Pediatr Radiol (1999) 29: 586–588


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# Aplasia of right internal carotid artery and hypopituitarism

Received: 21 July 1998
Accepted: 11 February 1999

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**Abstract** *Background.* The pathogenesis of congenital hypopituitarism is unknown in many cases. *Objective.* We report a case of congenital pan-anterior hypopituitarism in association with a complex vascular abnormality involving the central nervous system, nasal pyriform aperture stenosis, and a single central maxillary incisor. *Materials and methods.* MRI and MRA were used to define this patient's complex vascular anomaly. *Results.* The vascular abnormality consists of absence of the right common carotid artery, the right internal carotid artery, the A1 segment of the right anterior cerebral artery, the anterior communicating artery, and partial absence of the M1 segment of the right middle cerebral artery. *Conclusion.* This unusual vascular anomaly may contribute to the pathogenesis of some cases of congenital hypopituitarism and related midline abnormalities, or may result from a common defect that causes pituitary insufficiency.

## Introduction

Congenital hypopituitarism may be seen in association with a small pituitary gland and absent pituitary stalk [1]. We present clinical and magnetic resonance (MR) findings in an infant who had, in addition to hypopituitarism, nasal pyriform aperture stenosis, a single central maxillary incisor, and an unusual vascular anomaly involving the central nervous system. We postulate that one cause of pituitary hypoplasia is a disturbance of the pituitary blood supply during organogenesis.

## Case report

The patient was a female infant born vaginally after a 36-week uncomplicated pregnancy to a 21-year-old G1P1 mother; birth weight was 2952 g. The infant had respiratory distress due to narrowing of the nasal airway and was intubated. Computed tomography (CT) showed nasal pyriform aperture stenosis, which was worse on the left side. Nasal dilatation and stent placement were performed, resulting in partial improvement. The patient remained in the neonatal intensive care unit for 6 weeks. Following discharge from the hospital, she grew poorly in both weight and length. At age 5 months, the patient was rehospitalized after a hypoglycemic seizure. Despite intravenous glucose therapy, she continued to have intermittent hypoglycemia. Physical examination revealed a weight and length both significantly below the 5th percentile. There were no dysmorphic features. There was mild hepatomegaly, and the genitalia were normal. Neurological examination showed marked axial hypotonia with complete head lag and no spontaneous lifting of the head from supine or prone positions. Developmental age based on the Gesell developmental schedules was 13 weeks. Endocrinological evaluation showed central hypothyroidism, cortisol deficiency, and growth hormone deficiency, but no evidence of diabetes insipidus. The patient also had slight anemia, total and direct hyperbilirubinemia, and mild elevations of her transaminase levels. The child was started on GH, hydrocortisone, and thyroxine replacement. Normoglycemia was achieved and maintained, and the patient was discharged after 8 days. A single central maxillary incisor erupted at 15 months. At age 31 months, her weight was at the 10th percentile, her length was at the 25th percentile, and she had no developmental delay.

## Methods

MR imaging was performed at 5 months of age with a Siemens Magnetom Vision 1.5 T; image sequences: axial TR/TE 600/14, 2000/80, coronal 400/14, and sagittal 400/14 and axial 600/14 and

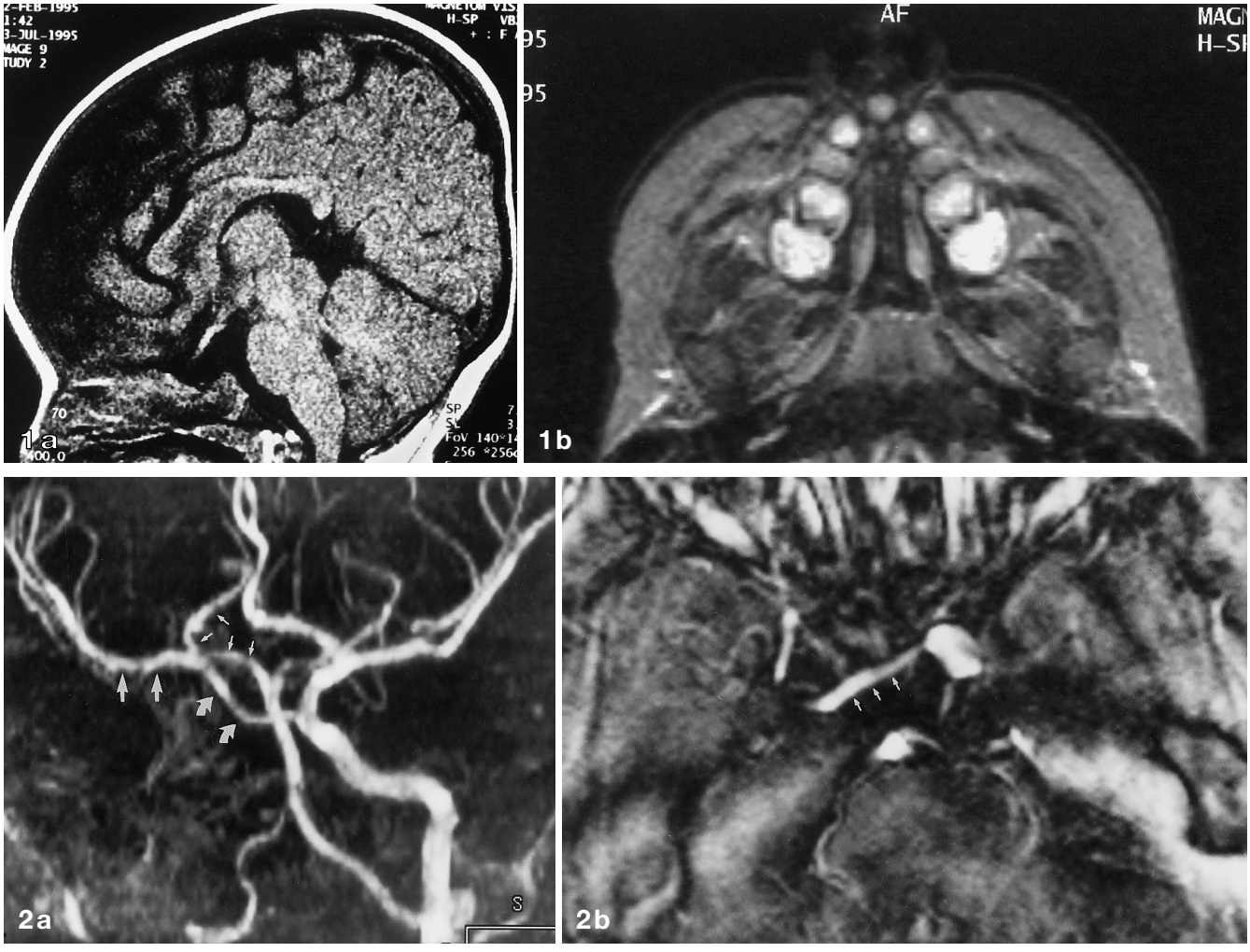


**Fig. 1 a, b** MRI. **a** T1-weighted (600/14) mid-sagittal image shows a very small amount of pituitary tissue in the base of the sella turcica. The pituitary stalk and the posterior-pituitary bright spot are absent. **b** T2-weighted (2000/810) axial image demonstrates a single central maxillary tooth bud (single incisor)

**Fig. 2 a, b** MRA. **a** AP view of 2D time-of-flight (78/10.4/70) MR angiogram shows absence of the right common carotid artery, the right internal carotid artery (ICA), the A1 segment of the right anterior cerebral artery, the anterior communicating artery, and partial absence of the M1 segment of the right middle cerebral artery (MCA). The intercavernous anastomosis *(curved arrows)* links the left ICA to the remaining M1 segment *(large arrows)* of the right MCA. The normal right posterior cerebral artery is indicated for clarity *(small arrows)*. **b** Axial view of a 3D time-of-flight (42/4.5/20) MR angiogram depicts an intercavernous vascular anastomosis *(small arrows)* between the cavernous portion of the left ICA and the right M1 segment, as well as absence of the right posterior communicating artery

coronal 400/14 before and after the administration of gadopentetate dimeglumine, respectively. An MR angiogram was performed at 15 months of age with a GE Signa 1.5 T, 2D, and 3D time-of-flight 78/10.4, flip angle 70, 42/4.5, flip angle 20, phase contrast, 29/9.5, flip angle 20.

## Results

On MR imaging, there was absence of the pituitary stalk and the posterior-pituitary bright spot. A minimal amount of pituitary tissue was present in the base of the sella turcica (Fig. 1 a). Myelination was mildly delayed. A single central maxillary tooth bud was present (Fig. 1 b). Absence of the signal void in the right internal carotid artery (ICA) prompted further evaluation with an MR angiogram that showed absence of the right common carotid artery, the right ICA, the A1 segment of the right anterior cerebral artery, the anterior communicating artery, and partial absence of the M1 segment of the right middle cerebral artery (MCA), all on

the right. An intercavernous vascular anastomosis linking the cavernous portion of the left ICA and the remaining M1 segment of the right MCA was seen (Fig. 2 a, b). The right A2 segment originated from the left A2 segment. Review of the head CT scan that had been performed previously revealed hypoplasia of the right carotid canal.

## Discussion

We report the first case of congenital pan-anterior hypopituitarism in association with a complex vascular abnormality involving absence of the right ICA and portions of its major branches, nasal pyriform aperture stenosis, and a single central maxillary incisor. Three previous cases of congenital hypopituitarism with absent ICA alone have been reported [1–3]. In addition, Lo et al. [4] reported one case and reviewed four others with nasal pyriform aperture stenosis, pituitary hypofunction, and a single maxillary incisor, which may represent a mild form of holoprosencephaly. Three of these latter patients had complete absence of the pituitary gland, with no information provided about associated vascular anomalies.

With the advent of MR imaging, the detection rate of specific radiological abnormalities in children with congenital hypopituitarism has increased significantly. These include decreased pituitary tissue in the sella turcica; interrupted, hypoplastic, or absent pituitary stalk; and an absent posterior-pituitary bright spot with or without ectopia typically near the median eminence [1]. Despite this array of possible MR findings, the proximate cause of congenital hypopituitarism (with or without actual MR abnormalities) is unclear in many cases. An increased incidence of birth asphyxia and breech delivery has been noted in these patients. It has been suggested that transient disruption of the blood supply to the pituitary and/or hypothalamus in the perinatal period can cause permanent damage and poor function. Another theory is that the pituitary stalk may be transected by the posterior clinoid plate or diaphragma sella during breech delivery. Congenital hypoplasia or dysplasia of pituitary and/or hypothalamus has also been implicated as a plausible mechanism.

The constellation of findings in our patient underscores the importance of an adequate blood supply to the developing pituitary gland and hypothalamus. ICA branches to the adenophypophyseal pocket develop on the 28th day of gestation. The formation of a capillary plexus supplied by a branch of the ICA on the external surface of the diencephalon precedes development of the primitive infundibular process [5]. In the case of an embryonic insult to the ICA resulting in vascular agenesis or aplasia, collateral sources of blood supply will form which, in our patient, involve an anomalous intercavernous vessel arising from the contralateral ICA. The developmental origin of one anomalous vessel is difficult to explain. One theory is that capillary branches of clival, capsular, or inferior hypophyseal arteries hypertrophy to form a single large vascular connection. Anterior-pituitary dysfunction in this situation may be related to the absence of the hypothalamic-hypophyseal-portal system.

In summary, vascular anomalies, while apparently rare, may play a causative role in some cases of congenital hypopituitarism or result from a common insult that affects midline structure development and pituitary function.

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