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Cutaneous Hemangiomas and Vascular Abnormalities: Persistence of Embryonic Vascularization

CASE REPORT

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SUMMARY: A 2-year-old girl presented with cutaneous facial, palpebral, back, perianal, and perineal hemangiomas and a subcutaneous neck and parotid voluminous hemangioma, associated with several extracranial vascular abnormalities, such as the absence of the ipsilateral internal carotid artery and hypoplasia of the common carotid arteries. Both vertebral arteries showed an embryonic appearance, without completing their maturation and without obliteration of all the right and left longitudinal neural arteries.

The association of facial hemangioma and abnormalities of the intra- or extracranial vessels was first described in 1978.¹ The disorder, known as “cutaneous hemangiomas: vascular anomaly complex,”^{2,3} PHACE,⁴ and Pascual-Castroviejo type II syndrome (PCII S),⁵ is possibly the most common neurocutaneous disorder. Persistence of the trigeminal artery and hypoplasia or absence of 1 carotid and/or vertebral artery is the most frequent congenital vascular malformation.^{2,3} We report a girl with cutaneous and subcutaneous hemangiomas in the face and in other parts of the body, who was studied by MR angiography (MRA), which revealed several rare embryonic vascular anomalies.

Case Report

A 2-year-old girl was first evaluated in our hospital at 1½ years of age because of facial hemangioma and left facial paresis. She was born at term with a weight of 3800 g after an uneventful pregnancy and delivery. During the first week of life, she had a cutaneous hemangioma in the left palpebral and forehead areas; and subcutaneous hemangiomas in the left parotid, retroauricular, and facial areas and in the back perianal and perineal areas. The hemangiomas showed a progressive growth and attained the largest size when she was 4 months of age, resulting in paresis of the facial area innervated by the mandibular nerve. The cutaneous hemangiomas were treated with laser therapy and corticoids administered orally during a 3-month period. Deep parotid and facial hemangiomas were treated with corticoids infiltrated locally. The palpebral and frontal superficial hemangioma disappeared almost completely in a short time. Deep parotid and facial hemangiomas presented recrudescence a short time after discontinuation of the corticoid therapy. After 1 year of age, all hemangiomas at all locations showed a slow regression. Psychomotor development was normal. The patient walked independently at 14 months of age and started to speak at 1 year. She had a normal vocabulary and pronunciation according to her age.

At 9 months of age, an intracranial MR imaging study showed normal supratentorial and infratentorial structures. MRA performed at 18 months of age revealed several intracranial and extracranial vascular abnormalities, including a voluminous hemangioma in the superficial and deep areas of the left neck and parotid, aplasia of the left internal carotid artery, hypoplasia of the left common carotid artery that arose in a low portion of the aortic arch, moderate hyper-

plasia of the right common and internal carotid arteries, and hypoplasia of the right external carotid artery (Fig 1A). Both anterior cerebral arteries arose from the right internal carotid artery, though this did not occur in the midline but in the left intracranial cavity, near the midline. Both vertebral arteries arose from the subclavian arteries and presented an embryonic appearance with persistence of all the cervical segmental arteries (Fig 1B, C). The left vertebral artery was larger than the right one. The basilar artery was normal and provided vascularization to the left cerebral hemisphere through the posterior communicating artery (Fig 2A, -B).

Discussion

The patient described in this article showed several of the already reported symptoms and signs,^{1,2} such as cutaneous and subcutaneous hemangiomas; agenesis and hypoplasia of some of the cerebral arteries; and other features, such as persistence of an embryonic state of the development of carotid and vertebral arteries. We are not aware of previous reports of these malformations.

The association of cutaneous hemangiomas with other intracranial or extracranial vascular and nonvascular anomalies has been known since 1978.¹ Mulliken and Glowacki (1982)⁶ differentiated 2 types of birthmarks, hemangiomas and vascular malformations, on the basis of endothelial characteristics. Hemangiomas are vascular tumors that have an initial period of evolution (increasing) and a second period of involution (decreasing), whereas the cutaneous vascular malformations change only their color or pigmentation but do not involute. Both vascular anomalies show some similarities and some differences.^{3,7} Similarities include the possibility of appearing anywhere in the body; the association with persistent embryonic arteries involving mostly the trigeminal artery; absence or hypoplasia of some of the main cerebral arteries, carotids, or vertebrals, most times ipsilateral to the facial vascular anomaly; cerebellar malformations; and, less often, of the cerebral hemispheres. The differences are that hemangiomas frequently are associated with subcutaneous internal organs, intracranial hemangiomas, or internal arteries with hemangiomatous and/or aneurysmal propensities that undergo the same process of increasing and regressing as the cutaneous hemangiomas,⁷ whereas the vascular malformations are not associated with these types of tumors.^{3,7}

Agenesis, aplasia, and hypoplasia of the internal carotid artery have been considered rare congenital anomalies. However, in the cutaneous hemangiomas and associated pathology (PCII S), absence or severe hypoplasia of an internal carotid artery has been described in 32%⁷ to 35%^{1,2} of patients in

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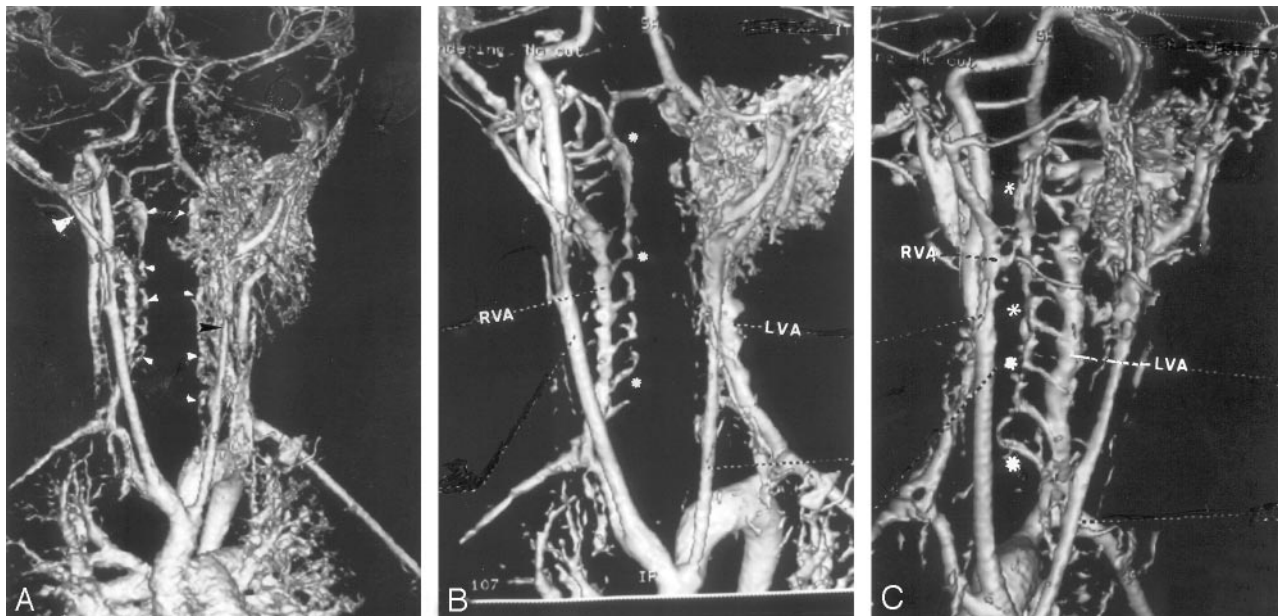


Fig 1. A, MRA in the coronal view shows a voluminous hemangioma in the left neck and parotid, a thin left common carotid artery that ends in the hemangioma (black arrowhead), right common and internal carotid arteries, and a hypoplastic external carotid artery (white arrowhead). Both vertebral arteries show an embryonic state with persistence of cervical segmental arteries (small white arrowhead). Both anterior cerebral arteries arise from the right internal carotid artery. B and C, The left (LVA) and right (RVA) vertebral arteries arise from the subclavian arteries but present an embryonic appearance, with persistence of all the cervical segmental arteries (arrowheads).

the largest series, in which persistence of the primitive trigeminal artery was found in 17%⁷ to 30%^{1,2} of patients. Absence of a carotid artery is compensated with collateral cerebral flow, mostly provided by the circle of Willis and via the persistent trigeminal artery.⁷

Collateral supply for the left cerebral hemisphere in our patient, who showed the absence of the left internal carotid artery, was accomplished through an ipsilateral hypertrophied posterior communicating artery that corresponded to the type 1A of the pathways of collateral blood flow described by Lie in 1968.⁸ The embryonic development of the cerebral arteries, including both their origin at the aortic arch and their intracranial trajectories, has been studied by several embryologists.^{9–11} The internal carotid artery appears in the embryo of 3 mm, and by 4 mm, this vessel begins to present an anterior division that is the outline of

the future anterior cerebral, middle cerebral, and anterior choroidal arteries. The posterior division gives origin to the posterior communicating and posterior cerebral arteries in the embryo of 5.3 mm. The common carotid artery is formed after the involution of the wall of the aorta between the third and the fourth arches in the embryo of 12–14 mm (35–36 days). The basilar artery is completely formed when the embryo has reached 7–12 mm (4½–5 weeks) by consolidation of the walls of the longitudinal neural arteries. The basilar artery can exist without the vertebral arteries having been formed.

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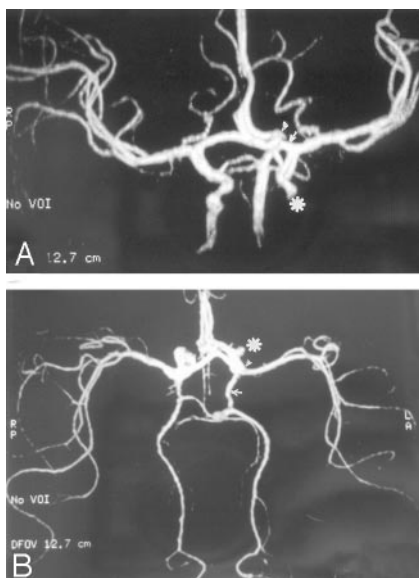


Fig 2. MRA. A and B, Coronal and axial views show both anterior cerebral arteries arising from the right internal carotid artery (arrowhead) in a very lateralized position into the left hemispherical cavity and the left carotid artery, communicating with the basilar artery through a large posterior communicating artery (arrow). Only the intracranial part of the left internal carotid artery is visible (asterisk).