral CCA or ICA can induce infarctions in the bilateral anterior circulation with further serious clinical implications. In addition, recognition of this anomaly can be important in preventing misdiagnosis of severe carotid stenosis or occlusion, and for planning neurosurgery by a transsphenoidal approach.

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