Hemangiomas of the head, neck, and chest with associated vascular and brain anomalies: a complex neurocutaneous syndrome.

I Pascual-Castroviejo, J Viaño, F Moreno, R Palencia, V Martínez Fernandez, S I Pascual-Pascual, A Martínez-Bermejo, J J García-Peñas and M C Roche

AJNR Am J Neuroradiol 1996, 17 (3) 461-471
http://www.ajnr.org/content/17/3/461

Hemangiomas of the Head, Neck, and Chest with Associated Vascular and Brain Anomalies: A Complex Neurocutaneous Syndrome

Ignacio Pascual-Castroviejo, Juan Viaño, Felipe Moreno, Rafael Palencia, Vicente Martínez Fernandez, Samuel I. Pascual-Pascual, Antonio Martínez-Bermejo, Juan J. García-Peñas, and María Carmen Roche

PURPOSE: To describe the vascular and nonvascular intracranial and extracranial anomalies associated with hemangiomas and vascular malformations of the face, neck, and/or chest. **METHODS:** Seventeen patients had a physical examination and imaging studies consisting of one or more of the following: pneumoencephalography, conventional carotid and vertebral arteriography, CT, MR imaging, and MR angiography. **RESULTS:** Conventional arteriography revealed persistence of the trigeminal artery in 5 cases, absence of internal or external carotid and/or vertebral arteries in 11 cases, persistence of intervertebral arteries in 1 case, deformities of the aortic arch in 3 cases, and anomalies of the intracranial arteries in 3 cases. MR angiography revealed persistence of the trigeminal artery in 1 case in which conventional arteriography failed to show the malformation, and permitted visualization of narrowing of the intracranial arteries. CT and MR imaging showed a cerebellar anomaly in 8 cases and cerebral cortical dysplasia with cerebral hemispheric hypoplasia in 1 case. Vascular and nonvascular anomalies appeared ipsilateral to the external vascular abnormalities in most cases. **CONCLUSION:** This study demonstrates the association of cutaneous angiomas with anomalies affecting intracranial and extracranial arteries, the cerebellum, and, less frequently, the cerebral hemispheres and aortic arch. This association constitutes a relatively frequent neurocutaneous disorder, which we call the *cutaneous hemangioma–vascular complex syndrome*.

Index terms: Face, neoplasms; Neck, neoplasms; Hemangioma; Phakomatoses; Children, diseases

AJNR Am J Neuroradiol 17:461–471, March 1996

Despite the high frequency of hemangiomas of the head and neck—about 11% (1) to 38% (2) of all cutaneous hemangiomas—the possible associated anomalies involving the central nervous system (CNS) were ignored until a few years ago. Previous reports (3, 4) described a syndrome in which the main features were female predominance; facial hemangioma usually located in the area of the first sensory trigeminal branch; presence of diverse embryological abnormalities of the intracranial and extracranial arteries; and a variable degree of cerebellar hypoplasia. These findings were first documented by conventional arteriography and computed tomography (CT). In recent years, the collection of cases has been expanded with the use of magnetic resonance (MR) imaging and MR angiography, including reports of hemangiomas in a variety of locations in the body, which provides a more complete picture of the magnitude of the disease. In this article, we present the clinical and imaging findings in 17 patients, and suggest that this disorder that we have called *cutaneous hemangioma–vascular*

Received April 12, 1995; accepted after revision September 25.

Supported by Caja Madrid.

From the Services of Pediatric Neurology (I.P-C., S.I.P-P., A M-B., M.C.R.) and Pediatric Cardiology (F.M.), Hospital Infantil La Paz, Madrid, Spain; the Section of Magnetic Resonance Imaging, Sanatorio Nuestra Señora del Rosario, Madrid, Spain (J.V., V.M.F.); the Section of Pediatric Neurology, Hospital Universitario, Valladolid, Spain (R.P.); and the Service of Pediatric Neurology, Hospital Torre Cárdenas, Almeria, Spain (J.J.G-P.).

Address reprint requests to Ignacio Pascual-Castroviejo, MD, Service of Pediatric Neurology, Hospital Infantil La Paz, Paseo de la Castellana 261, Madrid 28046, Spain.

AJNR 17:461–471, Mar 1996 0195-6108/96/1703–0461

complex syndrome be included as a neurocutaneous syndrome.

Materials and Methods

Clinical and imaging studies using pneumoencephalography, conventional arteriography, CT, MR imaging, and MR angiography were carried out in 17 patients (12 girls, 5 boys; age range, 1 month to 5 years at the time of initial examination) between 1966 and 1994. The children all had hemangiomas, located mainly on the face but also on the neck and chest. The main reason for neurologic consultation was the facial hemangioma and, secondarily, psychomotor delay. The initial studies consisted of the clinical examination, pneumoencephalography in 3 patients, CT in 6, conventional arteriography in 13, MR imaging in 8, and MR angiography in 6. Four of the patients (cases 1, 6, 7, and 11) examined by MR angiography had been studied by conventional arteriography several or many years earlier. Findings in cases 1 through 7 have been previously published (3–5).

Conventional arteriography was performed using a brachial catheter in a few patients and femoral catheterization in most. MR imaging and MR angiography were performed on a 1.5-T system. The MR angiographic studies were done using the following parameters: For the intracranial structures, three-dimensional time-of-flight imaging was done at 47/5.5/1 (repetition time/echo time/excitations); 20° flip angle; field of view, 13 cm; section thickness, 1.2 mm; and matrix, 512 × 256. For the carotid bifurcation, two-dimensional time-of-flight imaging was done at 43/8/1, 50° flip angle; field of view, 16 cm; section thickness, 1.9 mm; and matrix, 256 × 192.

Results

In 15 patients, a large portion of the hemangioma involved the facial and scalp dermatome innervated by the first sensory trigeminal branch. Only cases 12 and 17 had hemangiomas completely outside the zone innervated by the first sensory trigeminal branch. In 3 patients (cases 2, 8, and 14) the external vascular anomaly also involved other regions of the body.

A summary of clinical and imaging findings in all cases is given in Table 1. Vascular abnormalities are presented in Table 2, and nonvascular anomalies in Table 3.

Arterial malformations were detected with conventional arteriography and with MR angiography. Two types of anomalies appeared most frequently. They were persistent embryologic arteries and agenesis of some major arteries. Persistence of the trigeminal artery was found in five patients. Although conventional arteriography showed the vascular malformation in all but one case, MR angiography similarly showed the vascular malformations. In one case, MR angiography showed the persistent trigeminal artery after conventional arteriography had failed to show this vessel many years earlier (Fig 1). In this imaging study, we also were able to observe the diminution of the thickness and tortuosity of intracranial arteries, which showed a narrower lumen in the adult than in the child. The persistent trigeminal artery was detected ipsilateral to the facial angioma in all cases. However, in some patients, the vertebral artery, which connects with the trigeminal artery, had its origin on the contralateral side and crossed at the level of the clivus (Fig 2).

Absence of the internal carotid artery (Fig 3) was found in four cases and severe hypoplasia of one internal carotid artery was discovered in two cases. We believe that in case 17 there was hypoplasia of the internal carotid artery, but only images from a cardiac catheterization were available, which showed the aortic arch and the origin of the cerebral vessels.

Absence of an external carotid artery was seen in one patient. The vascular anomaly, however, was not ipsilateral to the facial angioma (Fig 4A). There were small branches that supplied the face and originated directly from the internal carotid artery.

The vertebral artery was absent in six patients. This vascular anomaly appeared ipsilateral to the hemangioma in all patients except in cases 6 and 9, in which the vertebral artery was absent on the contralateral side (Fig 1E and F). In these cases, however, the facial zone innervated by the third branch of the trigeminal nerve was partially occupied by hemangioma.

Other less severe vascular anomalies were seen in isolated cases. These included the presence of segmental intervertebral anastomoses; arterial angiomatous malformation involving the territory of the left carotid siphon with hypoplasia of the left posterior inferior cerebellar artery (PICA); extremely thin appearance of one posterior cerebral artery; and a common origin for both anterior cerebral arteries that corresponded to the same side as the facial hemangioma. The hemangiomas were supplied primarily by hypertrophied branches of the external carotid artery, the subclavian artery, or the ophthalmic artery in most cases.

Eight patients had cerebellar anomalies, which ranged from hemispheric hypoplasia with

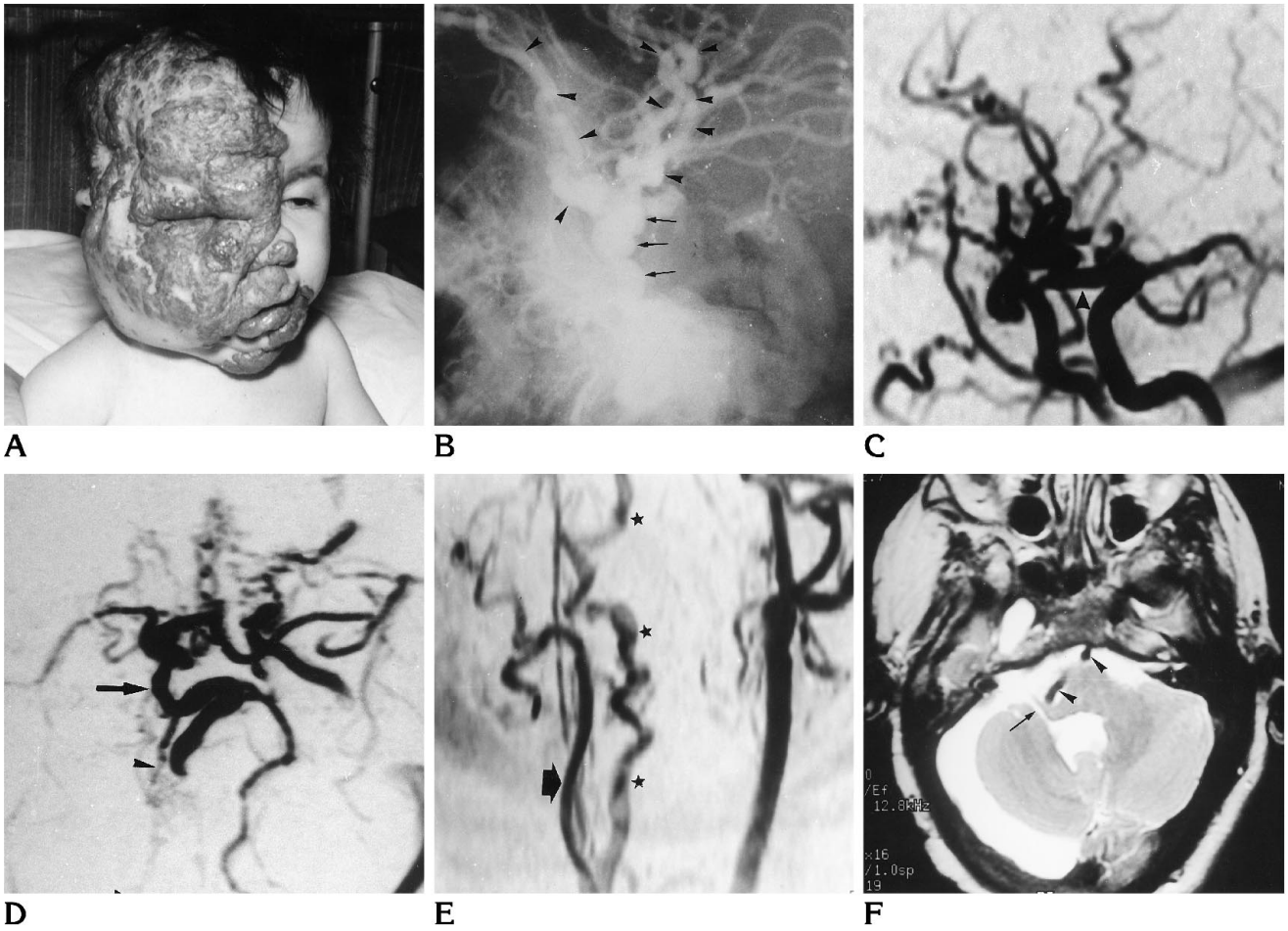

Fig 1. Case 6.

A, Photograph of 12-month old girl with a cavernous hemangioma shows involvement of all of the right hemiface and the left lower lip.

B, Conventional arteriogram in sagittal view shows an aneurysmal dilatation of the internal carotid artery (*arrows*) and most of its branches (*arrowheads*).

C and *D,* MR angiograms obtained when the patient was 28 years old. Three-dimensional arterial phase-contrast MR angiogram (velocity encoding, 60 cm/s; 25/8/1; flip angle, 20°) in sagittal view (*C*) shows the presence of a trigeminal artery (*arrowhead*) and disappearance of the aneurysmal image of the internal carotid artery and its branches seen on the conventional arteriogram. Three-dimensional phase-contrast MR angiogram (velocity encoding, 60 cm/s; 25/8/1; flip angle, 20°) in axial view (*D*) shows the trigeminal artery (*arrow*) and a decrease in the caliber of the vessels in the right cerebral hemisphere, especially in the posterior cerebral artery (*arrowheads*) and in the distal branches of both middle cerebral arteries.

E, Two-dimensional time-of-flight arterial MR angiogram (43/8/1; flip angle, 50°) in the coronal view at the level of the neck. On the right side, note irregularly enlarged vertebral artery (*stars*) and a thin carotid artery (*arrow*). On the left side, note a thick carotid artery and absence of vertebral artery.

F, Fast spin-echo axial MR image 3500/100/2 of the posterior fossa shows hypoplasia of the right cerebellar hemisphere; the fourth ventricle is open to the cisternal spaces through its inferior right lateral zone (*arrow*); the only vertebral artery image is also seen (*arrowheads*).

a possible small arachnoidal cyst without a defect of the posterior midline of the vermis (Fig 1F) to a major defect of the posterior midline of the cerebellum, constituting a Dandy-Walker malformation with hypoplasia of one hemisphere (Fig 5) or of both. The possibility of a larger number of cases with cerebellar malformation cannot be discounted, because CT, MR imaging, and pneumoventriculography were not performed in cases 2, 10, 12, 16, and 17. The unilateral hemispheric cerebellar hypoplasia appeared ipsilateral to the external hemangioma in all cases. Vascular deficiency related to the cerebellar anomaly was seen only in patient 5, who had a hypoplastic PICA ipsilateral to the hemispheric cerebellar hypoplasia.

TABLE 1: Clinical imaging and cardiologic abnormalities in 17 patients with 'cutaneous hemangioma–vascular complex syndrome'

Case	Sex/ Age	Type of Vascular Disease and Cutaneous Location	Imaging Studies Performed	Vascular Malformation	Other CNS	Cardiac Malformation	Present Age and Neurologic Sequelae
1	F/20 mo	Cavernous hemangioma; right side of scalp	CA, MR, MRA	Right trigeminal artery	None	None	20 y, retarded
2	F/5 y	Capillary angioma; left V1, external genitalia, feet	CA	Segmental intervertebral anastomoses	Not evaluated	Patent ductus arterious; aortic arch coarctation	25 y, borderline mental level
3	F/6 mo	Capillary angioma; left V1 and V2, right V3	CA, CC, CT	Absence of left ICA and VA; right VA supplies left trigeminal artery	Left hemispheric cerebellar hypoplasia	Triscuspid atresia; aortic arch atresia	Died at age 4 y
4	F/7 mo	Capillary angioma; left V1, V2, V3	PNEG, VCG, CA	Absence of left ICA and VA; vascularization via right ICA with angiomatosis and abnormal distribution	Dandy-Walker syndrome; dysplastic left ear	None	Lost to follow-up at 1 y of age
5	F/3 mo	Capillary angioma; left V1, V2, V3	CA, CT	Arterial angiomatous malformation in left carotid siphon; hypoplasia of left PICA	Hypoplasia left cerebellar hemisphere	None	21 y, borderline mental level
6	F/1 y	Cavernous hemangioma; right V1, V2, V3, and left V3	CA, CT, MR, MRA	Absence of left VA; right ICA hypoplastic; right trigeminal artery; hypoplastic right PCA	Thin pariental cortex in both hemispheres; Dandy-Walker malformation; right cerebellar hypoplasia	None	28 y, borderline mental level
7	F/3 y	Cavernous hemangioma; left V1	CA, CT, MR, MRA	Absence of left ICA	Absence of foramen lacerum; left cerebral hypoplasia and cortical dysplasia; left cerebellar hypoplasia	None	12 y, right hemiparesis, borderline mental level
8	F/3 mo	Cavernous hemangioma; left V1 and V2, left side of thoraxic and upper limb	CA, MR	Absence of left PCA	None	None	2 y, mildly retarded
9	F/6 mo	Cavernous hemangioma; right side of face, right V1, V2, V3, and left V3	CA, PNEG	Absence of the left VA; right trigeminal artery	Right cerebellar hypoplasia	None	21 y, borderline mental level

TABLE 1: Continued

Case	Sex/ Age	Type of Vascular Disease and Cutaneous Location	Imaging Studies Performed	Vascular Malformation	Other CNS	Cardiac Malformation	Present Age and Neurologic Sequelae
10	F/3 mo	Cavernous hemangioma; left V1 and V2	CA	Absence of the left VA	Not evaluated	None	23 y, borderline mental level
11	F/1 y	Cavernous hemangioma; left V1	CA, CT, MR, MRA	Left trigeminal artery; both ACA originated from the left ICA	Left brain hemiatrotrophy; right frontal lobe atrophy; corpus callosum atrophy	None	15 y, severe mental retardation
12	F/1 mo	Cavernous hemangioma; right V2	CA	Absence of the right external carotic artery	Absence of right ear	None	24 y, borderline mental level
13	M/6 mo	Capillary angioma; left V1	MR, MRA	None	Left hemispheric cerebellar hypoplasia	None	1½ y, none
14	M/4 mo	Right V1, V2, right side of neck	CT, MR	Not evaluated	Right cerebellar hypoplasia; Dandy-Walker syndrome	None	Lost to follow-up at 1 y of age
15	M/3 mo	Cavernous hemangioma; right V1 and V2	MR, MRA	Not evaluated	Dandy-Walker syndrome; right cerebellar hypoplasia	None	3 y, mildly retarded
16	M/2 mo	Cavernous hemangioma; bilateral V1 and V2, right mandibula and shoulder, left side of chest	CA	Absence of the left ICA	Not evaluated	None	Lost to follow-up at 1 year of age
17	M/1 mo	Cavernous hemangioma; left side of chest	CC	Absence of the left VA; hypoplastic left ICA	Intraabdominal hemangioma	Aortic arch coarctation	Died at age of 4 mo after aortic and intestinal surgery

Note.—ACA indicates anterior cerebral artery; CA, conventional arteriography; CC, cardiac catherism; CT, computed tomography; ICA, internal carotid artery; MR, magnetic resonance; MRA, magnetic resonance arteriography; PCA, posterior cerebral artery; PICA, posterior inferior cerebellar artery; PNEG, pneumoencephalography; VA, vertebral artery; VCG, ventriculography.

TABLE 2: Vascular abnormalities among 17 patients with 'cutaneous hemangioma–vascular complex syndrome'

Abnormality	No. of Cases
Persistence of the trigeminal artery	5
Absence of the internal carotid artery	4
Hypoplasia of the internal carotid artery	2
Absence of the external carotid artery	1
Absence of the vertebral artery	6
Presence of segmental intervertebral anastomosis	1
Intracranial angiomatous arteries	1
Hypoplastic posterior cerebral artery	1
Common origin for both cerebral arteries	1
Hypoplastic posterior inferior cerebellar artery	1
Aortic arch coarctation	2
Aortic arch artresia	1

In case 7, the left carotid artery was absent, and there was vascularization of both cerebral hemispheres through the right carotid artery; this child also had cerebral cortical dysplasia and cerebral hemispheric hypoplasia on the same side as the facial hemangioma and absence of the internal carotid artery (Fig 6). Bilateral cerebral and corpus callosal atrophy were seen in patient 11, who had cerebral palsy associated with birth hypoxia. Mental status was low for all patients, with the IQ at borderline normal or lower.

Discussion

Most large series of hemangiomas, including those located in the head and neck (1, 2, 6–9), have focused on their external appearance, histologic nature, and extraneurologic complications. Hemangiomas have been associated with skeletal changes (1), sternal malformation (10), constitutional deformities (8, 9), coarctation of the aorta (11, 12), midabdominal raphe (13), and sacral and genitourinary defects (14). Most often they are related to cosmetic abnormalities.

After an early report associated facial capillary hemangiomas with extracranial and intracranial vascular anomalies, cerebellar malformations, and cardiopathies with aortic arch deformities (3), several articles appeared in which isolated cases or small series with similar findings were described (15–19). Vascular anomalies are the most frequent malformation associated with cutaneous hemangioma–vascular complex syndrome. Absence of the internal carotid and/or vertebral arteries and persistence of the trigeminal artery are the most common malformations. In our series, they were ipsilateral to the external angioma in all cases except in patients 6, 9, and 12, who had agenesis of the contralateral vertebral artery (cases 6 and 9) or of the contralateral external carotid artery (case 12). In case 12, the vascular anomaly also could be related to agenesis of the ear, which occurred on the same side as the vascular agenesis. Conventional arteriography usually showed the vascular anomalies. With the advent of MR imaging and MR angiography, we were able to detect abnormalities that had not been discovered by conventional arteriography or by CT in two patients (cases 6 and 7). In case 6, we did not find persistence of the trigeminal artery until MR angiography revealed this vascular malformation 27 years later. In this case, apart from the presence of the trigeminal artery, we found an absence of the vertebral artery on the side contralateral to the facial hemangioma, and involution of the internal carotid artery and its branches ipsilateral to the facial hemangioma, which had appeared as aneurysmal vessels or megaarteries on a cerebral arteriogram obtained at 1 year of age (3). These changes of the cerebral vessels were similar to those observed in the facial angioma. In case 7, cerebral cortical dysplasia and cerebral hemispheric hypoplasia present on the same side as the agenesis of the internal carotid artery were shown on the MR study.

TABLE 3: Nonvascular abnormalities among 17 patients with 'cutaneous hemangioma–vascular complex syndrome'

Abnormality	No. of Cases
Cerebellar Anomalies	8
Dandy-Walker malformation	3
Cerebellar hemispheric hypoplasia	5
Cerebral cortical dysplasia	1
Cardiac malformations	2
Patent ductus arteriosus	1
Tricuspid atresia	1

Apart from our previous reports (3–5) and the present series, only isolated cases have been studied angiographically (15, 18), in which some vascular anomalies have been revealed. On the basis of these results, we believe that in patients with hemangioma of the face, scalp, neck, and chest, CT or MR studies should be performed together with conventional arteriography or, preferably, MR angiography. Vascular imaging should include the intracranial and extracranial vessels as well as the aortic arch.

All arteries that supply the brain, face, and

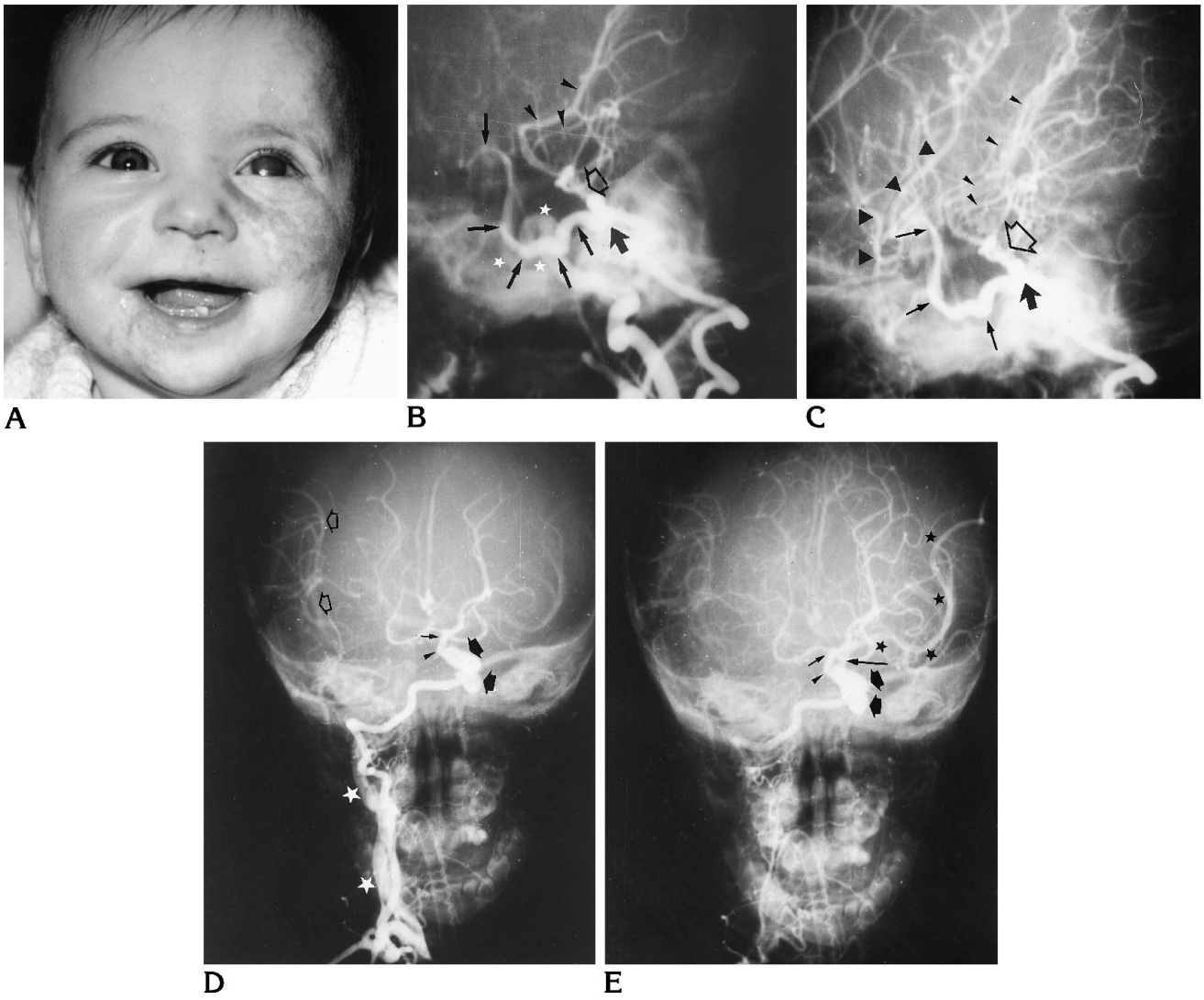

Fig 2. Case 3.

A, Photograph of a 6-month-old girl with capillary telangiectatic malformation involving the skin of the left facial zone innervated by the first and second sensory branches of the trigeminal nerve and a part of the right facial zone innervated by the third trigeminal branch.

B and *C,* Early and late phases of conventional arteriography performed in the lateral projection to obtain right carotid and vertebral arteriogram from right retrograde brachial injection. In the early phase (*B*), the carotid artery appears tortuous but shows a normal configuration at the carotid siphon (*stars*). The basilar artery bifurcates upon reaching the middle upper portion of the clivus (*solid thick arrow*), continuing its trajectory on one side (*open arrow*) to give rise to the superior cerebellar arteries and posterior cerebral arteries (*thin arrowheads*). It also gives rise to a thick and tortuous branch directed forward and superiorly (*solid thin arrows*) to form hemispheric ramifications, giving the impression that this last branch might be a persistent trigeminal artery that fills via the vertebral artery. In the late phase (*C*), the carotid artery is not seen, but the vertebrobasilar system shows along with filling of the middle cerebral artery (*thick arrowheads*) through the trigeminal artery (*solid thin arrows*).

D and *E,* Anteroposterior projection of the same study in early and late phases. The early (*D*) phase shows a normal distribution from the cerebral arteries (*open arrows*) arising from the right internal carotid artery and filling of the anterior cerebral arteries from the right carotid artery (*stars*). The basilar artery is situated far laterally to the left and bifurcates (*arrowhead*) to give continuity to the basilar artery on one side (*short thin arrow*) and, on the other side, to fill the left trigeminal artery (*thick arrows*) through which all left carotid branches except the anterior cerebral artery are supplied. The late phase (*E*) shows the same view of the vertebrobasilar system, but the superior portion of the trigeminal artery (*long arrow*) is seen as well as its continuity in the middle cerebral artery (*stars*).

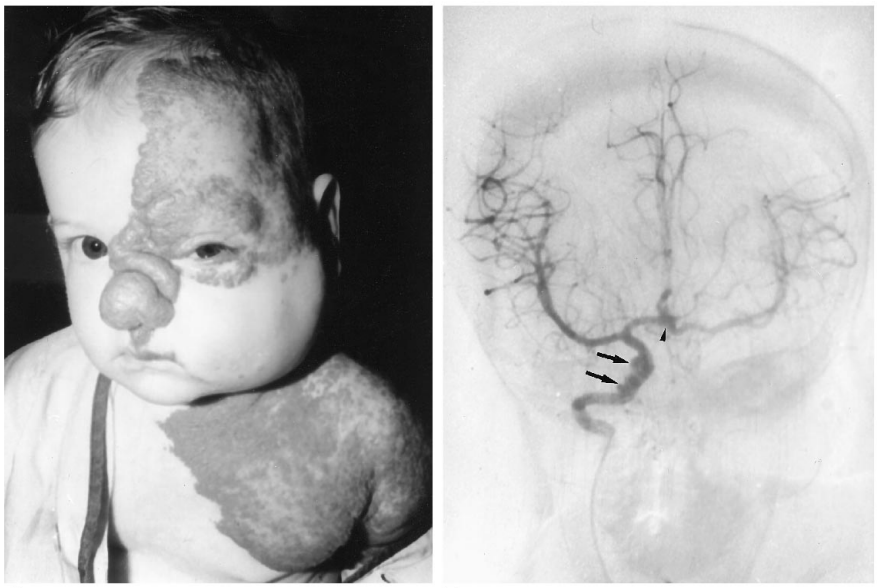

Fig 3. Case 8.

A, Photograph of a 6-month-old girl with cavernous hemangioma in the left facial zone innervated by the first and second branches of the trigeminal nerve, thorax, shoulder, and upper limb.

B, Cerebral arteriogram shows that both cerebral hemispheres are supplied by the right internal carotid artery through an enlarged anterior communicating artery (*arrowhead*). The carotid siphon has a very irregular appearance (*arrows*).

neck can be affected by angiomas of the face, neck, or chest. Primitive, internal and external carotid, and vertebral arteries (including their origin in the aortic arch), and even embryonic arteries that would disappear early in development, can be associated with these angiomas. To understand these vascular anomalies, it is necessary to place the embryonic development of the cerebral arteries, including both their origin at the aortic arch and their intracranial trajectories, into ontogenic perspective, a chronologic summary of which appeared in a previous article (4).

The most frequent vascular anomaly in patients with cutaneous hemangioma–vascular complex syndrome is persistence of the primitive trigeminal artery, which constitutes a carotid-basilar anastomosis. The primitive trigeminal artery may have received its name from its intimate relation with the gasserian ganglion (20). This assertion is confirmed by the findings observed in our patients for whom there was evidence of an association of the trigeminal artery and hemangioma in zones of the face and scalp innervated by the trigeminal nerve.

The trigeminal artery first appears when the embryo is 3 mm in size (3½ weeks). It is derived from the first aortic arch as the second of the three vascular arms; it passes dorsally, arching next to the medial zone of the gasserian ganglion. When the embryo reaches 4 mm (4 weeks), the trigeminal artery communicates with fragments of the walls of the neural longitudinal arteries, from which the basilar artery

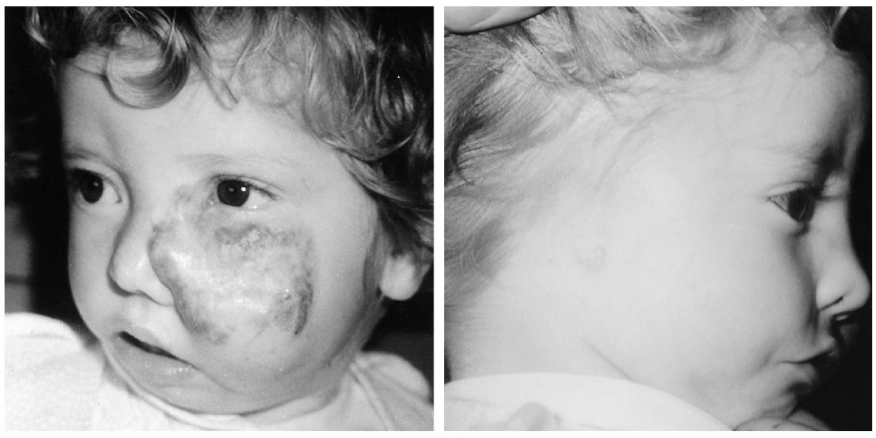

Fig 4. Case 12. Photographs of a 1-year-old girl with a hemangioma show central regression in the left cheek (*A*) and absence of the right ear (*B*).

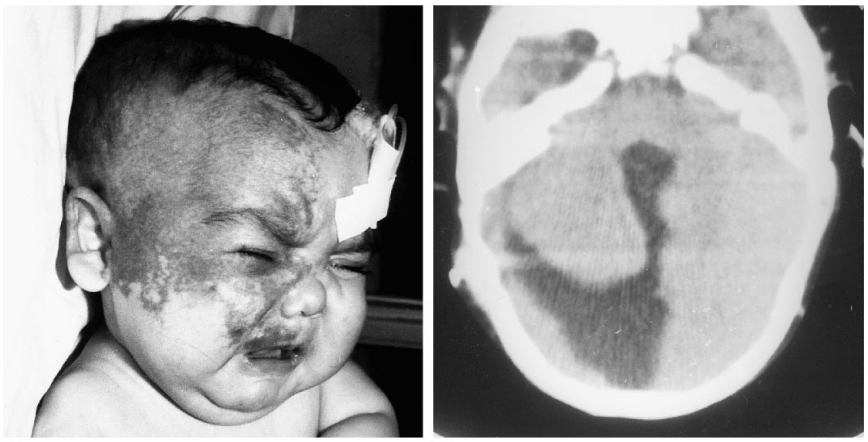

Fig 5. Case 14.

A, Photograph of a 4-month-old boy with capillary-venous stain involving the right facial zone innervated by the first and second sensory branches of the trigeminal nerve.

B, Axial CT scan shows a Dandy-Walker malformation with right cerebellar hypoplasia.

forms fully when the embryo is 7 to 12 mm (4½ to 5 weeks). The formation of the basilar artery is accompanied by the involution of the trigeminal artery or its annexation by the internal carotid artery, becoming complete in the embryo of 14 mm (5 to 5½ weeks). In the embryo of 9 to 11 mm (32 days), the completely formed basilar artery is still in an intermediate phase of anastomosis with the carotid and the vertebral systems. The vertebral arteries begin to form when the embryo has reached 9 mm (32 days) by longitudinal anastomosis between superior longitudinal segmental arms of the dorsal aorta; and their formation is nearly complete by the time the embryo measures 12.5 mm (35 days). But they still originate from the wall of the aorta. The maturity of the vertebral arteries is reached when the embryo is between 14 and 16 mm (36 to 40 days), with their origin having been displaced to the level of the ductus arteriosus. If the vascular development suffers arrest or occlusion between 32 and 36 days, the basilar artery could exist without the vertebral artery's having formed.

The internal carotid artery was absent in four patients and hypoplastic in two patients. This arterial anomaly was ipsilateral to the external angioma in all six cases. The internal carotid artery appears in the embryo at 3 mm, and by 4 mm this vessel begins to divide anteriorly, a division that will become 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. At this time, the anterior cerebral artery is well defined and extends from the third aortic arch. When the embryo is in the third and last phase of the brachial stage, with a length of approximately 7 to 12 mm (32 to 35 days), the internal carotid

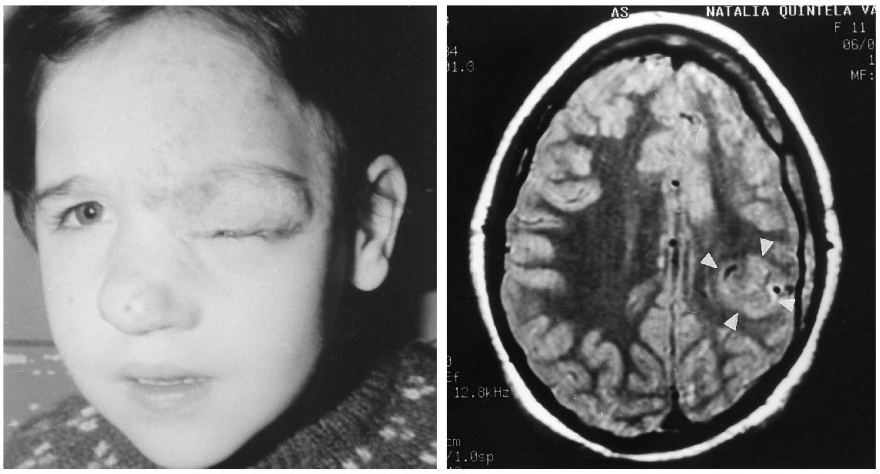

Fig 6. Case 7.

A, Photograph of a 3-year-old girl with a regressing hemangioma in the left facial zone innervated by the first sensory branch of the trigeminal nerve and nose.

B, Axial T2-weighted MR image (2000/100) obtained at 12 years of age shows a left hemispheric hypoplasia with cerebral cortical dysplasia (*arrowheads*).

artery is already filled with blood in its intracerebral course. The external carotid artery was absent in only one of our cases. However, it was discovered on the contralateral side of the facial angioma, although on the same side as an aplastic ear. Absence of the external carotid artery or some of its branches in mandibulofacial dysplasia with ear deformities was found in a large series of angiographic studies (21). There is disagreement with respect to the development of the external carotid arteries. Cogdon (22) concluded that they originate as an external growth from the aortic sac after the first and second aortic arches have disappeared. Their subsequent origin displaces the third aortic arch, becoming separated into a proximal part that forms the primitive carotid artery and a distal portion that gives origin to the proximal part of the internal carotid artery.

It is not difficult to distinguish the cutaneous hemangioma–vascular complex syndrome from other angioblastic neurocutaneous diseases, such as the Sturge-Weber syndrome and the Wyburn Mason syndrome. Cutaneous lesions are different in appearance (4). In Sturge-Weber syndrome, nevus flammeus consists of vascular ectasias that continue to increase in size with age. The location of all or part of the nevus flammeus in the zone corresponding to the first trigeminal branch is a fundamental characteristic of Sturge-Weber syndrome. Intracranial lesions in Sturge-Weber syndrome consist of leptomeningeal angiomatosis and progressive atrophy of the ipsilateral cerebral hemisphere. Arterial changes are rare, whereas absence of cortical veins and presence of deep hemispherical veins are common. The Wyburn Mason syndrome is not always accompanied by cutaneous nevus or angioma, but it usually includes angiomatous vessels in the retina. This syndrome always includes arteriovenous angiomas of the retina and cerebral cortex.

Dandy-Walker malformation has been related to angiomas located on the face or anywhere on the trunk or extremities (3, 4, 16, 17, 23–29). Less severe cerebellar defects, mostly of the type of unilateral hypoplasia located on the same side as the facial angioma, have been reported frequently (3, 4, 15, 17, 19). The time of intrauterine development of Dandy-Walker malformation remains uncertain, although the malformation of the posterior fossa and the associated anomalies suggest a teratogenic influence between embryonic weeks 4 and 6½. Ear malformations, exemplified in our study by case 4, correspond to a phase of development at the embryonic stage of 13 mm (6½ weeks' gestation), and the malformation in the origin of the carotid and vertebral arteries in the aortic arch corresponds to between 4½ and 6½ weeks' gestation, at which time the embryo measures 7 to 14 mm (4).

Segmental intervertebral anastomosis was seen in only one patient (case 2), who had an associated patent ductus arteriosus and aortic coarctation, and seems not to be related to the external hemangioma or vascular malformation but rather to a common finding in cardiac malformations. Aortic arch anomalies, atresia, or coarctation were found in three patients. Cardiac malformations associated with the aortic arch defects appeared in two cases. One patient had an intraabdominal hemangioma affecting an extensive part of the intestines.

As compared with the number of cases reported in a major series of patients with cutaneous hemangiomas (1, 7) and with those described in reports of the most frequent classic neurocutaneous diseases—neurofibromatosis type 1 (30), tuberous sclerosis (31), and nevus of Ito (32)—cutaneous hemangioma–vascular complex syndrome may be one of the most common neurocutaneous diseases.

Acknowledgment

We thank Harvey B. Sarnet, MD, (Seattle) for help in preparing the manuscript in an understandable English.

References

1. Boyd JB, Mulliken JB, Kaban LB, Upton J, Murray JE. Skeletal changes associated with vascular malformations. *Plast Reconstr Surg* 1984;74:789–795
2. Margileth AM, Museles M. Cutaneous hemangiomas in children. *JAMA* 1965;194:223–226
3. Pascual-Castroviejo I. Vascular and nonvascular intracranial malformations associated with external capillary hemangiomas. *Neuroradiology* 1978;16:82–84
4. Pascual-Castroviejo I. The association of extracranial and intracranial vascular malformations in children. *Can J Neurol Sci* 1985; 12:139–148
5. Pascual-Castroviejo I, Viaño J, Pascual-Pascual S-I, Martinez V. Facial hemangioma, agenesis of the internal carotid artery and cerebral cortex dysplasia: case report. *Neuroradiology* 1995;37: 692–695
6. Glowacki J, Mulliken JB. Mast cells in hemangiomas and vascular malformations. *Pediatrics* 1982;70:48–51
7. Finn MC, Glowacki J, Mulliken JB. Congenital vascular lesions: clinical application of a new classification. *J Pediatr Surg* 1983; 18:894–900

8. Burns AJ, Kaplan LC, Mulliken JB. Is there an association between hemangioma and syndromes with dysmorphic features? *Pediatrics* 1991;88:1257–1267
9. Baker LL, Dillon WP, Hieshima GB, Dowd CF, Frieden I. Hemangiomas and vascular malformations of the head and neck: MR characterization. *AJNR Am J Neuroradiol* 1993;14:307–314
10. Hersh JH, Woterfiell D, Rutledge J, et al. Sternal malformation/vascular dysplasia association. *Am J Med Genet* 1985;21:177–186
11. Schneeweiss A, Blieden LC, Shem-Tov A, Motro M, Feigel A, Neufeld HN. Coarctation of the aorta with congenital hemangioma of the face and neck and aneurysm or dilatation of subclavian or innominate artery: a new syndrome? *Chest* 1982;82:186–187
12. Vaillant L, Lorette G, Chantepie A, et al. Multiple cutaneous hemangiomas and coarctation of the aorta with right aortic arch. *Pediatrics* 1988;81:707–710
13. Igarashi M, Uchida H, Kajaii T. Supraumbilical midabdominal raphe and facial cavernous hemangiomas. *Clin Genet* 1985;27:196–198
14. Goldberg NS, Hebert AA, Sterly NB. Sacral hemangiomas and multiple congenital abnormalities. *Arch Dermatol* 1986;122:684–687
15. Mizuno Y, Kurokawa T, Numaguchi Y, Goya M. Facial hemangioma with cerebrovascular anomalies and cerebellar hypoplasia. *Brain Dev* 1982;4:375–378
16. Pascual-Castroviejo I, Velez A, Pascual-Pascual SI, Roche MC, Villarejo F. Dandy-Walker malformation: analysis of 38 cases. *Childs Nerv Syst* 1991;7:88–97
17. Reese V, Frieden IJ, Paller AS, et al. Association of facial hemangiomas with Dandy-Walker and other posterior fossa malformations. *J Pediatr* 1993;122:379–384
18. Murotani K, Hiramoto M. Agenesis of the internal carotid artery with a large hemangioma of the tongue. *Neuroradiology* 1985;27:357–359
19. Goh WHS, Lo R. A new 3 C syndrome: cerebellar hypoplasia, cavernous hemangioma and coarctation of the aorta. *Dev Med Child Neurol* 1993;35:637–641
20. Padget DH. The development of the cranial arteries in the human embryo. *Contrib Embryol* 1948;32:205–262
21. Palencia Luaces R, Pascual-Castroviejo I. Displasias maxilofaciales. *Bol Med Hosp Infant Mex* 1973;30:501–528
22. Congdon ED. Transformation of aortic arch system during the development of the human embryo. *Contrib Embryol* 1922;14:47–110
23. Dandy WE, Blackfan KD. Internal hydrocephalus: an experimental, clinical and pathological study. *Am J Dis Child* 1914;8:406–482
24. Taggart JK Jr, Walker AE. Congenital atresia of the foramens of Luschka and Magendie. *Arch Neurol* 1942;48:583–612
25. Hart MN, Malamud N, Ellis WG. The Dandy-Walker syndrome: a clinicopathological study based on 28 cases. *Neurology* 1972;22:771–780
26. Stoeter P, Marquardt B. Kombinierte Hirn-und Gefässmissbildung bei einem dreijährigen kind: Dandy-Walker syndrome: Arteria primitive trigemina und Hemangiom. *Fortschr Rontgenstr* 1978;128:680–683
27. Nova HR. Familial communicating hydrocephalus, posterior cerebellar agenesis, mega cisterna magna and portwine nevi. *J Neurosurg* 1979;52:862–865
28. James HE, Kaiser GL, Shut L, Bruce DA. Problems of diagnosis and treatment in the Dandy-Walker syndrome. *Childs Brain* 1979;5:24–30
29. Hirsch JF, Pierre-Kahn A, Renier D, Sainte-Rose C, Hoppe-Hirsch E. The Dandy-Walker malformation: a review of 40 cases. *J Neurosurg* 1984;61:515–522
30. Pascual-Castroviejo I. Complications of neurofibromatosis type 1 in a series of 197 children. In: Fukuyama Y, Suzuki Y, Kamoshita S, Casaer P, eds. *Fetal and Perinatal Neurology*. Basel: Karger, 1992;162–73
31. Gomez MR. Criteria for diagnosis. In: *Tuberous Sclerosis*, 2nd ed. New York: Raven Press, 1988;9–19
32. Pascual-Castroviejo I, Rodriguez-Lopez L, De la Cruz Medina M, Salamanca-Maesso C, Roche Herrero C. Hypomelanosis of Ito. Neurological complications in 34 cases. *Can J Neurol Sci* 1988;15:124–129