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M C Chen, H M Liu and K M Huang

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# Agenesis of the Internal Carotid Artery Associated with Neurofibromatosis Type II

Ming-Chang Chen, Hon-Man Liu, and Kou-Mou Huang

**Summary:** We report a case of agenesis of the internal carotid artery associated with neurofibromatosis type II. Common carotid arteriograms showed no visualization of the internal carotid artery. CT revealed absence of the corresponding carotid canal. MR documented absence of the internal carotid artery in the cavernous sinus.

**Index terms:** Arteries, abnormalities and anomalies; Arteries, carotid (internal); Neurofibromatosis

Congenital absence of the internal carotid artery is a rare anomaly. Since Tode first described absence of the internal carotid artery in 1787 (1), fewer than 100 cases have been reported in the literature (2).

When the internal carotid artery is congenitally absent, collateral circulation develops through the circle of Willis from the basilar artery and the opposite internal carotid artery to supply the involved hemisphere. Therefore, neurologic deficits are few (2). The ophthalmic artery on the affected side sometimes comes from the external carotid artery but may arise from a branch of the middle cerebral artery (3).

Agenesis of the internal carotid artery associated with other congenital malformations is uncommon (4, 5). We report a case of agenesis of the internal carotid artery occurring in a patient with neurofibromatosis type II.

## Case Report

A 14-year-old girl was admitted with the chief complaints of poor vision for 7 years and headache for several months. On physical examination, several subcutaneous nodules were noted on her forehead and extremities. The largest one had been removed. Pathologic analysis showed it to be a neurofibroma. Her father and her elder sister had similar cutaneous problems.

Magnetic resonance was performed and revealed lesions in both internal auditory canals, Meckel's caves and the right parietal region were hypointense to gray matter on T1-weighted images and hyperintense on T2-weighted images. The lesions enhanced, and bilateral acoustic neuromas, bilateral trigeminal ganglion neuromas, and meningioma (Fig 1A) were diagnosed. This feature suggested the diagnosis of neurofibromatosis type II (4). There was no visualization of the normal signal void of the cavernous portion of left internal carotid artery (Fig 1B). The convexity meningioma was surgically removed.

A left common carotid angiogram revealed no evidence of the internal carotid artery originating from the common carotid artery, but the accessory meningeal artery recanalized the distal cavernous portion of the internal carotid artery (Figs 1C and 1D). A hypoplastic ophthalmic artery opacified late from carotid collaterals. A left vertebral angiogram demonstrated collateral flow to the left anterior and middle cerebral arteries from the left posterior communicating artery. A right common carotid angiogram revealed collateral flow to the left anterior cerebral artery. No aneurysm was found on the angiograms. Computed tomographic scans of the temporal bones showed an absence of the left carotid canal (Figs 1E and 1F).

## Discussion

There is no universal agreement on the developmental origin of the common carotid artery and its two branches, the internal and external carotid arteries (1). Two theories try to explain the embryology of the internal carotid artery (2). However, the sequence of events leading to total absence of the internal carotid artery still is unknown (4).

Varying degrees of embryonic insult to the internal carotid artery cause agenesis, aplasia, or hypoplasia of an internal carotid artery (4, 6, 7).

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From the Department of Radiology, Medical College and Hospital, National Taiwan University, Taipei.

Address reprint requests to Ming-Chang Chen, Department of Radiology, Medical College and Hospital, National Taiwan University, 1, Chang-Te St Taipei City, 100, Taiwan, Republic of China.

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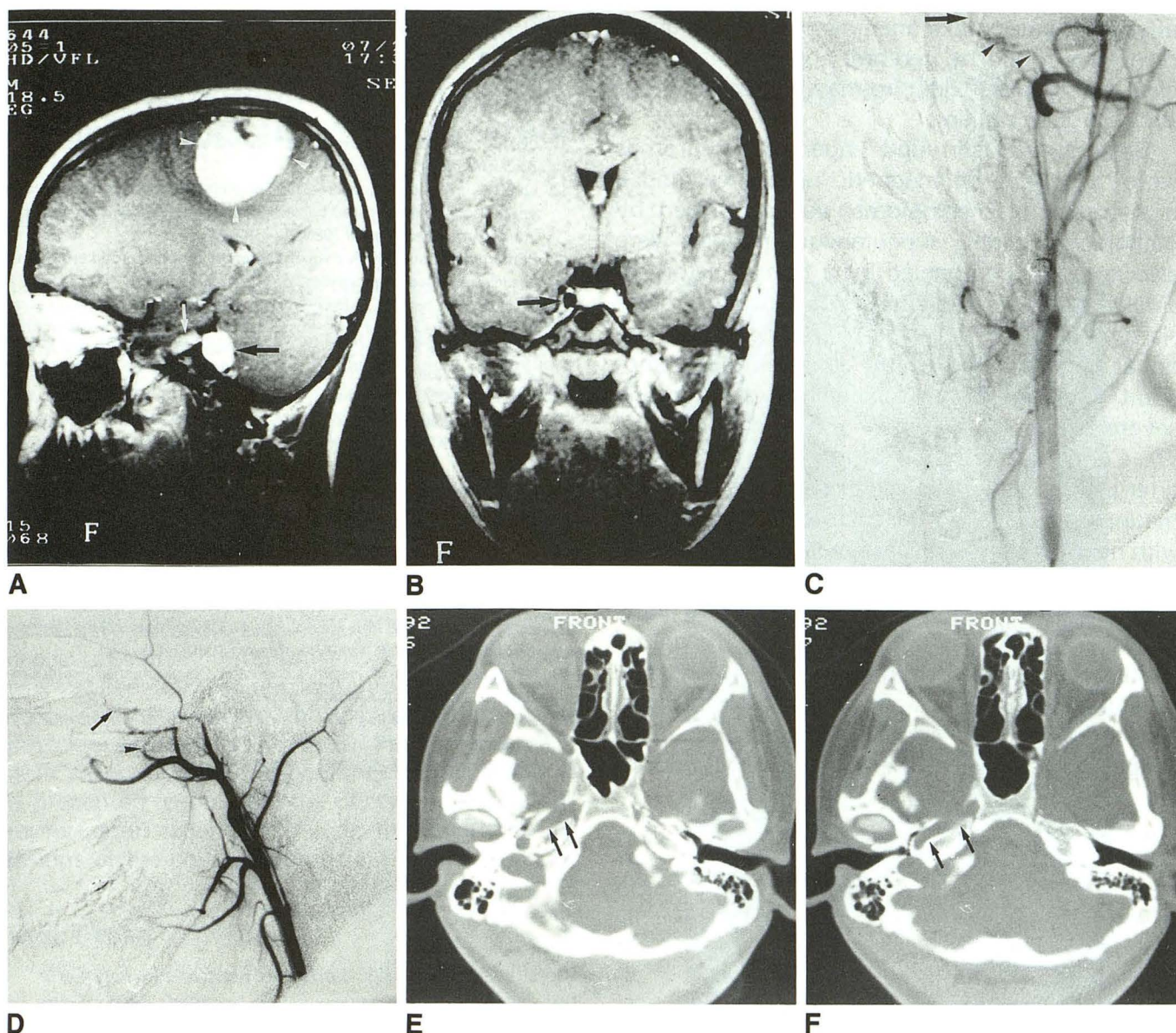


Fig. 1. A, Right parasagittal T1-weighted image with gadopentetate dimeglumine enhancement (500/15/2 [repetition time/echo time/excitations]). A meningioma (*arrowheads*) at the high convexity is shown (pathologic proof after surgical removal). Trigeminal neuroma (*white arrow*) and acoustic neuroma (*black arrow*) also are diagnosed by magnetic resonance.

B, Coronal T1-weighted image with gadopentetate dimeglumine enhancement (600/15). A normal right internal carotid artery (*arrow*) is noted as a signal void, but there is no evidence of left internal carotid artery within the cavernous sinus.

C and D, Left common carotid arteriogram, anteroposterior and lateral projections. There is no evidence of the internal carotid artery arising from the common carotid artery, but the accessory meningeal artery (*arrowhead*) recanalizes the distal cavernous portion of the internal carotid artery (*arrow*).

E and F, Computed tomographic scans of the temporal bones demonstrate absence of the left carotid canal. *Arrows* reveal the normal right carotid canal.

Agenesis and aplasia can be differentiated by the absence or presence, respectively, of some remnant of the vessel, and by demonstration of the carotid canal at the skull base by computed tomography or hypocycloidal tomography of the temporal bone (2, 8). Hypoplasia is underdevelopment of the internal carotid artery.

In this case, neither a remnant of the internal carotid artery nor the carotid canal at the skull base were shown by computed tomography or angiography.

The recognition of congenital absence of the internal carotid artery is of clinical significance. Several reports have documented a high associ-



ation with intracranial aneurysm (2, 7). No aneurysm was found by cerebral angiograms in this case, but regular follow-up may be necessary for such a young patient.

Bilateral acoustic and trigeminal neuromas were diagnosed by magnetic resonance, and a high-convexity meningioma was confirmed by pathology. Multiple subcutaneous nodules were also noted, accompanied by a family history of similar lesions. The diagnosis of neurofibromatosis type II was established according to the National Institutes of Health Consensus Development Conference as a distinct disorder among several proposed categories of neurofibromatosis (9). Congenital absence of the internal carotid artery combined with other congenital malformations is rare (4, 5). Vascular lesions associated with neurofibromatosis I are mainly characterized by stenosis, occlusion, rupture, and aneurysm or fistula formation of large and medium-size arteries (10). Vascular disease is not yet a recognized manifestation of neurofibromatosis type II (10), possibly because types I and II are distinct disorders with different genetic linkages (10). This congenital absence of the internal carotid artery

with neurofibromatosis type II may be a chance occurrence.

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