CONGENITAL ABSENCE OF THE INTERNAL CAROTID ARTERY ASSOCIATED WITH CEREBRAL HEMIATROPHY, ABSENCE OF THE EXTERNAL CAROTID ARTERY, AND PERSISTENCE OF THE STAPEDIAL ARTERY*

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CONGENITAL absence of the internal carotid artery has been rarely reported. Since Tode[44] first described absence of the internal carotid artery in 1787, the authors are aware of reports of 25 cases[4–7, 9, 12, 15, 16, 19, 22–24, 26, 30–33, 40, 41, 44–48, 50] of unilateral total absence, 6 cases[11, 14, 18, 25, 38, 49] of bilateral total absence, 5 cases[20, 22, 23, 39] of unilateral partial absence and 1 case[10] of bilateral partial absence. Of this number, only 9 cases[4, 6, 16, 22, 23, 26, 40, 41, 47] of unilateral and 1 case[25] of bilateral total absence were demonstrated angiographically. Each case of partial absence was demonstrated by angiography. We are presently reporting the sixth case of unilateral partial absence and the tenth case of unilateral total absence of the internal carotid artery demonstrated by angiography known to us.

The case of partial absence of the internal carotid artery presented here is unique in that we feel that it represents the first report of demonstration of a vascular etiology of primary cerebral hemiatrophy in a living patient. Carotid angiography demonstrated absence of the proximal left internal carotid artery with maintenance of the petrous (possibly distal cervical) and more distal portions of the internal carotid artery by means of collateral flow through a branch of the external carotid artery in a patient with a classical history of primary cerebral hemiatrophy.

Parker and Gaede[30] have recently reported the postmortem finding of agenesis of the left internal carotid, left middle cerebral, and left posterior communicating arteries combined with marked hypoplasia and poor myelination of the entire left pyramidal system in a patient with a typical history of primary cerebral hemiatrophy. This apparently was the first definitive demonstration of a vascular etiology of primary cerebral hemiatrophy.

The case of unilateral total absence of the internal carotid artery presented here is also unique particularly from an anatomic point of view. Associated contralateral absence of the external carotid artery with its usual branches arising from the internal carotid artery and persistence of the stapedial artery were demonstrated by angiography.

The embryology of the carotid arteries has been well described by Padget.[29] She states that the internal carotid arteries, which first appear in the 3 mm. (24 days) stage, are formed from the terminal segments of the paired dorsal aortae and from the third branchial arches. At the 4 mm. (28 days) stage, anterior (primordial anterior cerebral, middle cerebral and anterior choroidal arteries) and posterior branches are formed. The primordial stem of the external carotid artery is the proximal aspect of the ventral pharyngeal artery which is first seen at the 5–6 mm. (29 days) stage. Definitive branches of the external carotid artery (thyroid and lingual branches) are first seen during the 12–14 mm. (35 days) stage as the proximal part of the ventral pharyngeal artery. The common carotid artery is also seen during the 12–14 mm. stage as obliteration of the

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segment of paired aortae between the third and fourth aortic arches approaches. The appearance of the definitive common carotid artery occurs in the 16–18 mm. (40 days) stage.

Padget[29] states that anastomosis between the primitive hyoid artery, a branch of the internal carotid artery, and distal end of the ventral pharyngeal artery is present during the 12–14 mm. stage. Conceivably, an insult which results in loss of patency of the proximal internal carotid artery during this stage could prevent involution of the primitive anastomosis, thereby allowing persistence of patency of the distal internal carotid artery.

The nature of the sequence of events which lead to total absence of the internal carotid artery is unknown. However, Keen[18] suggests that unilateral absence of the internal carotid artery may be due to mechanical causes in early development such as pressure effects, excessive bending of the cephalic end of the embryo to one side or the other, effects of amniotic adhesions, etc.

Congenital absence of the external carotid artery with its usual branches arising from the internal carotid artery may be explained by either failure of the stem of the external carotid artery to develop or involution of the stem of the external carotid artery following annexation by the internal carotid artery of all external carotid branches.

REPORT OF CASES

Case 1. DW. This 41 year old white male was admitted to the Los Angeles County-University of Southern California Medical Center for evaluation of generalized central nervous system deterioration. His past history obtained from older members of his family was remarkable in that a seizure disorder, marked right hemiparesis and mental retardation had been present since birth. In spite of his handicap, he had been able to care for himself until about 3 months prior to admission to this center at which time he was institutionalized at a state hospital.

Because of increasing somnolence, he was admitted to this center. Examination at that time revealed a lethargic patient with marked right hemiparesis and atrophic musculature on the right side of his body. Plain skull film roentgenogram (Fig. 1) revealed the typical findings of cerebral hemiatrophy as described by Dyke *et al.*[8] A brain scan indicated suggestive increased uptake in the right hemisphere. Right common carotid angiography (not shown) on February 25, 1971 demonstrated a very large right internal carotid artery, right-to-left shift of the midline cerebral vessels, spontaneous opacification of the left anterior and middle cerebral arteries, and left-sided ventricular dilatation according to the appearance of the left thalamo-striate vein. These findings were felt to represent the result of left cerebral hemiatrophy. However, in view of the equivocal brain scan, serial scanning was recommended. The patient was then transferred to an affiliated neurologic disease chronic care and rehabilitation hospital.

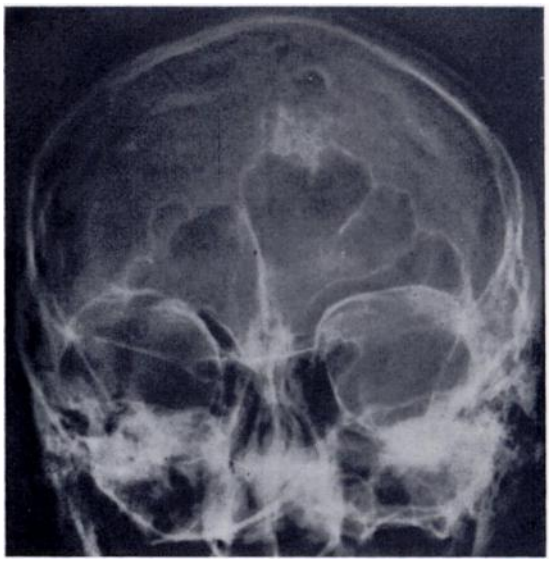

Fig. 1. Case 1. Plain skull roentgenogram demonstrates left-sided hypertrophy of the following: frontal and ethmoidal sinuses; calvarium; temporal bone air cells; and anterior clinoid process. Also seen are elevation of the left sphenoidal ridge and left side of planum sphenoidale; and tilting of the crista galli to the left.

However, because of continued deterioration of his neurologic status (increase in stupor and onset of progressive left hemiparesis), he was readmitted to LAC-USC Medical Center for re-evaluation. At this time lumbar puncture revealed an increase in both the pressure and

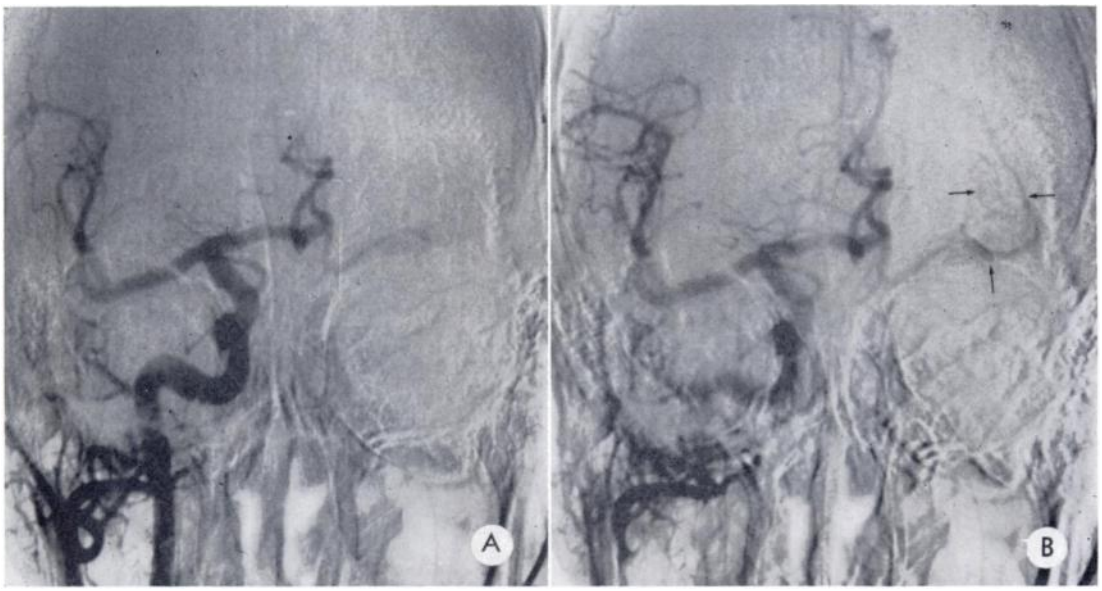

Fig. 2. Case 1. (*A*) Right common carotid arteriogram demonstrates large right internal carotid, right anterior cerebral, and anterior communicating arteries and spontaneous opacification of a smaller proximal left anterior cerebral artery. (*B*) Exposure made ½ second after *A* demonstrates right-to-left shift of anterior cerebral artery branches and spontaneous opacification of branches of the left middle cerebral artery (small arrows).

protein content of the cerebrospinal fluid. A repeat brain scan revealed a large discrete focus of increased uptake in the right hemisphere. Bilateral carotid and left brachial cerebral angiography (Fig. 2 *B*) on August 8, 1971 revealed a marked increase in the right-to-left shift of the midline vessels. This occurred since the angiogram of 6 months earlier, without discrete localization of a mass and absence of the left internal carotid artery from the usual region of its origin to the vicinity of the junction of the cervical and petrous portions where it was noted to fill in an antegrade fashion through the ascending pharyngeal branch of the external carotid artery. Combined ventriculography and pneumoencephalography disclosed the presence of a large suprasylvian mass compressing the right lateral

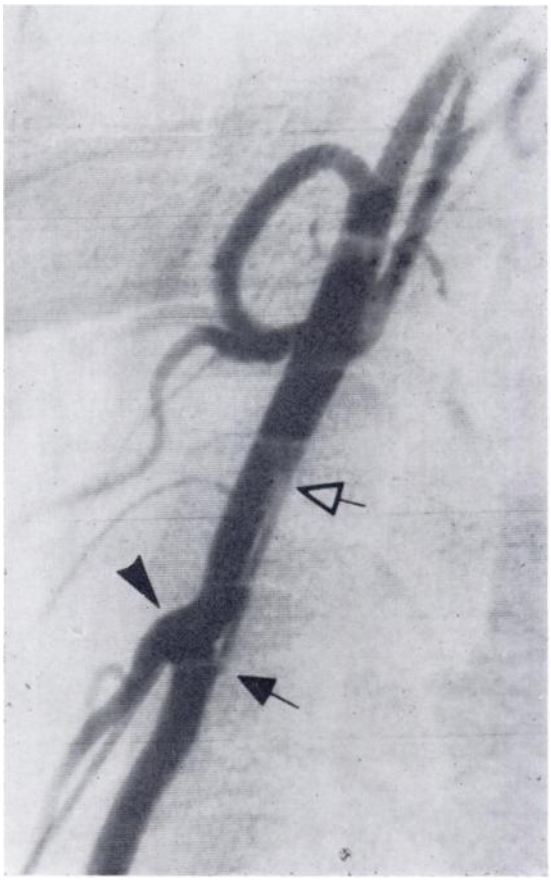

Fig. 3. Case 1. Left "common" carotid arteriogram demonstrates the origin (closed arrow) of a small branch arising from the posterior surface of the "common" carotid artery opposite the superior thyroid artery (arrowhead) and rejoining (open arrow) the external carotid artery a few centimeters higher. This probably represents either a simple fenestration or an aborted proximal internal carotid artery.

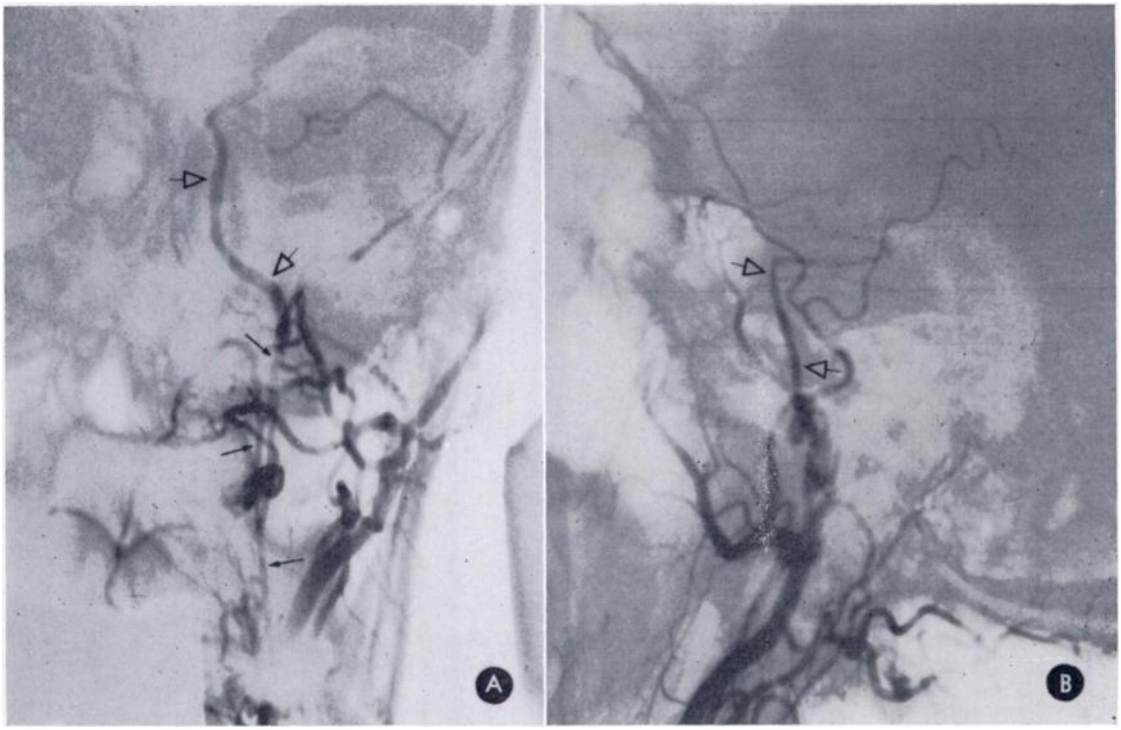

Fig. 4. Case 1. (*A* and *B*) Left carotid arteriogram demonstrates the petrous and cavernous portions of the left internal carotid artery (open arrows) being filled *via* the ascending pharyngeal artery (small closed arrows). Note opacification of the ophthalmic artery *via* both the internal carotid and middle meningeal arteries.

ventricle. At surgery a large malignant ependymoma was resected. The postoperative course was unremarkable and the patient was returned to the affiliated chronic care neurologic disease hospital for rehabilitation.

Case II. E.P. This 66 year old black female was admitted to the LAC-USC Medical Center on April 10, 1969 following a fall from which she sustained injuries to the left side of her head and face. Her past history was unremarkable except for alcoholism.

At the time of admission, she was dysphasic and exhibited a left VII nerve weakness. The day after admission, she began to have left facial seizures. Two days later, right body seizures commenced which progressed to status epilepticus in about 24 hours. At this time, cerebral angiography was requested to exclude the presence of a subdural hematoma.

Left carotid angiography (Fig. 7; and 8, *A* and *B*) revealed congenital absence of the left internal carotid artery, origin of the left ophthalmic artery from the left middle meningeal artery, and a small, probably traumatic, aneurysm of the left superficial temporal artery. Right carotid angiography (Fig. 10, *A C*) demonstrated small aneurysms of the cavernous portion of the right internal carotid artery, visualization of the anterior cerebral artery system on the right side only, absence of the external carotid artery with its usual branches arising from the internal carotid artery, petrous internal carotid artery origin of the middle meningeal artery (persistent stapedial artery), and an unusual course of the petrous internal carotid artery. Left brachial cerebral angiography (Fig. 9, *A* and *B*) demonstrated a large arterial stem on the left at the termination of the basilar artery which gave origin to the left posterior cerebral artery and continued as a markedly enlarged left posterior communicating artery giving rise to the left middle and anterior cerebral arteries or the left middle cerebral artery alone, which in turn gave rise to the left anterior cerebral artery. Also noted were a large avascular mantle over the left cerebral hemisphere having the appearance of a subacute subdural hematoma and a probable aneurysm (appropriate views were not taken

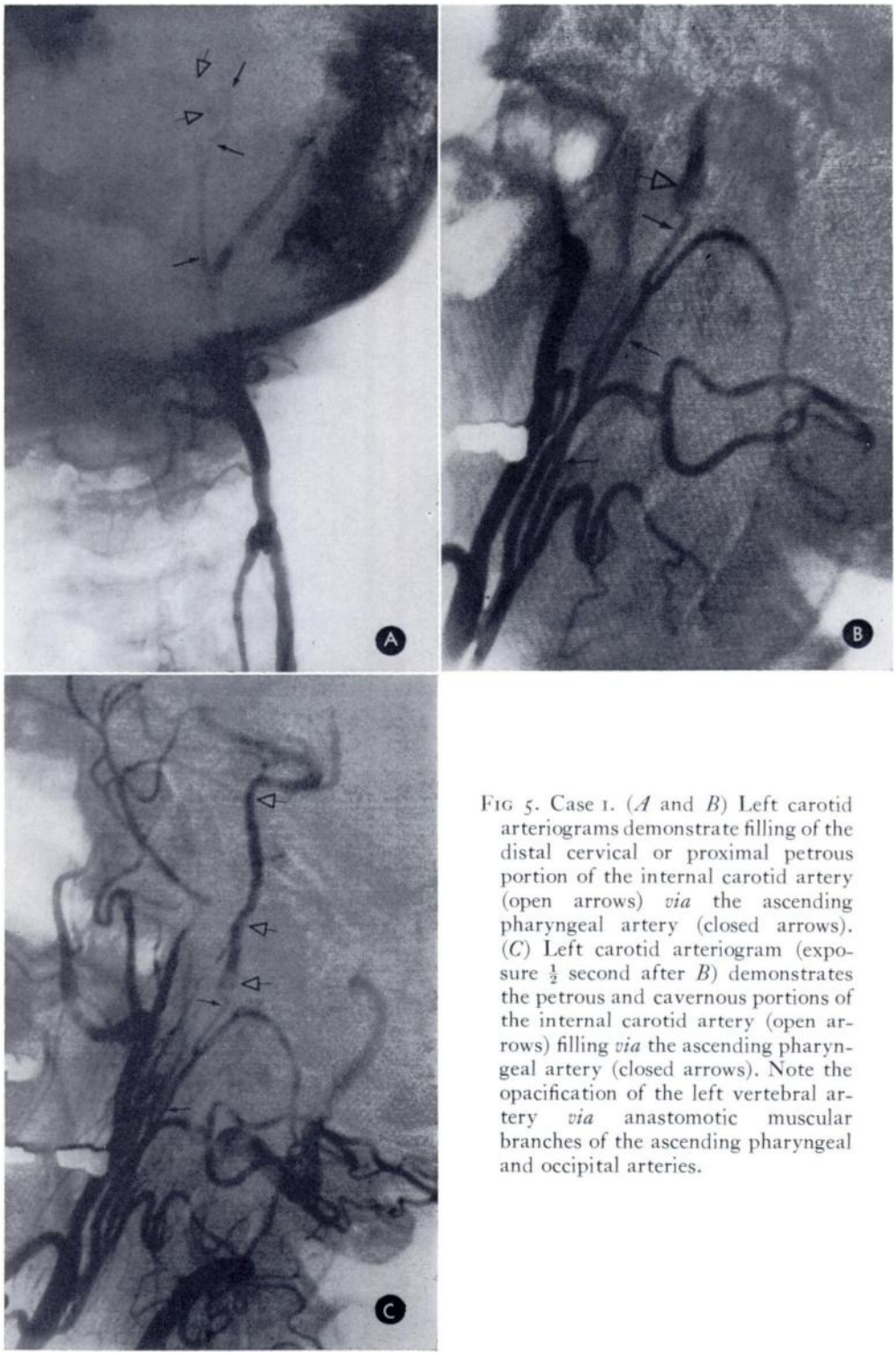

FIG 5. Case 1. (*A* and *B*) Left carotid arteriograms demonstrate filling of the distal cervical or proximal petrous portion of the internal carotid artery (open arrows) *via* the ascending pharyngeal artery (closed arrows). (*C*) Left carotid arteriogram (exposure $\frac{1}{2}$ second after *B*) demonstrates the petrous and cavernous portions of the internal carotid artery (open arrows) filling *via* the ascending pharyngeal artery (closed arrows). Note the opacification of the left vertebral artery *via* anastomotic muscular branches of the ascending pharyngeal and occipital arteries.

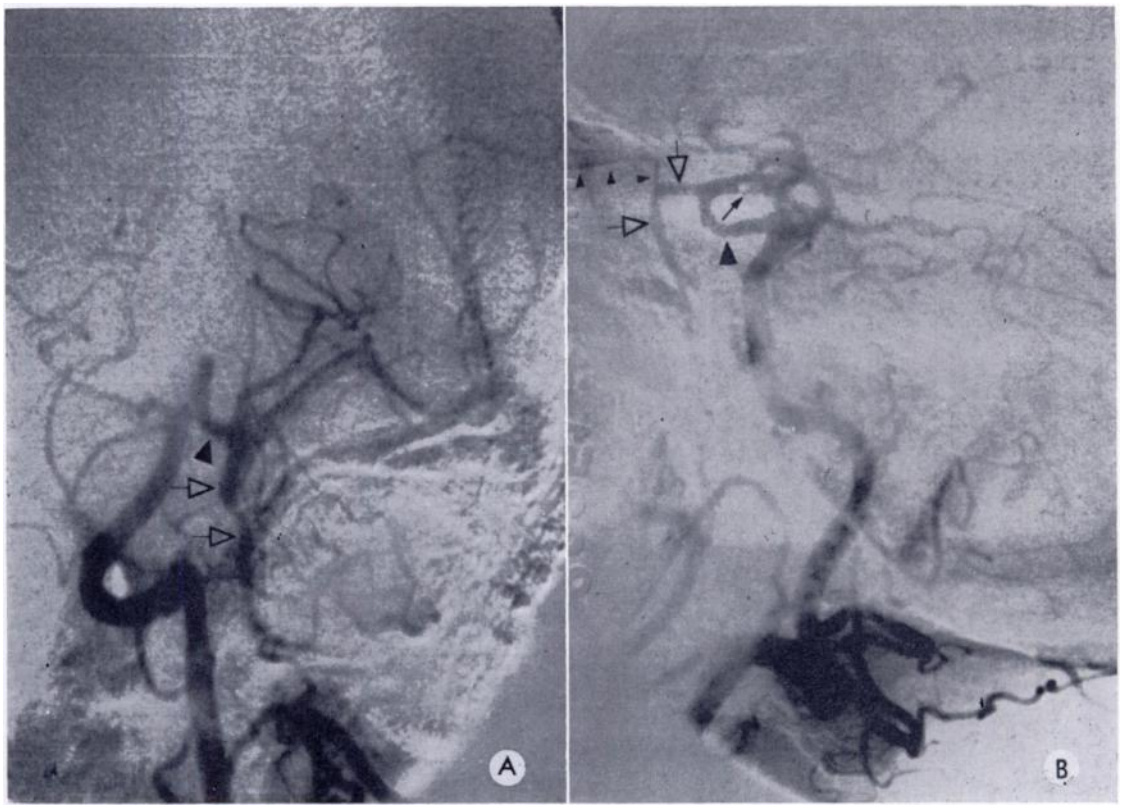

Fig. 6. Case 1. (*A*) Left brachial cerebral arteriogram with digital compression of the left "common" carotid artery demonstrates continuation of the basilar artery as a large arterial stem ("proximal left posterior cerebral artery") (arrowhead) on the left which gives origin to the left posterior cerebral and posterior communicating arteries and retrograde opacification of the internal carotid artery (arrows). The posterior communicating and internal carotid arteries are superimposed at the level of the upper arrow. (*B*) Left brachial cerebral arteriogram exposed simultaneously with *A* demonstrates a large posterior communicating artery (large arrowhead), retrograde opacification of the internal carotid (open arrows) and ophthalmic (small arrowheads) arteries, and an area of nonopacification (closed arrow) secondary to the right internal carotid artery contributing nonopacified blood to the left middle cerebral artery.

to unequivocally confirm or exclude the presence of an aneurysm, since this was of secondary importance at that time) of the left anterior cerebral artery near the usual vicinity of the origin of the anterior communicating artery.

Immediately after angiography, the patient was taken to surgery and a large subacute subdural hematoma evacuated. The patient expired on the third postoperative day. An autopsy was not performed.

DISCUSSION

As suggested by Taveras and Wood,[42] and Parker and Gaede,[30] the name primary cerebral hemiatrophy is a misnomer since what occurs is lack of cerebral development rather than atrophy. The terminology cerebral hemihypoplasia or unilateral cerebral hypoplasia as suggested by them gives a more accurate description. However, since the name primary cerebral hemiatrophy is firmly implanted in medical literature and its meaning is well understood, we have chosen to continue its use here.

Alpers and Dear[2] state that in primary or congenital cerebral hemiatrophy, cerebral symptoms (seizures, mental deficiency, and hemiparesis) are present from birth. They feel that it is the result of congenital and hereditary causes or from intrauterine transplacental insult.

Serres[37] in 1860 first proposed a vascular

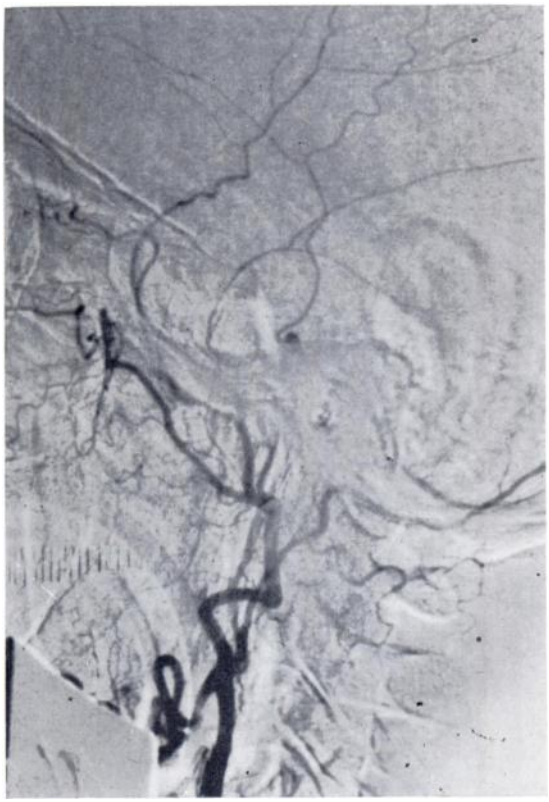

FIG. 7. Case II. Left "common" carotid arteriogram demonstrates absence of internal carotid artery.

pathogenesis; however, his theory has had few supporters since significant vascular disease has seldom been detected.[2,3,17,28,35–37] He felt that cerebral hemiatrophy was the result of anomalies of the cerebral arteries.

Case 1 presented here had a history of seizures, marked right hemiparesis, and mental retardation since birth. Bilateral carotid angiography revealed absence of the proximal left internal carotid artery and a large right internal carotid artery. Not even a suggestion of the typical common carotid bifurcation was noted on the left. However, a branch was noted to arise from the posterior surface of the "common" carotid artery just below the level of the origin of the superior thyroid artery (Fig. 3). This branch ascended for about 2 cm. and joined the external carotid artery. This is felt to be analogous to the branch of the common carotid artery which joined the internal carotid artery about 1 cm. distal to the carotid bifurcation as described by Adachi[1] and probably represents either an aborted attempt at formation of the proximal internal carotid artery or a simple carotid fenestration. The proximal

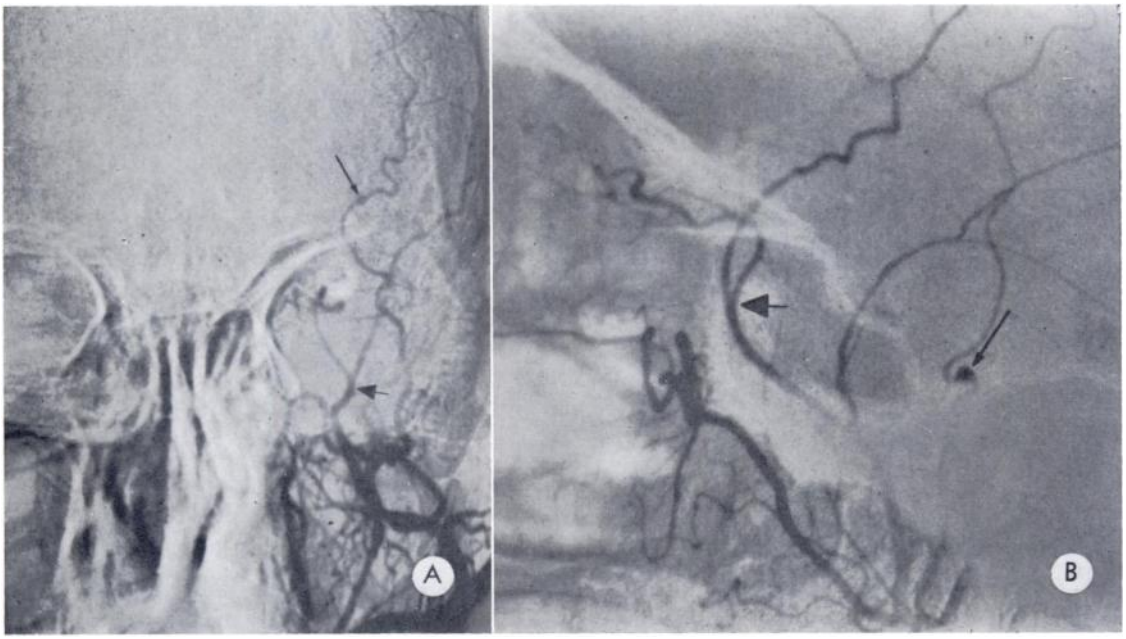

FIG. 8. Case II. (*A* and *B*) Left "common" carotid arteriograms demonstrate absence of internal carotid artery, origin of the ophthalmic artery from the middle meningeal artery (large arrow), and a small aneurysm of the superficial temporal artery (small arrow).

petrous or distal cervical portion of the left internal carotid artery was seen to communicate with the ascending pharyngeal artery which is a communication not known to have been previously reported. Lie,[23] however, has reported the angiographic demonstration of anastomoses between the ascending pharyngeal artery and the cavernous and distal petrous portions of the internal carotid artery. We, too, have encountered several examples of anastomoses between the ascending pharyngeal artery and the distal petrous and cavernous portions of the internal carotid artery in cases of proximal internal carotid occlusion.

No previous reports of angiographic confirmation of significant cerebrovascular deficiency in association with primary cerebral hemiatrophy are known to us. However, Parker and Gaede[30] have recently reported the postmortem discovery of unilateral internal carotid artery absence associated with primary cerebral hemiatrophy. These 2 cases lend strong support to the view that at least one etiology of primary cerebral hemiatrophy is vascular in origin.

The plain skull film roentgenogram (Fig. 1) of Case 1 demonstrates the roentgenographic findings of cerebral hemiatrophy reported by Dyke *et al.*[8] These findings

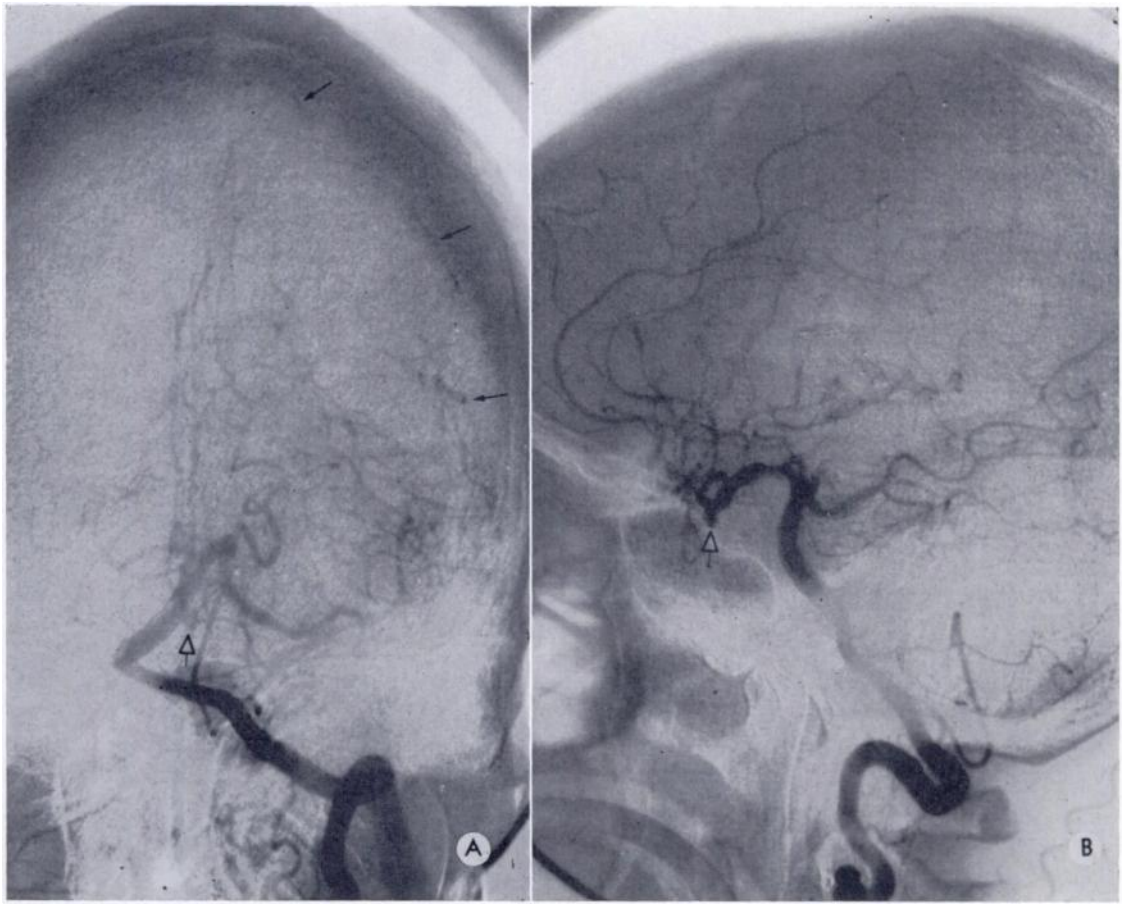

Fig. 9. Case 11. (*A* and *B*) Left brachial cerebral arteriograms demonstrate opacification of the left anterior cerebral and middle cerebral arteries, a probable aneurysm near the region of the anterior communicating artery (large arrow), and displacement of the peripheral middle and anterior cerebral arteries from the inner table of the skull by a subacute subdural hematoma (small arrows). Note the absence of dilution of contrast medium by nonopacified blood in the anterior and middle cerebral arteries indicating the absence of significant contribution by an anomalous internal carotid artery.

may be seen in primary and secondary cerebral hemiatrophy. In general, the bone changes tend to be more severe in the primary form. However, in view of considerable variation of cerebral and bone changes in both forms, plain skull film roentgenographic findings are of limited use in differentiating between primary and secondary cerebral hemiatrophy.

Even though agenesis of the internal carotid artery was first described in 1787 only few such cases have been reported. The most striking clinical significance of this anomaly is the association with intracranial aneurysms. Of the 31 known reported cases of total absence of the internal carotid, 7 have presented with subarachnoid hemorrhage, 4 resulting from anterior communicating aneurysms, 2 resulting from basilar artery aneurysms, and 1 was the result of a middle cerebral aneurysm. Case II presented here was noted to have aneurysms of the cavernous internal carotid artery on the right and a probable intradural aneurysm on the left near the origin of the anterior communicating artery.

Two patients with agenesis of the internal carotid artery without demonstrable aneurysm, in addition to the case associated with primary cerebral hemiatrophy reported by Parker and Gaede,[30] have been reported who presented with neurologic deficits. Verbiest[47] reported that 1 of these 2 patients presented with paralysis of several cranial nerves secondary to compression by a dilated loop of the basilar artery. The second patient reported by Hussain *et al.*,[16] presented with left hemiparesis following head trauma and was shown surgically and angiographically to have congenital absence of the right internal carotid artery. The case of Hussain *et al.* did not demonstrate an acute surgical intracranial lesion and it was felt that minor head trauma was sufficient to alter the cerebral hemodynamics to produce hemiparesis.

The anomalies of the Circle of Willis angiographically demonstrated here in total absence of the internal carotid artery are quite similar to the autopsy findings of several previously recorded cases[11,12,24,46] with 2 significant exceptions. The exceptions are: (1) the ipsilateral ophthalmic artery in this patient originated from the middle meningeal artery rather than the middle cerebral artery; and (2) the pericallosal arteries in this patient originated from their respective sides rather than both originating from the contralateral side.

It is generally felt that, in order to conclusively diagnose congenital total absence of an internal carotid artery, its absence should be confirmed surgically or absence of the carotid canal should be demonstrated either roentgenographically or by dissection. However, Case II is felt to represent an example of congenital total absence of the left internal carotid artery in spite of lack of neck exploration and temporal bone dissection and/or tomography. This is felt to be the situation because of lack of demonstrable remnants of the proximal and supraclinoidal internal carotid artery as is usually demonstrable in cases of acquired occlusion. Additional evidence supporting congenital absence is the origin of the left ophthalmic artery from the left middle meningeal artery, which is an extremely rare phenomenon, whereas the inverse, *i.e.*, ophthalmic artery origin of the middle meningeal artery, occurs much more frequently. Origin of the left internal carotid artery from the aorta is even rarer than internal carotid artery absence[23] and is doubtful, since the left anterior and middle cerebral arteries were well opacified *via* the basilar artery without evidence of dilution of contrast material by nonopacified blood entering from an anomalous internal carotid artery. The associated extremely rare vascular aberration on the right of absence of the external carotid artery with its usual branches arising from the internal carotid artery combined with persistence of the stapedial artery lend even more supportive evidence for a congenital basis of absence of the left internal carotid artery.

Angiographic demonstration of oblitera-

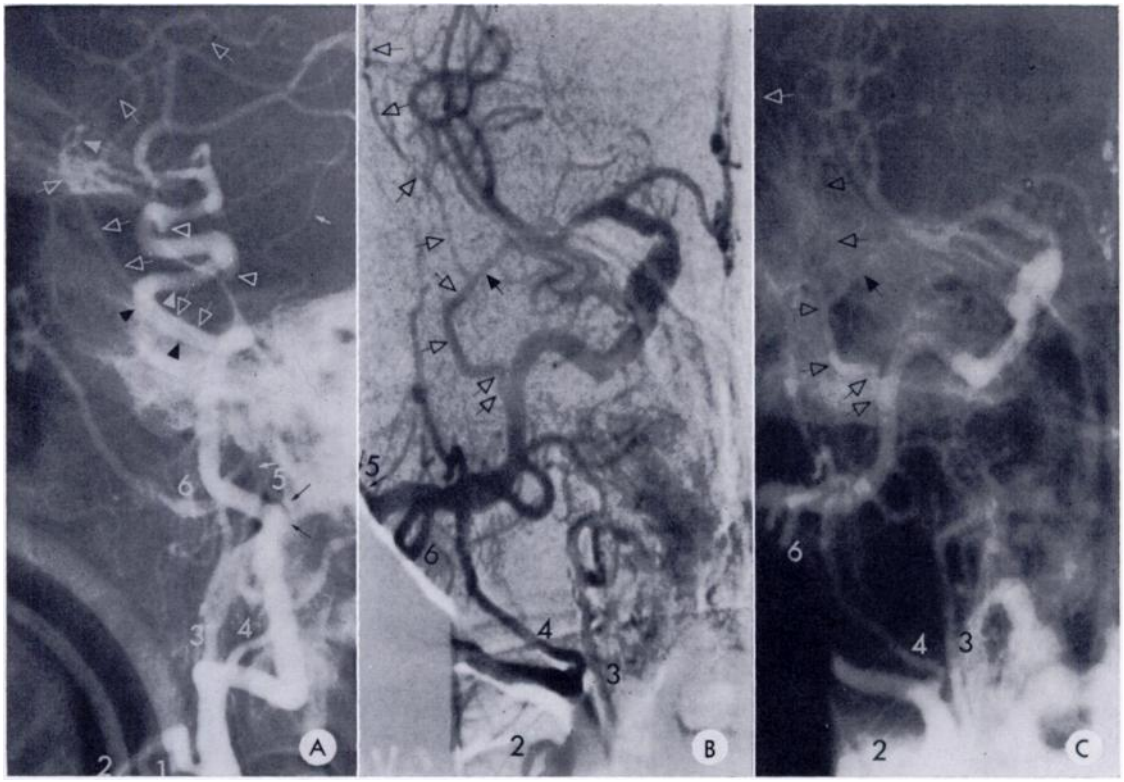

Fig. 10. Case 11. (*A–C*) Right "common" carotid arteriograms demonstrate absence of the external carotid artery with its usual branches (lingual (1), facial (2), ascending pharyngeal (3), occipital (4), superficial temporal (5, small closed arrows), and internal maxillary (6) arteries) arising from the tortuous internal carotid artery, origin of the middle meningeal artery (persistent stapedial artery) (open arrows) from the proximal petrous portion of the internal carotid artery, an ophthalmic artery branch (large closed arrow) of the middle meningeal artery, an elongated anteriorly convexed course of the distal petrous portion (closed arrowheads) of the internal carotid artery, opacification of only the right anterior cerebral artery and its branches, and small aneurysms (open arrowheads) of the cavernous portion of the internal carotid artery. Note in *A* the double density along the inferior limb of the anteriorly convexed distal petrous internal carotid artery indicating superimposition of the middle meningeal artery upon this segment. Note in *C* that the origin of the middle meningeal artery (open arrows) from the proximal petrous internal carotid is identifiable in spite of the density of the overlying petrous temporal bone.

tion of the distal external carotid artery with its usual branches arising from the internal carotid artery has been previously reported by Seidel.[36] In his case the proximal stem of the external carotid artery was present, whereas in Case 11 presented here no evidence of a proximal stem was noted. Angiographic demonstration of usual branches of the external carotid artery arising from the internal carotid artery without associated external carotid absence has been reported previously by several authors.[10,21,27,43]

Persistence of the stapedial artery is an extremely rare anomaly in man. The authors' review of the literature has revealed a single[13] previous angiographic demonstration. According to Padget[29] the stapedial artery appears in the 14 mm. (35 days) embryo as a branch of the hyoid artery. The stapedial artery has 2 main divisions, *viz.* maxillomandibular and supraorbital. The maxillomandibular division represents the stem of the future middle meningeal artery. The supraorbital branch represents the extended course of the future middle meningeal artery. By approximately the 40 mm. (52 days) stage all branches of the

stapedial artery have usually been annexed by the ophthalmic and external carotid arteries and the proximal stem of the stapedial artery has usually involuted.

The elongated, anteriorly convexed course of the distal petrous portion of the right internal carotid artery (Fig. 10, *B* and *C*) is quite similar to that demonstrated by Guinto and associates.[13] The significance of the association of the course of distal petrous portion of the internal carotid artery with persistence of the stapedial artery is unknown at this time.

The primary angiographic difference in the persistent stapedial artery demonstrated here and the previously reported Case 13 is that a branch (Fig. 10, *A–C*) of the middle meningeal artery either joins the ophthalmic artery or forms the principal part of the ophthalmic artery. Padget[29] feels that this should be occasionally expected, since in the 20 mm. (40 days) embryo the supraorbital branch of the stapedial artery forms an anastomosis with the ophthalmic artery. This, combined with failure of the internal maxillary artery to annex the involuting stapedial artery, is the explanation for ophthalmic artery origin of the middle meningeal artery. The inverse, *i.e.* middle meningeal artery origin of the ophthalmic artery, as shown on the left side (Fig. 8, *A* and *B*) in Case 11 may be explained by the stapedial-ophthalmic artery anastomosis combined with involution of the embryonic internal carotid artery after annexation of the stapedial artery by the internal maxillary artery.

SUMMARY

Unilateral congenital absence of the internal carotid artery, either partial or total, has been rarely demonstrated. We are presenting 1 case of each entity.

The case of partial absence is unique in that it is felt to represent the first known case of demonstration of a vascular etiology of primary cerebral hemiatrophy in a living patient.

The case of unilateral total absence of the internal carotid was associated with contralateral absence of the external carotid artery and persistence of the stapedial artery which were incidental findings in a patient who presented with a subdural hematoma following head trauma.

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