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Intracranial contrast-enhancing masses in infants with capillary haemangioma of the head and neck: intracranial capillary haemangioma?

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Abstract Contrast-enhancing intracranial masses are rarely found in infants with extracranial capillary haemangiomas (CH). We aimed to assess their nature and progression in three patients undergoing CT and/or MRI. The changes in size of both extra- and intracranial lesions were recorded. In a fourth case, a single examination was obtained. All patients harboured one or two enhancing intracranial nodular, meningeal-based lesions. Diffuse leptomeningeal enhancement of the cerebellar surface was also seen in one, which disappeared at follow-up. In all but one of the cases, the intracranial lesions were on the same side as the extracranial CH. These lesions and the extracranial CH demonstrated parallel changes in size (suggesting that both represent CH) during follow-up of 1–2 years: the size of intracranial lesions and the extracranial CH decreased in two cases, whereas it was unchanged in the third. One patient had a persistent trigeminal artery, while another had cerebellar atrophy with high signal in the cortex on T2-weighted images. In some cases, extracranial CH are part of PHACE syndrome; the association with intracranial CH might represent a peculiar phenotype of this rare vascular phakomatosis. As extracranial CH are known to regress spontaneously in the majority of cases, a conservative approach is recommended also for presumed intracranial CH; surgery should be avoided unless follow-up studies demonstrate growth.

Key words Haemangioma, capillary · Magnetic resonance imaging

Introduction

Capillary haemangioma (CH) is the most common tumour of infancy, occurring in up to 12% of Caucasian infants in the first year of life [1]. It is usually found in the head and neck and is more common in girls, with a 3:1 to 5:1 preponderance [2]. It typically involves the skin, scalp, and orbit, especially the lids, with frequent extension to the rectus muscles [3]. Microscopy shows small, tightly packed capillary-like vascular spaces bordered by benign, flattened endothelial cells and embedded in a delicate collagenous stroma [1, 4–6]. Less than one third of CH are present at birth [2], but most appear by the first month of life [6]. They typically demonstrate rapid growth up to 6–8 months, followed by a plateau phase between 8 and 12 months [2]. High blood flow and pressure within the mass lead to vascular dilatation and perivascular collagenisation [1], which accounts for their well-known tendency to spontaneous involution [1–4], beginning by 12 months and continuing until 5–10 years [2].

These lesions are distinctly uncommon in the intracranial compartment, only one histologically proven intracranial CH having been reported [1]. We describe four cases of extracranial CH with associated intracranial, meningeal-based contrast enhancing masses which we diagnosed as CH, based on their neuroradiological features and changes in size during follow-up.

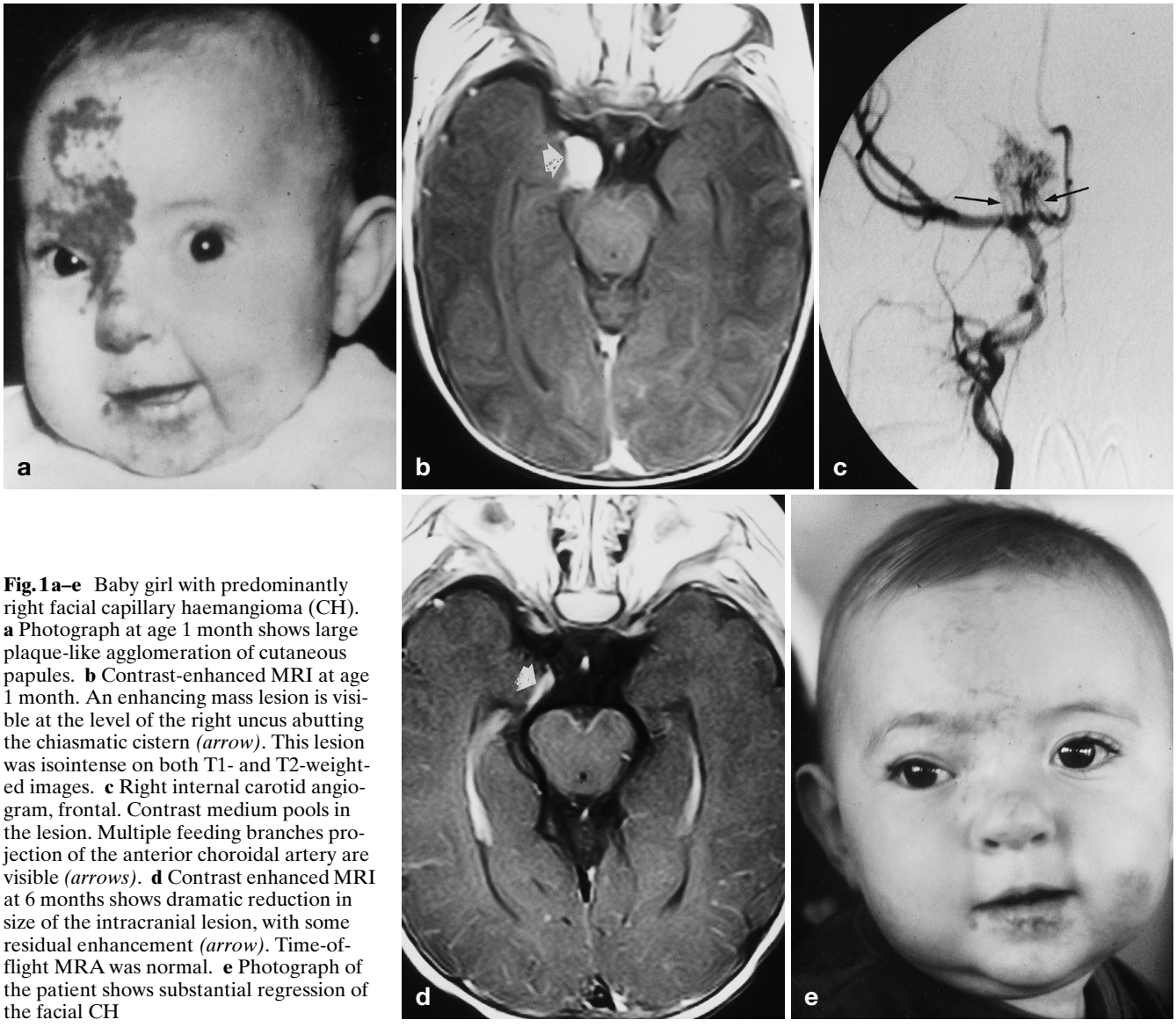

Fig. 1 a–e Baby girl with predominantly right facial capillary haemangioma (CH). **a** Photograph at age 1 month shows large plaque-like agglomeration of cutaneous papules. **b** Contrast-enhanced MRI at age 1 month. An enhancing mass lesion is visible at the level of the right uncus abutting the chiasmatic cistern *(arrow)*. This lesion was isointense on both T1- and T2-weighted images. **c** Right internal carotid angiogram, frontal. Contrast medium pools in the lesion. Multiple feeding branches projection of the anterior choroidal artery are visible *(arrows)*. **d** Contrast enhanced MRI at 6 months shows dramatic reduction in size of the intracranial lesion, with some residual enhancement *(arrow)*. Time-of-flight MRA was normal. **e** Photograph of the patient shows substantial regression of the facial CH

Methods

We studied four infants with CH of the head and neck who also had intracranial contrast-enhancing masses. All presented with large, plaque-like agglomerations of reddish cutaneous papules which became evident during the first month of life (Fig. 1). These lesions involved the soft tissues of the face and scalp without strict dermatomal distribution, although they were mainly on one side of the midline; contralateral extension was present in one case, and the orbits were involved in two. Neurological examination was normal and developmental history disclosed mild psychomotor delay in all cases. Family histories were consistently uneventful.

The intracranial lesions were discovered when CT or MRI was performed to assess the extracranial masses. Digital subtraction (DSA) and magnetic resonance angiography (MRA) were performed in one case at presentation. We obtained follow-up studies in three patients, which included CT, MRI and MRA. Embolisation was performed to treat a huge extracranial CH in case 3 after unsuccessful surgery. No attempt was made to treat the intracranial lesions.

Results

In all patients, CT and/or MRI disclosed nodular intracranial lesions displaying marked contrast enhancement (Figs. 1–4); one child had two separate masses (Fig. 3). There were three lesions in the cerebellopontine angle (Figs. 2–4), one in the unco-hippocampal region (Fig. 1), and one in the hypothalamus (Fig. 3). Three lesions were on the same side as the extracranial CH, one midline, and one contralateral. Diffuse lepto-

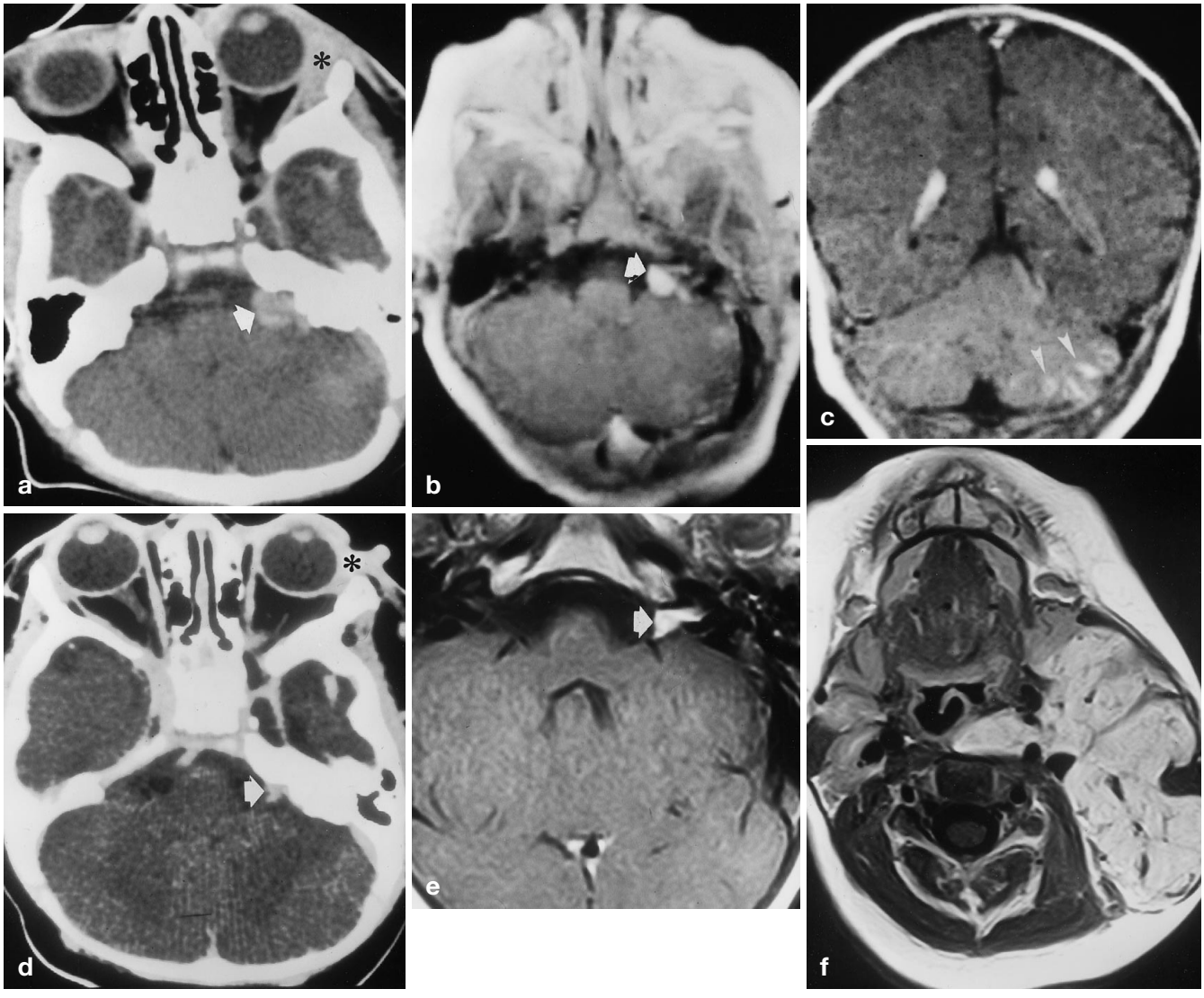

Fig. 2 a–f Baby girl with a left orbitofacial CH. **a** Contrast-enhanced CT at age 1.5 months shows enhancing left orbital portion of a large extracranial CH *(asterisk)* and a rounded enhancing mass in the left cerebellopontine angle (CPA) *(arrow)*. **b, c** Contrast-enhanced axial and coronal T1-weighted images. The enhancing mass in the left CPA is clearly visible *(arrow)*. Pial enhancement on the surface of the left cerebellar hemisphere is also seen *(arrowheads)*. **d** Contrast-enhanced CT at 4.5 months shows involution of the orbital CH *(asterisk)* and marked reduction in size of the left CPA lesion *(arrow)*. **e, f** Contrast enhanced MRI at age 15 months. An enhancing lesion is still visible in the left CPA, grossly unchanged since the previous CT. No enhancement along the pial surface of the cerebellum is visible. A huge mass has now developed in the left neck. Note the prominent flow voids within the mass, typical of CH

meningeal enhancement was noted on the surface of the left cerebellar hemisphere in one patient (Fig. 2). Only one patient underwent both MRA and DSA at presentation; both methods revealed abnormal intracranial vascularity, but DSA in far greater detail (Fig. 1).

Case 4 was lost at follow-up. Cases 1 and 2 received no therapy. Case 3 was given systemic steroid therapy (prednisone, 2 mg/kg/day, 2 cycles of 25 days in 6 months) after two unsuccessful attempts at surgical excision.

Follow-up ranged from 6 to 24 months. Marked or subtotal regression of the extracranial CH was seen within 6 months in two cases (Figs. 1, 2), whereas the mass was grossly unchanged in another (Fig. 3). The intracranial lesions behaved in parallel with the extracranial CH; marked or subtotal involution was seen simultaneously with the regression of the extracranial CH in two patients (Figs. 1, 2), while the intracranial and

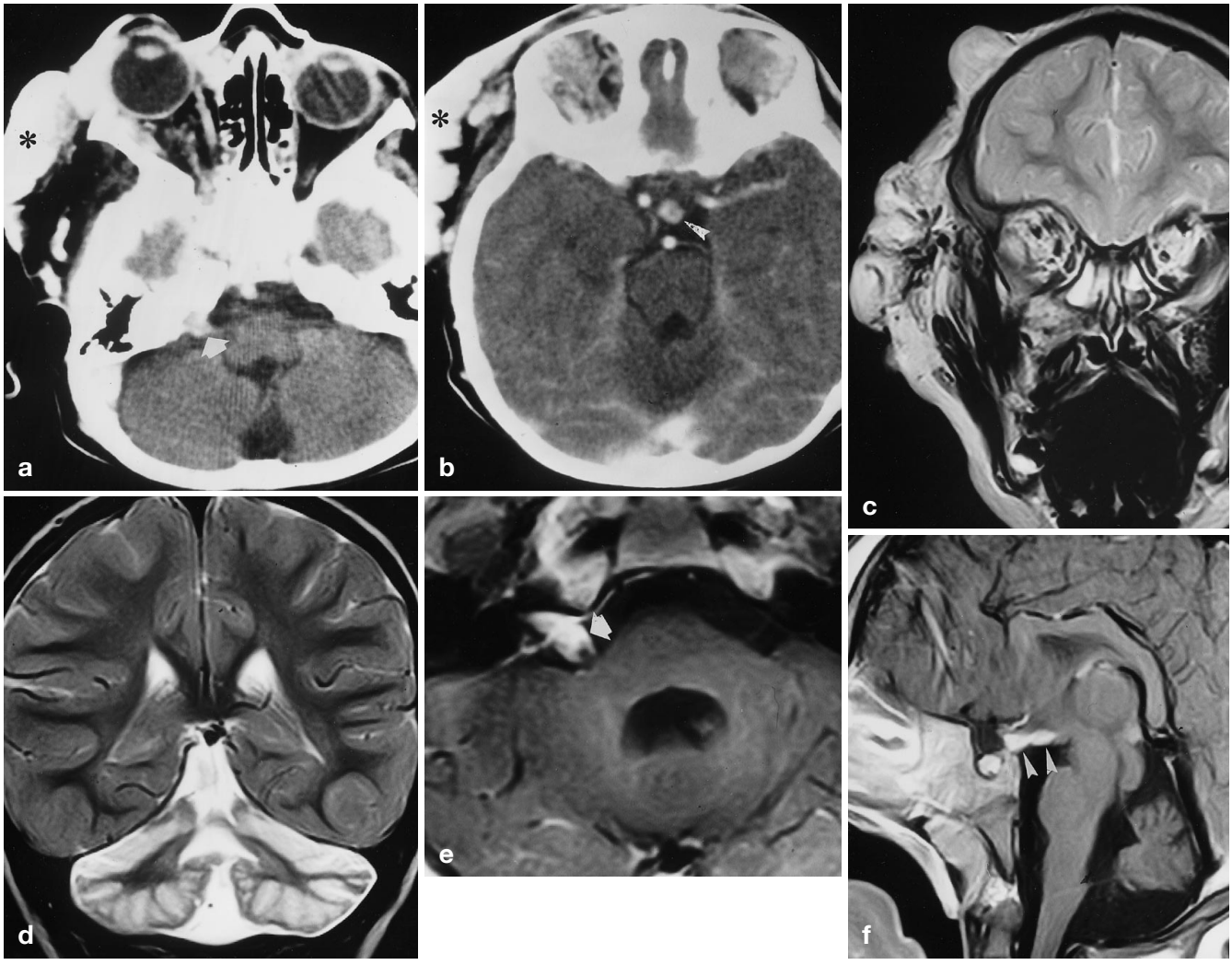

Fig. 3 a–f Baby girl with right CH of the face and scalp. **a, b** Contrast enhanced CT at age 1.5 years shows large, right orbitofacial CH *(asterisk)*. Abnormal enhancement is seen in the ipsilateral CPA *(arrow)* and in the hypothalamus *(arrowhead)* **c–f** MRI at age 2 years **c, d** T2-weighted **e, f** contrast-enhanced T1-weighted images. These show the extent of the extracranial CH, which is essentially unchanged. The cerebellar cortex appears atrophic and gives diffuse high signal. The intracranial lesions are grossly unchanged *(arrows)* although better seen than on CT

extracranial lesions did not change in the third (Fig. 3). The diffuse leptomeningeal enhancement seen initially on the surface of the cerebellum in case 2 disappeared altogether by 3 months (Fig. 2). In this case, a large CH appeared on the left side of the neck during follow-up (Fig. 2); it compressed the upper airway, causing respiratory distress, and was therefore excised. No increase in the intracranial lesions was seen at this time.

MRI at 2 years in patient 3 displayed cerebellar atrophy with high signal in the cortex on T2-weighted images; no contrast enhancement was seen. The patient developed cerebellar ataxia. As the size of the extracranial CH was unchanged after 7 months and two cycles of systemic steroid therapy, endovascular treatment was attempted. Contour particles were injected via the superficial temporal artery, the main feeding vessel. As the mass was also fed by a hypertrophied ophthalmic artery, large calibre particles were chosen to avoid embolisation of the intracranial circulation. The procedure was followed by partial involution of the mass with no side effects.

Discussion

Although extracranial CH may extend intracranially through skull base foramina [4], only one isolated, histologically proved intracranial CH has been reported [1]. It was an extra-axial, dural-based mass with strong contrast enhancement on both CT and MRI, in a child

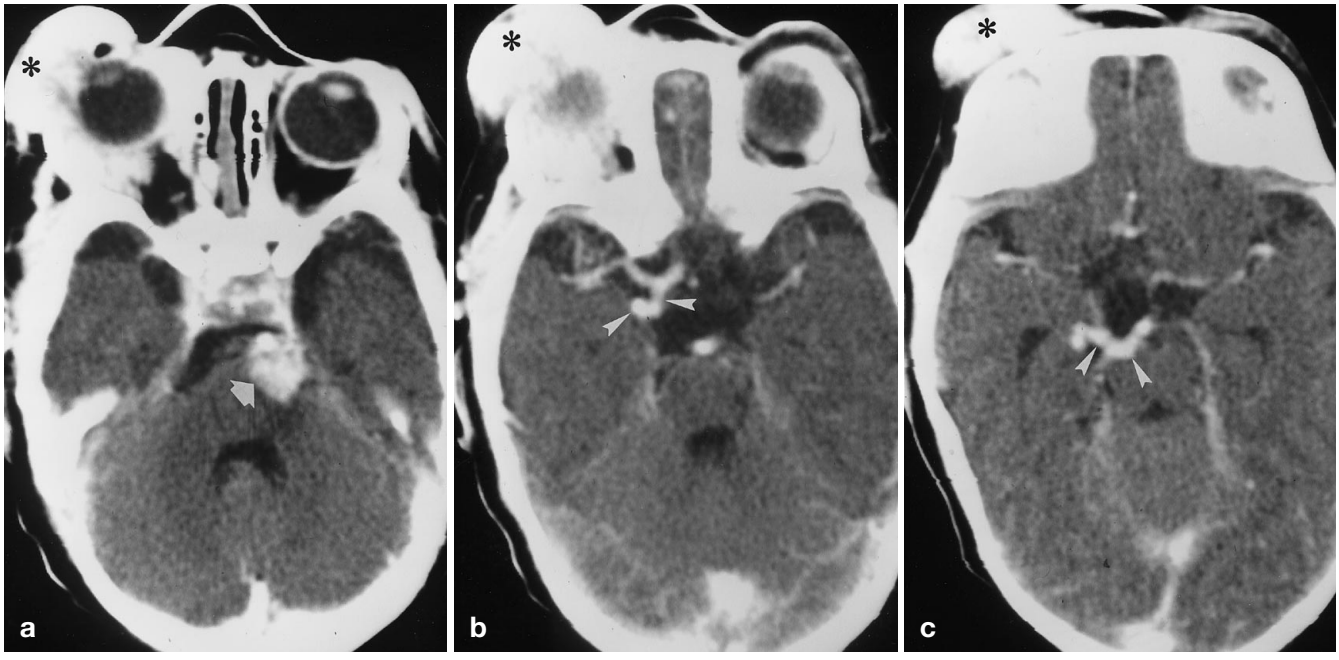

Fig. 4 a–c Newborn girl with right orbitofacial CH. **a–c** Contrast-enhanced CT. A large, right-sided cutaneous haemangioma is clearly visible *(asterisks)*. An enhancing mass lesion occupies the contralateral CPA *(arrow)*. A persistent trigeminal artery *(arrowheads)* is present

with no skin abnormality. We are aware of no prior reports of the association of head and neck and histologically proven intracranial CH; however, Pascual-Castroviejo [7] reported a facial CH associated with "angiomatous malformations" at the left carotid siphon and hypothalamic area. Frieden et al. [8] reported right facial CH associated with a "2 × 2 cm focus of abnormal blood vessels along the anterior aspect of the right cerebellar pontine angle", but the pertinent MR images were not shown. Bar-Sever et al. [9] reported the coexistence of a multicentric hepatic haemangioendothelioma with a large mass in the brain of a preterm infant; although histology was not obtained, they suspected an intracranial CH on CT and scintigraphy and on regression with interferon therapy.

On MRI, extracranial CH are well-circumscribed lesions, isointense on T1- and giving high signal on T2-weighted images, reflecting their content of unclotted blood [1, 3]. Inhomogenous signal within the mass may represent areas of clot due to previous haemorrhage or signal void from enlarged vascular spaces [3]. Typically, CH enhance markedly with contrast medium on both CT and MRI [1]. All lesions in our series, both extracranial and intracranial, enhanced markedly and homogeneously on CT and MRI. The angiographic features of CH are quite characteristic [5]. The tumour is fed by enlarged branches of normal arteries; usually, several vessels fan out in a radial pattern, supplying individual lobules of the lesion. Intense contrast-medium staining is the expression of pooling of contrast medium in the vascular spaces within the mass; early draining veins are often present. Such features are typical of extracranial CH, but were also seen on DSA in one intracranial lesion in our series (Fig. 1).

Although the anatomical distribution of CH includes the intracranial compartment [1], the imaging features of intracranial CH are nonspecific. We were therefore unable to exclude other extra-axial neoplasms on imaging findings. However, we hypothesised that these masses were intracranial CH on account of: 1. the presence of an extracranial CH; 2. the parallel changes in size of the extra- and intracranial lesions; and 3. the suggestive angiographic features in one case.

One of these infants (case 2) also showed marked, diffuse leptomeningeal contrast enhancement on the surface of the left cerebellar hemisphere (Fig. 2), which disappeared in parallel with the regression of the other vascular lesions. This could represent pial vascular dysplasia or a diffuse variety of CH. Other common causes of diffuse leptomeningeal enhancement, such as inflammatory meningitis [10], seemed unlikely, due to the spontaneous regression and the lack of a consonant clinical picture. The Sturge-Weber syndrome was unlikely, as the abnormal contrast enhancement was in the posterior cranial fossa, which is atypical [11, 12], and because of the absence of other features, including a port-wine stain. In case 3, cerebellar atrophy with high signal in the cortex was detected in a child who developed cerebellar ataxia (Fig. 3). The cause of this was not

Table 1 Case material: clinical and neuroradiological findings (*CPA* cerebellopontine angle, *CH* capillary haemangioma)

Case	Sex	Age	Neurological signs	Imaging studies at presentation	Location of extracranial CH	Location of intracranial lesions	Chosen treatment	Follow-up imaging studies	Follow-up results	Adjunct treatment
1	F	1 month	None	MRI, MRA, DSA	Prevailingly right side, face and lips	Right uncohippocampal	Watchful waiting	MRI, MRA	Dramatic involution of both extracranial CH and intracranial lesion	None
2	F	1.5 months	None	CT, MRI	Left side, face and orbit	Left CPA; Leptomeningeal enhancement at cerebellar surface	Watchful waiting	CT, MRI	Marked involution of both extracranial CH and intracranial lesion; leptomeningeal enhancement disappeared; large CH appeared in left cervical region	Surgical excision of cervical CH
3	F	1.5 years	None at presentation developed ataxia	CT	Right side, face and scalp	Right CPA; Hypothalamus	Systemic steroid therapy	MRI, MRA, DSA	Substantially unchanged lesions; cerebellar atrophy with hyperintense cortex	Embolisation of extracranial CH
4	F	1 month	None	CT	Right side, face and orbit	Left CPA; persistent trigeminal artery	Systemic steroid therapy	Lost at follow-up	Lost at follow-up	None

clear; cerebellar atrophy is often of unknown aetiology [13].

One infant (case 2) developed a bulky mass in the neck after the regression of her facial CH and intracranial lesions. The observation that some tumours, cutaneous and intracranial, may regress while new ones appear raises the suspicion that, at least in some cases, CH may represent part of a phakomatosis. This term is used to describe congenital conditions with cutaneous, central nervous system and often ocular involvement [11, 12], whose inheritance patterns are variable. Recently, the terms PHACE syndrome [8] and cutaneous haemangioma-vascular complex (CHVC) syndrome [14] have been coined to describe the non-random association of extracranial capillary haemangiomas, posterior cranial fossa malformations including the Dandy-Walker malformation and cerebellar hemisphere hypoplasia, arterial anomalies such as persistent trigeminal artery or hypo- or aplasia of the internal and external carotid and vertebral arteries, coarctation of the aorta and eye abnormalities. Cutaneous CH in this syndrome are generally large, plaque-like and unilateral, as in our cases. They do not strictly adhere to a dermatomal distribution [8, 15]. Most affected patients are girls, and most cases reported so far sporadic [8, 15]. Incomplete phenotypic expression has been described [16], and it appears that the complete spectrum of intracranial abnormalities which may be found with an extracranial CH has not yet been completely assessed. We speculate that the association of extracranial and presumed intracranial CH which grow and regress spontaneously may represent a particular phenotype of this syndrome; this consideration is supported by the added association of a persistent trigeminal artery in case 4. CH typically display rapid neonatal growth due to endothelial proliferation, followed by diminishing hyperplasia and progressive fibrosis [17]. We speculate that, at least in some cases, this spontaneous regression, rather than representing a simple effect of the haemodynamics within the lesion, could result from self-limited growth, a phenomenon seen in phakomatoses such as neurofibromatosis type I, in which spontaneous regression of intracranial "gliomas" has been reported [18].

We suggest that all patients with CH of the head and neck should undergo screening contrast-enhanced imaging to detect silent intracranial lesions, and that, as extracranial CH are known to regress spontaneously or with systemic steroids or interferon [19–21], surgery should be avoided unless follow-up studies demonstrate growth or, more importantly, the clinical picture deteriorates.

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