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Total Agenesis of the Internal Carotid Artery

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Congenital absence of the internal carotid artery is extremely rare. Angiographic demonstrations of three cases of unilateral total agenesis and one case of bilateral total agenesis of the internal carotid artery are reported. Hypocycloidal tomography confirmed the absence of the appropriate carotid canals. A brief description of the embryology of the internal carotid artery is given.

Since Tode (cited in [10]) first described congenital absence of the internal carotid artery in 1787, there have been 34 previously reported cases [1–27] (Cohen MM, Kristiansen K; Koberwein; Quain; and Wyeth JA, all cited in [10]; Wernitz A, cited in [28]) of unilateral total absence of the internal carotid artery and eight previously reported cases [28–35] of bilateral total absence of the internal carotid artery. Congenital total absence of the internal carotid artery has been demonstrated by angiography unilaterally in 17 cases [1, 6, 8–10, 12–14, 19, 22, 24, 26, 27, 36] and bilaterally in two cases [30, 33]. Seven cases [10, 37–42] of unilateral partial congenital absence of the internal carotid artery with an intercavernous anastomosis between the contralateral "normal" and ipsilateral partially aplastic internal carotid arteries have been identified by angiography. We offer angiographic demonstrations of unilateral total agenesis of the internal carotid artery in three new cases and a fourth case with bilateral total agenesis. Confirmatory hypocycloidal tomographs of the petrous bones demonstrating absence of the appropriate carotid canals are presented.

An excellent description of the embryology of the human carotid arteries is that of Padget [43]. She stated that the internal carotid arteries first appear at the 3 mm (24 day) stage and are formed from the terminal segments of the paired dorsal aorta and from the third branchial arches. The definitive common carotid artery and most of the definitive branches of the external carotid artery are present by the 16–18 mm (40 day) stage.

The sequence of events that lead to absence of the internal carotid artery is unknown. However, Keen (32) suggested 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. He offered no explanation of the embryogenesis of bilateral absence of the internal carotid artery.

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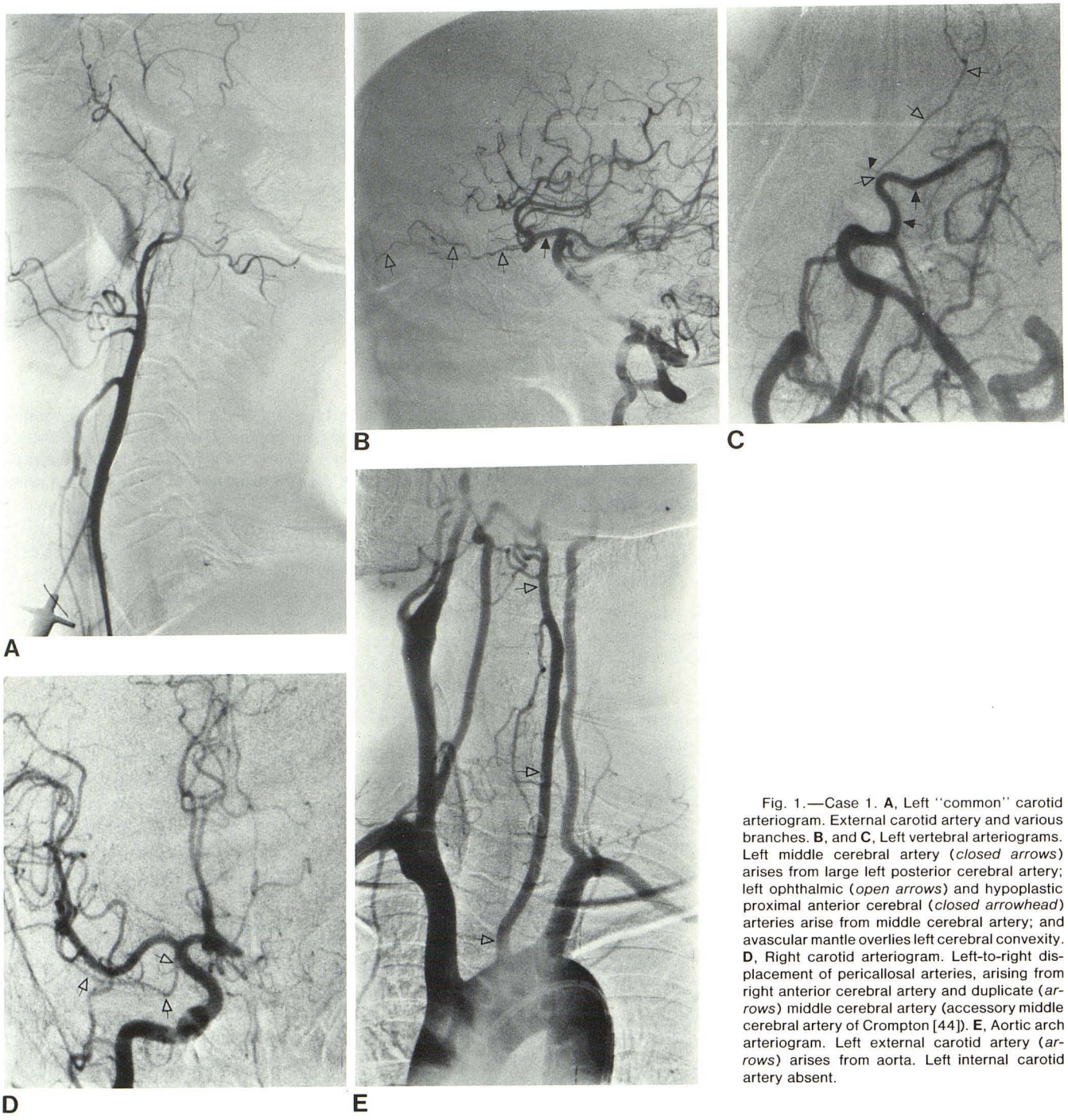

Fig. 1.—Case 1. **A**, Left "common" carotid arteriogram. External carotid artery and various branches. **B**, and **C**, Left vertebral arteriograms. Left middle cerebral artery (*closed arrows*) arises from large left posterior cerebral artery; left ophthalmic (*open arrows*) and hypoplastic proximal anterior cerebral (*closed arrowhead*) arteries arise from middle cerebral artery; and avascular mantle overlies left cerebral convexity. **D**, Right carotid arteriogram. Left-to-right displacement of pericallosal arteries, arising from right anterior cerebral artery and duplicate (*arrows*) middle cerebral artery (accessory middle cerebral artery of Crompton [44]). **E**, Aortic arch arteriogram. Left external carotid artery (*arrows*) arises from aorta. Left internal carotid artery absent.

Case Reports

Case 1

A 56-year-old woman had an unremarkable history except for several weeks of recurrent headaches and 1 day of marked memory impairment. Admission physical examination was unremarkable. A radionuclide brain scan demonstrated subcalvarial activity in the left parietal region, thought to be secondary to a subdural hematoma or meningioma.

Her condition worsened with the onset of emesis episodes and increasing confusion and lethargy. Left carotid angiography (fig. 1A) demonstrated only the external carotid artery, which was believed to be secondary to agenesis of the internal carotid artery. Right brachial cerebral angiography (not shown) immediately after left carotid angiography demonstrated a large left chronic subdural hematoma and origin of the left middle cerebral artery from an enlarged left posterior cerebral artery. After angiography, a large chronic subdural hematoma was surgically evacuated.

Because of persistent lethargy repeat angiography was per-

formed. Selective left vertebral angiography (figs. 1B and 1C) demonstrated origin of the left middle cerebral artery from the left posterior cerebral artery and origin of the ophthalmic and hypoplastic proximal anterior cerebral arteries from the left middle cerebral artery. Right carotid angiography (fig. 1D) demonstrated left-to-right displacement of the pericallosal arteries, both of which were supplied by the right anterior cerebral artery, and duplication of the right middle cerebral artery (accessory middle cerebral artery of Crompton [44]). Aortic arch angiography (fig. 1E) demonstrated no separate origin to the left internal carotid artery.

Temporal bone hypocycloidal tomography (fig. 2) failed to demonstrate a left carotid canal. However, a small vertical cleft about 1.5 cm (uncorrected) in diameter was noted anteromedial to the left external auditory canal in the usual region of the proximal part of the carotid canal. Since no evidence of a horizontal component of this small vertical cleft in the left temporal bone was noted, the cleft was not believed to represent a hypoplastic carotid canal. The patient was discharged 3 weeks after surgery without neurologic deficit.

Case 2

A 73-year-old man was admitted for evaluation prior to planned left superficial temporal–middle cerebral artery anastomosis. Earlier aortic arch angiography for suspected transient ischemic attacks at another hospital was interpreted as complete occlusion of the left internal carotid artery. Left carotid (fig. 3A) and aortic arch (fig. 3B) angiography on the day after admission demonstrated complete absence of the left internal carotid artery. Postangiography Stenver projection hypocycloidal tomography of the petrous parts of the temporal bones (figs. 3C and 3D) demonstrated absence of the left carotid canal with normal right internal carotid canal.

Case 3

A 57-year-old white woman was admitted with chest pain and fainting spells. Physical examination was normal. Cardiac catheterization revealed normal coronary arteries. The esophageal motility test was within normal limits. Right and left common carotid angiograms (figs. 4A and 4B) revealed no evidence of internal carotid arteries originating from the common carotid arteries. No occlusion was suspected. To evaluate the intracranial circulation, left vertebral angiography (figs. 4C and 4D) was performed which demonstrated the right and left anterior and middle cerebral arteries filling from posterior communicating arteries on both sides. The left vertebral artery was larger than normal. The aortic arch study showed the right and left common carotid arteries with no separate origin to suggest evidence of internal carotid arteries on either side (fig. 4E). There was a small atheromatous plaque at the origin of the left subclavian artery with no high-grade obstruction. Hypocycloidal tomography of the petrous parts of the temporal bones (fig. 5) failed to demonstrate either left or right carotid canal.

Case 4

A 14-year-old boy was asymptomatic until 3 months before admission when he noted an odd sensation in his throat followed shortly by a noticeable voice change and difficulty swallowing. Indirect laryngoscopy demonstrated paralysis of the left vocal cord and neurologic examination revealed palsies of left cranial nerves IX–XII, a left Horner syndrome, and left cervical adenopathy. Cranial computed tomography (CT) and metrizamide CT cisternography were normal. Hypocycloidal tomography in submentovertical projection (fig. 6A) demonstrated absence of the left carotid canal. Biopsy of an enlarged left cervical lymph node to confirm or exclude lymphoma revealed only chronic inflammatory lymphoid hyperplasia. Left carotid (fig. 6B), left vertebral (fig. 6C), and aortic arch (fig. 6D) angiography were unremarkable except for complete absence of the left internal carotid artery and anomalous origin of the left vertebral artery from the aorta. The vertebral arteries were symmetrical with normal courses and calibers and the basilar artery was normal in course and caliber. Therefore, the patient's symptom complex was believed to be secondary to causes unrelated to his vascular anomaly. He was discharged without a definitive diagnosis.

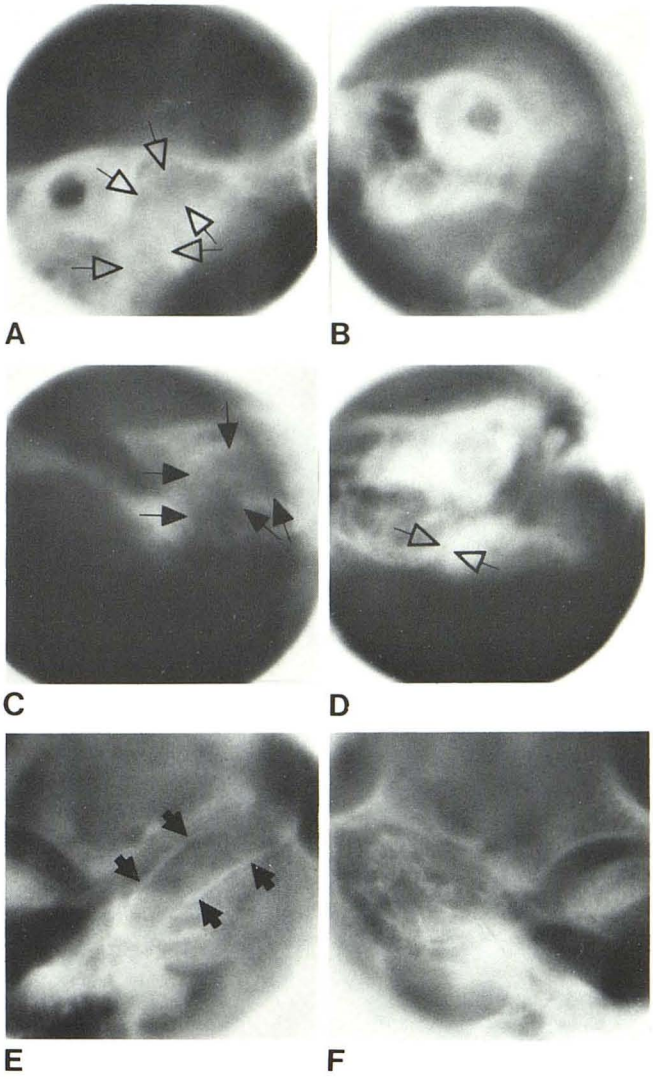

Fig. 2.—Case 1. Hypocycloidal tomograms of both temporal bones. Lateral (**A** and **B**), anteroposterior (**C** and **D**), and submentovertical views (**E** and **F**). Normal right (**A**, **C**, and **E**) (*closed arrows*) and absent left (**B**, **D**, and **F**) carotid canals. Small vertical cleft (**D**, *open arrows*) in left temporal bone near usually located carotid canal is not believed to represent hypoplastic carotid canal.

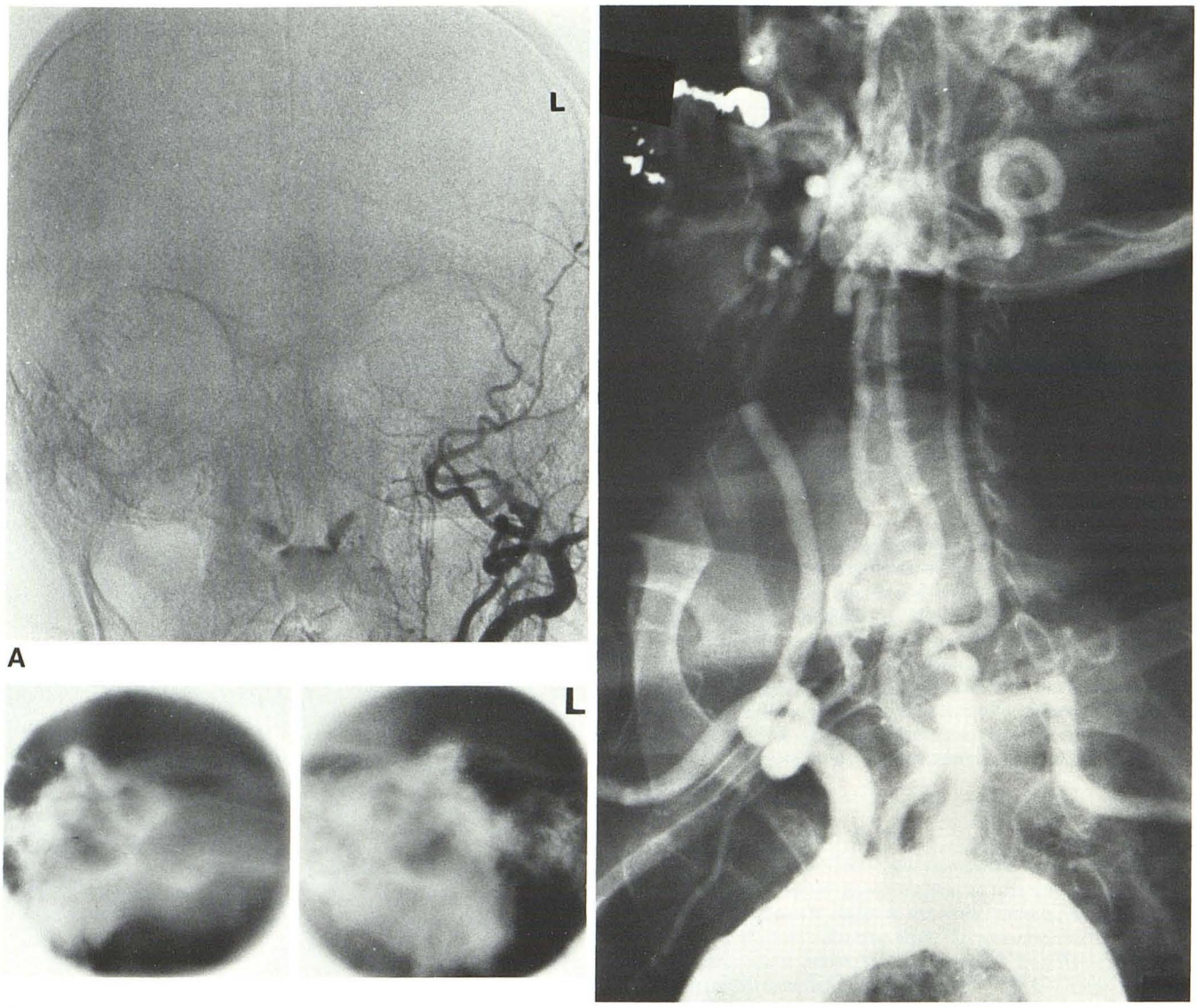

Fig. 3.—Case 2. **A**, Frontal view of left carotid angiogram. No evidence of internal carotid artery. **B**, Aortic arch angiogram. Bifurcation of right common carotid artery and origin of left "common" carotid artery, but no separate origin to left internal carotid artery. **C**, Polytomogram of right temporal bone, Stenver position. Horizontal and vertical parts of right carotid canal. **D**, Polytomogram of left temporal bone, Stenver view. No evidence of carotid canal.

Discussion

Since first described in 1787, 42 cases of congenital absence of the internal carotid artery have been reported. In 12 of these, subarachnoid hemorrhage due to ruptured intracranial aneurysms was the initial finding. Our four patients had neither aneurysms nor hemorrhage. The literature reveals a left-side predilection of unilateral total agenesis of the internal carotid artery at a ratio of about 3:1. Therefore, it is not surprising that our three cases of unilateral agenesis involved the left internal carotid artery.

At least four previous patients with congenital total absence of the internal carotid artery without associated ruptured aneurysms had neurologic deficits. The first, reported by Verbiest [26], had paralysis of several cranial nerves secondary to compression by a dilated loop of basilar artery. Hussain et al. [8] reported a case with left hemiparesis following head trauma who was shown surgically and angiographically to have complete absence of the right internal carotid artery. In the absence of an intracranial lesion it was believed that minor head trauma was sufficient to alter the cerebral hemodynamics to produce hemiparesis. The third case, reported by Parker and Gaede [15], was associated with primary cerebral hemiatrophy secondary to agenesis of the left internal carotid, middle cerebral, and posterior communicating arteries. The fourth patient, reported by Teal et

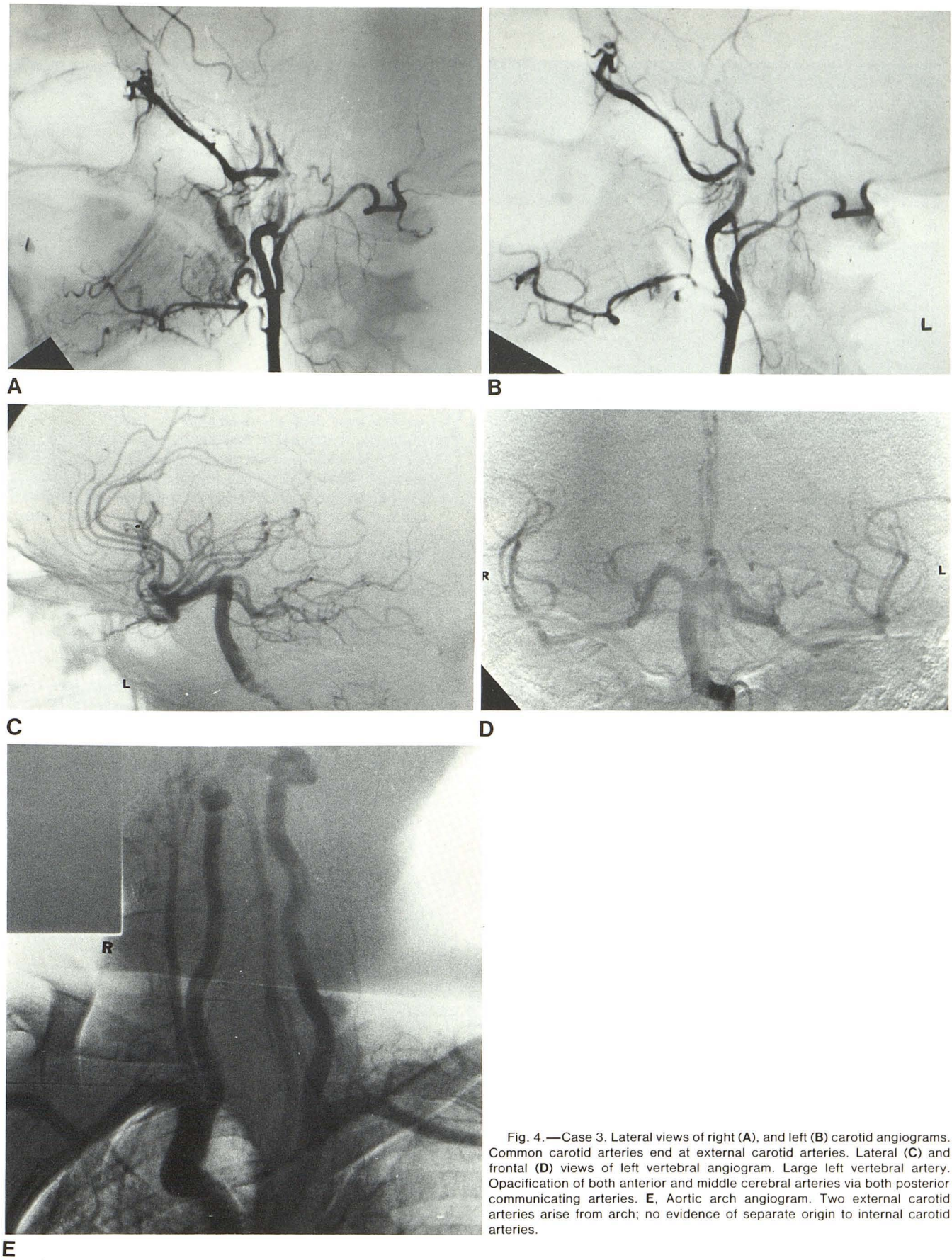

Fig. 4.—Case 3. Lateral views of right (**A**), and left (**B**) carotid angiograms. Common carotid arteries end at external carotid arteries. Lateral (**C**) and frontal (**D**) views of left vertebral angiogram. Large left vertebral artery. Opacification of both anterior and middle cerebral arteries via both posterior communicating arteries. **E**, Aortic arch angiogram. Two external carotid arteries arise from arch; no evidence of separate origin to internal carotid arteries.

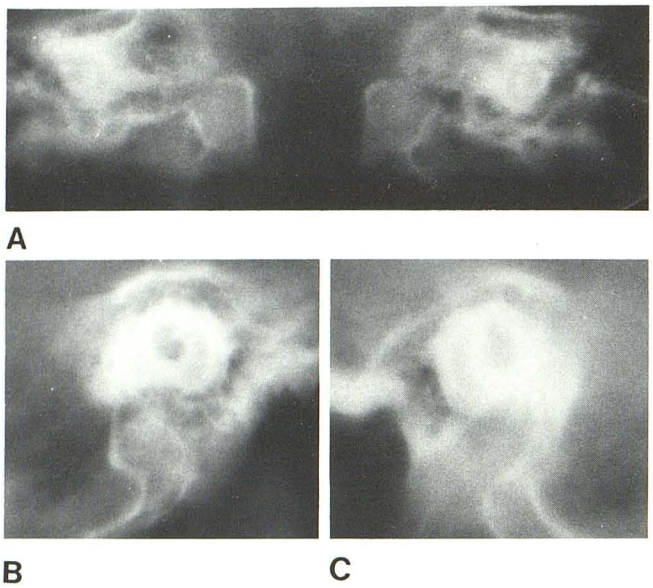

Fig. 5.—Case 3. Frontal (**A**) and lateral (**B** and **C**) polytomograms of petrous bones. No evidence of carotid canal bilaterally.

al. [22], had dysphasia, facial nerve weakness, and seizure secondary to a large subacute subdural hematoma.

Origin of the ophthalmic artery from the ipsilateral middle cerebral artery and supply of both pericallosal arteries from the contralateral anterior cerebral artery in case 1 are identical to several previously reported cases [5, 11, 25, 37] of unilateral congenital absence of the internal carotid artery demonstrated anatomically at autopsy. Duplication of the right middle cerebral artery (accessory middle cerebral artery of Crompton [44]) is believed to represent an incidental anomaly and is not directly involved in the sequence of events that led to agenesis of the left internal carotid artery.

It is debatable whether or not case 1 had agenesis or atresia of the left internal carotid artery. In true agenesis of the internal carotid artery, the carotid canal probably should be totally absent. In atresia of the internal carotid artery, the carotid canal should be markedly hypoplastic since a fibrous, atretic internal carotid artery should persist. In case 1, temporal bone hypocycloidal tomography (fig. 2D) demonstrated a lucency that may have represented a markedly hypoplastic proximal vertical part of the left carotid canal. However, in view of failure to demonstrate a complete horizontal part, we believe that the small vertical cleft does not represent a hypoplastic carotid canal. Admittedly, a thread-like, atretic mid and distal petrous internal carotid artery should be present in association with a carotid canal so small that it might not be detectable by hypocycloidal tomography or by dissection.

The explanation of the small vertical cleft in the left temporal bone of case 1 is unknown to the authors. To be considered among several possible etiologies are an aborted carotid canal, the inferior tympanic canaliculus, an anomalous anteriorly placed neural compartment of the jugular fossa, and site of passage of a meningeal branch of the ascending pharyngeal artery through the base of the skull.

Hypocycloidal tomography of the petrous parts of the temporal bones of cases 2–4 demonstrated no evidence of even a remnant of the involved carotid canals. However, fewer projections were obtained in these cases than were obtained in case 1. We believe the combination of angiographic evidence of absence of the internal carotid arteries and hypocycloidal tomographic evidence of absence of the appropriate carotid canals represents sufficient evidence that our cases are congenital rather than acquired absence of the internal carotid artery.

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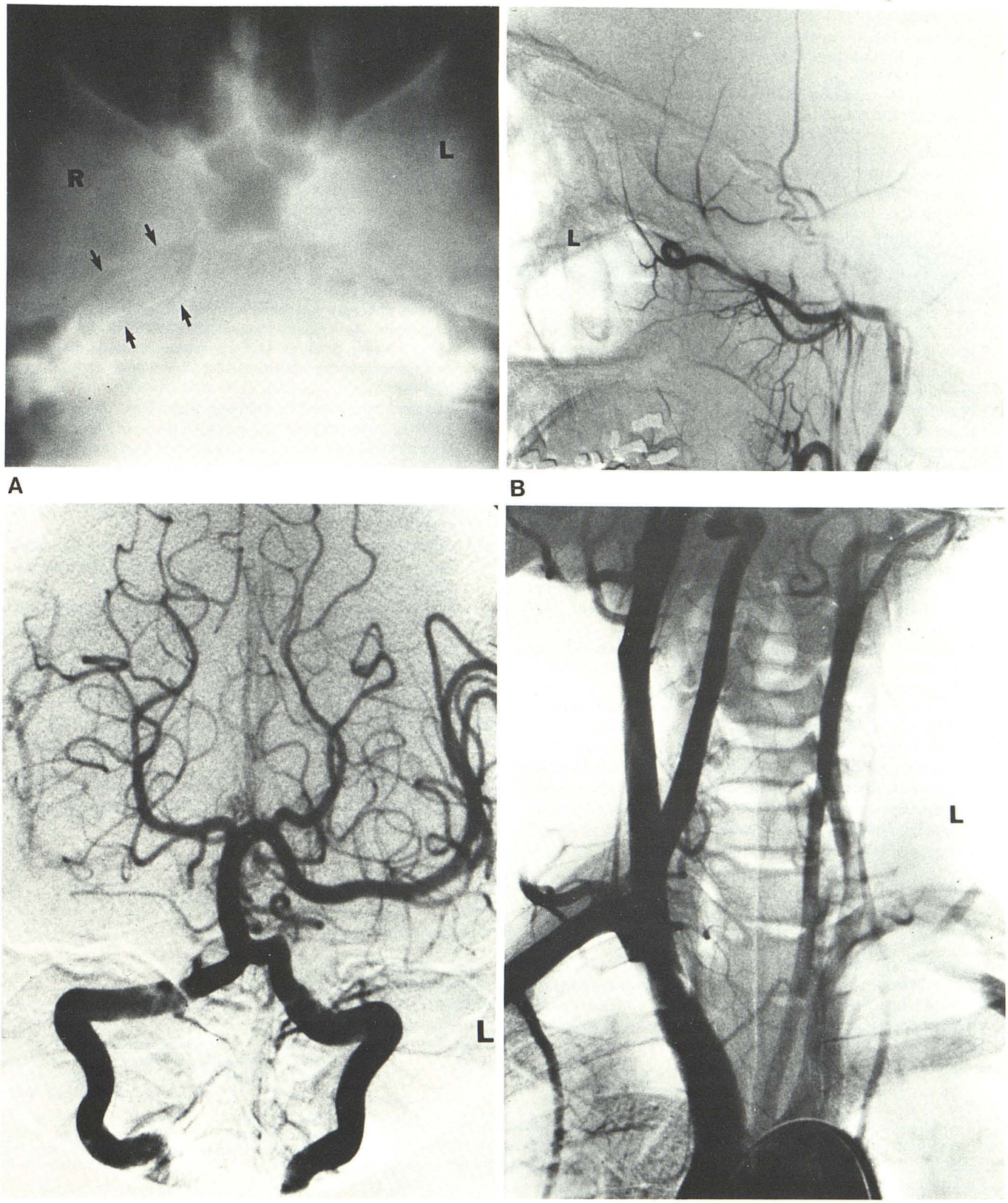

Fig. 6.—Case 5. **A**, Polytomogram of skull, submentovertical view. Right carotid canal (*arrows*) with no evidence of canal on left side. **B**, Lateral view of left carotid angiogram. Left external carotid artery and branches. **C**, Frontal view of left vertebral angiogram. Patent vertebrobasilar system. Opacification of left middle cerebral artery via prominent left posterior communicating artery with flash filling of right middle cerebral artery. Retrograde opacification of right vertebral artery. **D**, Aortic arch angiogram. No separate origin to left internal carotid artery. Separate origin to left vertebral artery from arch.

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