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Facial haemangioma, agenesis of the internal carotid artery and dysplasia of cerebral cortex: case report

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Abstract We describe a girl with a facial haemangioma, associated with other vascular anomalies: agenesis of the internal carotid artery, cerebral cortical dysplasia and hypoplasia of the cerebral hemisphere on the same side of the angioma. We studied the patient by conventional arteriography, T1- and T2-weighted MRI and magnetic resonance angiography.

Key words Facial haemangioma · Agenesis of internal carotid artery Cortical dysplasia · Magnetic resonance angiography

Introduction

The association of facial capillary haemangioma and abnormalities of the cerebral arteries in their intra- or extracranial course was first described in 1978 [1]. These abnormalities are always on the same side as the haemangioma and consist mainly of persistence of embryonic extracranial arteries, such as the intervertebral arteries or intracranial vessels such as the trigeminal artery, and absence of the internal carotid artery. Less frequently other malformations of the cerebellum, heart and aortic arch can be detected. The relationship between different types of facial angioma and vascular abnormalities and embryonic development was described in a later report [2]. The 8 female patients described in the first publication were studied by conventional transfemoral or brachial arteriography.

Magnetic resonance angiography (MRA) allows us to investigate the cerebral and cerebellar parenchyma, intra- and extracranial vessels, aortic arch and origin of cerebral arteries, all in a single examination. We report a girl studied by MRI and MRA in whom a facial haemangioma was accompanied by cerebral cortical dysgenesis, in addition to other commonly associated malformations [1, 2].

Case report

An 11-year-old girl was first studied in our service when she was 3 years old because of a facial haemangioma and delayed language acquisition. Pregnancy had come to full term without other complications than a small vaginal haemorrhage in the 3rd month. Delivery was normal and weight at birth was 3200 g. The family history disclosed a third-degree parental consanguinity, but two older brothers were normal.

An angiomatous spot was seen on the left frontal area at birth, extending in a few days over the upper eyelid and margin of the nose, causing deformity of the affected areas and closure of the left eye. The angioma was treated with corticosteroids, undergoing partial regression but reverting to its initial size upon cessation of therapy. Motor development was slightly delayed. The patient started to walk at 14 months of age but showing a significant lack of dexterity. Speech began at 2 years of age, with delay in acquisition of vocabulary and ability to pronounce. The patient suffered two generalized seizures when she was 1 year old, coincident with febrile episodes. She was treated with sodium valproate for 3 years without recurrence. CT at 18 months of age showed slight atrophy of the left cerebral hemisphere.

Examination at 3 years of age disclosed motor and language retardation. There was a capillary-cavernous haemangioma ex-

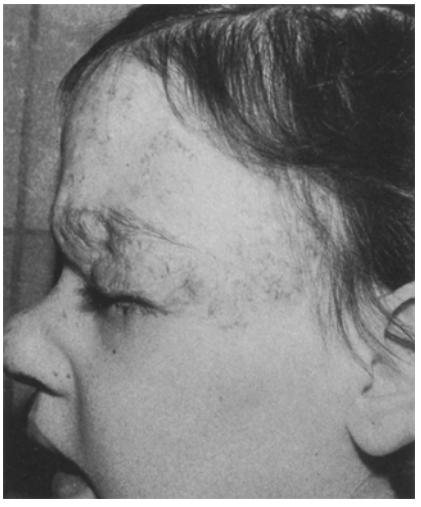


Fig. 1 Capillary-cavernous haemangioma on the left forehead, eyelids and nose

tending over the left forehead, eye and nose (Fig. 1). It was regressing but still covered both eyelids completely, causing convergent strabismus and loss of vision in the left eye. Funduscopy showed left optic atrophy. EEG and plain skull films were normal, although the basal projection revealed absence of the left carotid canal. Cerebral angiography showed no left internal carotid artery; both hemispheres were supplied through the right internal carotid artery (Fig. 2), which gave origin to the anterior and middle cerebral arterial systems of both sides, via an enlarged anterior communicating artery. The left external carotid artery was slightly enlarged and several hypertrophied branches supplied the facial angioma (Fig. 3), especially the vessel of the orbital region and the superficial temporal artery. The patient underwent plastic surgery on several occasions between the ages of 3 and 11 years, with significant improvement of her appearance. At the age of 11 she has a slight right hemiparesis and prefers to use her left hand. Her school performance is only slightly worse than that of other children her age.

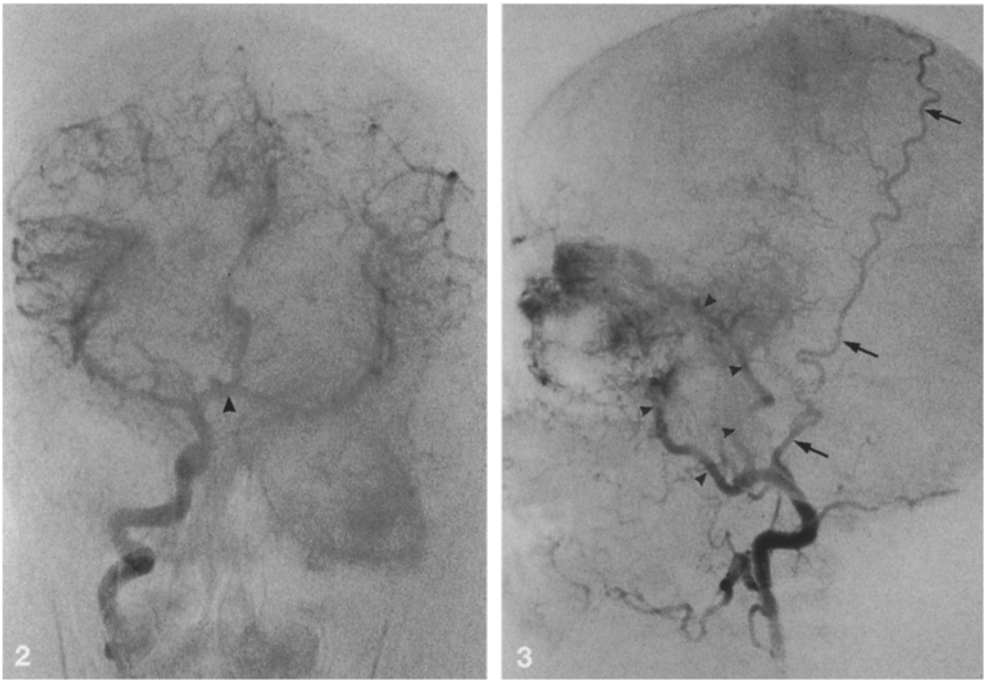
T1- and T2-weighted MRI showed mild left cerebral hypoplasia and a nodular area of cortical dysplasia criss-crossed by vessels in the left hemisphere (Fig. 4), and a primitive Sylvian sulcus, especially evident in superficial sagittal sections (Fig. 5). MRA showed images similar to those obtained by conventional angiography when the patient was 3 years old (Fig. 6). On the left the common carotid artery continued as the left external carotid artery, without the normal bifurcation into internal and external carotid arteries (Fig. 7). The vertebrobasilar arterial system was normal. The arch of the aorta and the origins of the carotid arteries were normal, although the left common carotid artery was narrower than the right (Fig. 8).

Discussion

A number of case reports and small series have appeared in which an angioma and one or more of the above-mentioned malformations were associated. Mouth or facial angiomas have been found in association with cerebellar defects, including the Dandy-Walker syndrome [3–5], absence of the internal carotid artery [6] and aortic arch malformations [7, 8]. The embryological explanation of these associations has been described [2]. The reason for the female predominance remains uncertain; a possible X chromosome alteration needs to be confirmed by molecular genetics.

Fig. 2 Cerebral arteriography. Both cerebral hemispheres are supplied by the right internal carotid artery through an enlarged anterior communicating artery *(arrowhead)*

Fig. 3 Left carotid arteriography showing only the external carotid artery. The haemangioma is supplied by two large branches *(arrowheads)* of the external carotid artery. Enlarged parietal branch of superficial temporal artery *(arrows)*



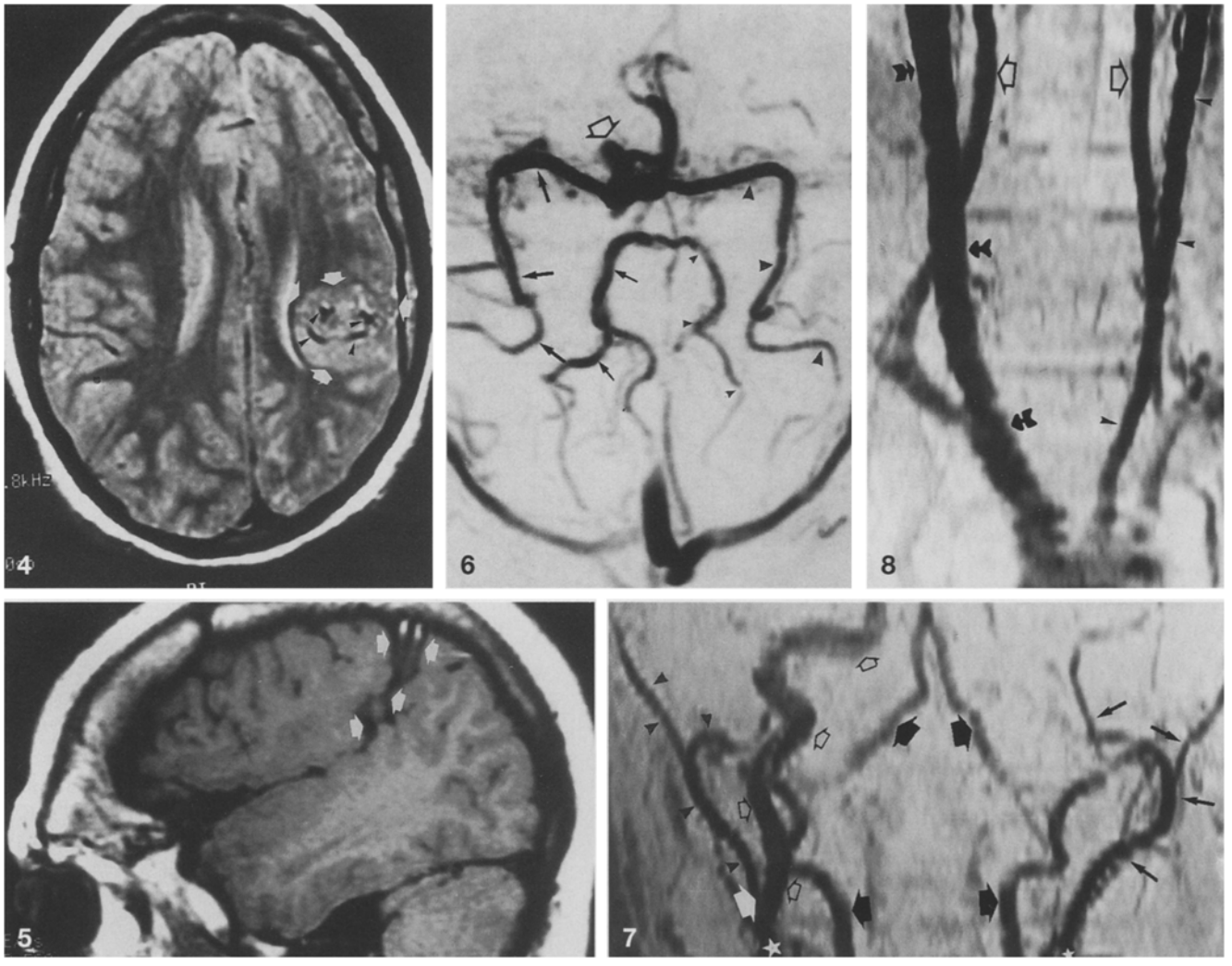


Fig. 4 Axial T2-weighted (200/100) image shows a left hemisphere mass *(arrows)*. Several large vessels *(arrowheads)* run within the infoldings of an abnormal grey matter mass. The left hemisphere is abnormally small

Fig. 5 Sagittal T1-weighted (680/25) image in a superficial plane reveals a primitive Sylvian sulcus *(arrows)*

Fig. 6 Axial MRA reveals that all the supratentorial arteries originate from the right internal carotid artery *(open arrow)*. The left middle *(large arrowheads)* and posterior *(small arrowheads)* cerebral arteries are smaller than the right middle *(thick arrows)* and posterior *(thin arrows)* cerebral arteries

Fig. 7 Coronal MRA of the neck. On the right, the common carotid artery *(star)* has a normal bifurcation *(thick white arrow)*, giving rise to a large internal carotid artery *(open arrows)* and a normal external carotid artery *(arrowheads)*. On the left, there is no internal carotid artery, and the external carotid artery *(arrows)* is the continuation of the common carotid *(small star)*. Both vertebral arteries *(thick black arrows)* appear normal

Fig. 8 Coronal MRA shows the arch of the aorta and the origin of both carotid and vertebral arteries. The right common carotid artery *(arrows)* is wider than the left *(arrowheads)*. The two vertebral arteries are the same size *(open arrows)*

It was difficult to study the complete intra- and extracranial course of the cerebral arteries, including their origin from the arch of the aorta in a single examination, and to obtain good-quality images by conventional angiographic techniques. The advent of MRA has conferred a considerable diagnostic advance.

Disorders of neuronal migration had not been previously described with facial haemangioma and absence of internal carotid artery, as in this case, neither had hypoplastia of one cerebral hemisphere. Given the absence of other possible causes, it seems reasonable to link this hypoplasia to the lack of an ipsilateral internal carotid artery, in spite of apparently good blood supply through the large anterior communicating artery. Agenesis of the left carotid canal is consistent with the absence of left internal carotid artery.

In this patient, the cortical dysplasia is associated with hypoplasia of the cerebral hemisphere, on the same side as the absent internal carotid artery, with compression of the lateral ventricle by a heterotopic nodule. It

seems difficult to establish a relationship between these changes and the absence of the internal carotid artery, because all of them have been described in subjects without arterial malformations [9].

The abnormally thick cortex shows a deep fissure that does not represent schizencephaly, but rather the primitive Sylvian sulcus which appears in poorly developed cerebral hemispheres [10, 11], and has been correlated with normal but abundant venous drainage in cases of pachygyria. MRI also revealed multiple blood vessels within the heterotopic mass [9], probably representing the long pial vessels often seen running along the deep sulci of dysplasic cortex [10]. Heterotopic nodules can be seen as isolated abnormalities or associated with other disorders such as the Chiari II malformation, pachygyria or polymicrogyria [12, 13].

Alterations of neuronal migration have been found in several syndromes [14], most of them hereditary, originating in the first 20 weeks of gestation and without involvement of vascular structures. Disturbances of neuronal migration occurring in the second half of gestation have only recently been recognized [15, 16]. Diverse injuries, such as ischaemic and/or hypoxic events on the pial surface, can produce retraction of radial glial fibres. Neuroblasts which cannot complete migration end up as heterotopic neurons.

Several theories try to explain the possible aetiological mechanisms of migrational disorders in the corticosubcortical zone of the cerebral hemispheres. A malformation known as marginal glioneuronal heterotopia arises on the pial surface of the cerebral mantle. Damage to the pial membrane allows this barrier to be breached by migratory nerve cells and glia, which form a nodule over the cerebral cortex and occasionally lead to fusion of contiguous convolutions [17, 18]. Hypervascularity of the external cortical layers is usually present, and reciprocal invasion of the cerebral cortex by meningeal vessels may occur [18]. Another mechanism of cerebral cortical damage is the unique distribution of watershed infarcts of the fetal and neonatal cortex that do not occur in older children [16]. This radial pattern of distribution is imposed by immature blood vessels which penetrate the cortex from the meninges perpendicular to the brain surface [16]. Migrating neuroblasts in this zone tend to cuff these vessels in the presence of ischaemia, leaving columns of perivascular aggregates of neuroblasts [19, 20].

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