

Guillaume Gorincour · Victor Kokta · Françoise Rypens
Laurent Garel · Julie Powell · Josée Dubois

Imaging characteristics of two subtypes of congenital hemangiomas: rapidly involuting congenital hemangiomas and non-involuting congenital hemangiomas

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Abstract Background: Common infantile hemangiomas (COMMON) occur in approximately 10% of infants by the age of 1 year, with a female predominance. Some hemangiomas can be fully developed at birth and are thus called congenital hemangiomas (CH). Within this population, two courses have been identified: rapidly involuting CH (RICH) and non-involuting CH (NICH). Little has been reported on the clinical prognosis and imaging features of these entities. **Objective:** To describe the imaging characteristics of two subtypes of CH, i.e. RICH and NICH, and to compare them with COMMON. **Materials and methods:** We retrospectively gathered data on 26 children presenting with CH, i.e. lesions fully developed at birth. These lesions were divided into two groups according to the clinical course: suspected RICH ($n=8$) and suspected NICH ($n=18$). We used US, CT or MRI and angiography to identify the gross anatomy and structure and the vascularization. Imaging findings were compared with the clinical course and pathology results, when available. The imaging findings in these patients were compared retrospectively with those in 26 patients with COMMON randomly chosen from the database of our multidisciplinary clinic. **Results:** When compared with COMMON imaging characteristics, NICH and RICH had distinctive features on US such as being heterogeneous (72% of NICH and 62.5% of RICH vs 42.3% of COMMON), visible vessels (72% of NICH and 62.5% of RICH vs 15.4% of COMMON), calcifications (17% of

NICH and 37.5% of RICH vs no case of COMMON). On CT and/or MRI, we compared imaging features such as well-defined limits (67% of NICH and 60% of RICH vs 100% of COMMON), and fat stranding (29.4% of NICH and RICH vs 7.7% of COMMON). **Conclusion:** Distinctive imaging characteristics are observed in cases of CH with US findings of visible vessels and calcifications statistically significant.

Keywords Infantile hemangioma · Congenital hemangioma · Rapidly involuting congenital hemangioma · Non-involuting congenital hemangioma · Ultrasound · CT · MR

Introduction

Common infantile hemangiomas (COMMON) occur in approximately 10% of infants by the age of 1 year, with a female predominance. Their evolutive pattern comprises an actively proliferating, highly angiogenic phase followed by spontaneous regression of angiogenesis, a decrease in size of the lesion, and, finally, the complete disappearance of the soft-tissue mass [1, 2]. Some hemangiomas can be fully developed at birth and are thus called congenital hemangiomas (CH), because they do not follow the typical evolutive pattern of COMMON and do not grow after birth. The incidence remains unknown. Within the CH population, two courses have been identified through the extensive work of vascular anomalies teams in Boston and Paris [1–3]: rapidly involuting congenital hemangiomas (RICH) and non-involuting congenital hemangiomas (NICH). RICH are defined by a spontaneous and complete resolution before the age of 14 months, and NICH by a proportional growth with the child and no regression whatsoever. Few reports are available concerning the clinical prognosis and imaging features of these entities.

We describe the clinical, radiological and pathological findings in 26 patients with CH and we also aimed to determine which features distinguish NICH and RICH from each other, as well as from COMMON.

G. Gorincour · F. Rypens · L. Garel · J. Dubois (✉)
Department of Medical Imaging, Sainte Justine Hospital,
3175 Cote Sainte Catherine, Montreal,
QC, H3T 1C5, Canada
E-mail: josee-dubois@ssss.gouv.qc.ca
Tel.: +1-514-3454637
Fax: +1-514-3454816

V. Kokta
Department of Pathology, Sainte Justine Hospital,
Montreal, QC, Canada

J. Powell
Department of Dermatology, Sainte Justine Hospital,
Montreal, QC, Canada

Materials and methods

We retrospectively reviewed the children presenting at our vascular malformation clinic with a congenital hemangioma, i.e. a lesion fully grown at birth without a subsequent proliferative phase. We identified 26 such patients who were examined by the attending dermatologist and referred to the Medical Imaging Department for US examination, CT and/or MRI, and (in some cases) angiography. The clinical workload at our multidisciplinary vascular malformation clinic is approximately 300 patients per year consisting of approximately 30% of COMMON, and 1% NICH and RICH.

They were classified as suspected RICH ($n=8$) and suspected NICH ($n=18$) according to the clinical course, i.e. spontaneous resolution within 14 months after birth (RICH) or proportional growth with the child and no regression (NICH).

US examinations ($n=26$) were performed using a constant technique by experienced pediatric radiologists (J.D., F.R., L.G.). A 5000 HDI scanner (ATL, Seattle, Wash.) with a linear 8- to 12-MHz transducer was used. Color Doppler sonograms were obtained by low-pulse frequency and a wall filter. Pulse-repetition frequency was increased only if aliasing occurred. Gray-scale US outlined the size, the contours, and the echogenicity of the

lesion and looked for the presence of visible vessels within or around the lesion, as well as the presence of calcification(s). Color Doppler US estimated vessel density, counting the number of color Doppler signals in a 1-cm² area of the most highly vascularized part of the lesion. Pulsed Doppler US was also performed, with maximal systolic Doppler shift (increased 2 kHz) being measured in kilohertz; the resistance index was also established. Shunts were considered present when arterial diastolic speed was elevated and arteriolization within veins was identified. The volume of the lesions was calculated sonographically using the three-dimensional method available in the software.

CT scans were performed with a single detector Picker PQ 5000 CT scanner (Picker International, Highland Heights, Ohio). Slice thickness (3–5 mm depending on the location), pitch, kV, mAs and fields of view depended on the body region in question. All CT scans ($n=7$) were performed without and then with contrast medium enhancement. The CT scan images were examined for density before contrast medium enhancement, the presence of calcification(s), the limits of the lesion, the fat stranding and the type of enhancement.

MRI was performed using a 1.5-T Magnetom Symphony (Siemens, Erlangen, Germany) with T1-, T2- and Fat-Sat-weighted sequences before and after gadolinium

Table 1 Clinical and evolutive features, suspected subgroup and pathological diagnosis, if any, in our 26 patients with CH (RICH rapidly involuting congenital hemangioma, NICH non-involuting congenital hemangioma)

Patient no.	Age	Sex	Location	Volume (cm ³)	Cutaneous appearance	Evolution	Suspected subgroup	Pathology
1	10 years	Female	Right knee	27	Blue	Regression at age 9 months	RICH	0
2	10 years	Female	Head	76	Blue	Regression at age 1 year	RICH	RICH
3	6 years	Male	Left thigh	480	Red–blue	Regression at age 1 year	RICH	0
4	6 years	Female	Trunk	32	Telangiectatic halo	Stability	NICH	0
5	6 years	Male	Left cheek	30	Red–blue	Stability	NICH	NICH
6	16 years	Female	Left leg	22	Blue with veins	Stability	NICH	NICH
7	11 years	Male	Right thigh	5	Blue	Stability	NICH	NICH
8	2 years	Female	Left chest	10	Blue	Stability	NICH	NICH
9	17 years	Female	Thorax	10	Telangiectasias	Stability	NICH	NICH
10	8 years	Female	Hip	55	White–blue halo	Stability	NICH	0
11	9 years	Female	Head	45	Blue–red	Regression at age 7 months	RICH	0
12	4 years	Male	Shoulder	11	Red with a blue halo	Stability	NICH	NICH
13	14 years	Female	Buttock	14	Red ulcerated	Stability, ulceration	NICH	NICH
14	3 years	Male	Right knee	40	Red ulcerated	Regression at age 7 months	RICH	0
15	11 years	Female	Wrist	16	Red	Stability	NICH	NICH
16	7 months	Female	Right thigh	63	Red	Regression at age 2 months	RICH	RICH
17	13 years	Male	Right arm	12	Red	Stability	NICH	0
18	7 years	Female	Right elbow	15	Red	Stability	NICH	0
19	1 year	Female	Right arm	15	Blue with white halo	Regression at age 1 year	RICH	0
20	12 years	Male	Right forearm	40	Blue vein, telangiectatic halo	Stability	NICH	0
21	11 years	Female	Lip	25	Blue–red	Stability	NICH	0
22	10 months	Male	Head	25	Blue with white halo	Regression at age 4 months	RICH	0
23	8 years	Male	Lip	10	Blue–red	Stability	NICH	0
24	10 years	Male	Right arm	123	White with telangiectasias	Stability	NICH	0
25	18 years	Male	Right elbow	175	Red	Stability	NICH	NICH
26	8 years	Female	Nose	20	White with telangiectasias	Stability	NICH	0

Table 2 Imaging findings in the suspected RICH subgroup (eight patients) (*Vessels* visible tubular vascular structures on color Doppler, compared with subcutaneous fat)

Patient no.	US						CT		
	Echostructure	Vessels	Calcification	Number of vessels (cm ⁻³)	Systolic frequency (kHz)	Shunt	Density before contrast	Calcification	Well-defined limits
1	Isoechogenic	No	No	3-4	>2	No	Isodense	No	No
2	Heterogeneous	Yes	Yes	3-4	>2	Yes	Isodense	Yes	Yes
3	Heterogeneous	Yes	No	>5	>2	No	Heterogeneous	No	No
11	Heterogeneous	Yes	Yes	2-3	>2.2	No	Homogeneous	Yes	Yes
14	Isoechogenic	No	No	>5	>2	No	-	-	-
16	Isoechogenic	No	Yes	>5	>2	No	-	-	-
19	Heterogeneous	Yes	No	>5	>2	No	-	-	-
22	Heterogeneous	Yes	No	>5	>2	No	-	-	-

enhancement. Slices of 3–5 mm were used, depending on the location. MRI examinations ($n=12$) studied signal intensities on T1- and T2-weighted sequences before contrast medium enhancement, as well as the presence of calcification(s), flow void phenomenon, the limits of the lesion, the presence of edema, fat stranding and the type of enhancement.

Supraselective angiography was performed in five patients with suspected NICH using a Toshiba KXO-80A (Toshiba, Tokyo, Japan) and looked for evidence of abnormalities of vessel size, shunts, dilated (ectatic) and/or stagnating veins, aneurysm and capillary blush. The indication for angiography was initially therapeutic in all cases (and to a lesser extent for differential diagnosis with more aggressive tumors such as fibrosarcoma). Sonographic shunts that were not identified on the angiograms were considered microshunts.

Pathological examinations ($n=11$) were reviewed by a single dermatopathologist who followed the criteria

defined by Enjolras, Mulliken, North and colleagues [4–6] but who was blinded to the clinical, evolutive (non-involuting), and radiological findings, i.e. to the suspected subgroup of the lesions. Involuting lesions were biopsied incisionally because of clinical atypical findings. Non-involuting lesions were fully operated upon, mainly because of esthetic or functional disturbances. A lesion was classified as a NICH when composed of large lobules of small vessels with arteriovenous or arteriolymphatic microfistulae, and/or hobnailed endothelial cells. Some lesions had few lobules. Conversely, a lesion was classified as a RICH when composed of variably sized vascular lobules around a large central draining vein and adjacent fibrosis, without any microfistulae or hobnailed endothelium. Regressive endothelial changes such as dystrophic calcifications and fibrosis were prominent. Negative GLUT1 immunostaining was completed on all cases to exclude infantile hemangioma. Given the regressive nature of RICH, most were not

Table 3 Imaging findings in the suspected NICH subgroup (18 patients) (*Vessels* visible tubular vascular structures on color Doppler, *isosignal* compared with subcutaneous fat)

Patient no.	US						CT			
	Echostructure	Vessels	Calcification	Number of vessels (cm ⁻³)	Systolic frequency (kHz)	Shunt	Density before contrast	Calcification	Well-defined limits	Enhancement with contrast
4	Heterogeneous	Yes	No	>5	>2	Yes	-	-	-	-
5	Heterogeneous	Yes	No	>5	1-2	No	-	-	-	-
6	Heterogeneous	Yes	No	1	1	No	-	-	-	-
7	Heterogeneous	Yes	Yes	1-2	>2	No	-	-	-	-
8	Isoechogenic	No	No	3-4	>2	No	-	-	-	-
9	Heterogeneous	Yes	Yes	>5	>8	Yes	-	-	-	-
10	Isoechogenic	No	No	>5	>3	No	-	-	-	-
12	Heterogeneous	Yes	No	3-4	>2	No	-	-	-	-
13	Heterogeneous	Yes	No	>5	>2	Yes	-	-	-	-
15	Isoechogenic	No	No	>5	>2	No	Isodense	No	Yes	Homogeneous
17	Heterogeneous	Yes	No	2-3	1	No	-	-	-	-
18	Heterogeneous	Yes	No	3-4	>2	Yes	-	-	-	-
20	Heterogeneous	Yes	No	>5	>2	Yes	Isodense	No	No	Homogeneous
21	Heterogeneous	Yes	No	>5	>2	No	-	-	-	-
23	Heterogeneous	Yes	No	>5	>2	No	-	-	-	-
24	Heterogeneous	Yes	Yes	2	1	No	Heterogeneous	Yes	Yes	Heterogeneous
25	Isoechogenic	No	No	>5	>2	No	-	-	-	-
26	Isoechogenic	No	No	>5	>2	No	-	-	-	-

T1(2) spontaneous signal on T1(2)-weighted images, *Gadolinium* pattern of enhancement after gadolinium; *isoechogenic, isodense, isosignal*

Enhancement with contrast	Fat stranding	MRI							
		T1	T2	Edema	Gadolinium	Flow void phenomenon	Well-defined limits	Fat stranding	Prominent veins
Homogeneous	Yes	–	–	–	–	–	–	–	–
Homogeneous	No	Isosignal	Hypersignal	No	Homogeneous	Yes	Yes	No	No
Heterogeneous	Yes	–	–	–	–	–	–	–	–
Homogeneous	No	–	–	–	–	–	–	–	–
–	–	–	–	–	–	–	–	–	–
–	–	–	–	–	–	–	–	–	–
–	–	–	–	–	–	–	–	–	–
–	–	Isosignal	Heterogeneous	No	Homogeneous	No	Yes	No	No

biopsied and therefore not available for pathological examination.

Then, the imaging features in our NICH and RICH patients were retrospectively compared with those (US in all cases, plus CT and/or MRI) in 26 other patients with COMMON chosen retrospectively and randomly from our multidisciplinary clinic database within 2 years.

Results

Clinical and evolutive data as well as suspected subgroup and final pathological diagnosis, if any, are summarized in Table 1.

The sex ratio was 15 females to 11 males, confirming the absence of female preponderance, unlike COMMON. The predilection for some locations was also confirmed with the head or the limbs near a joint involved in 23 patients (88%). Clinically, RICH were rather blue-col-

ored, and telangiectasias were only seen with NICH. The mean regression time of RICH was 8.1 months (minimum 2 months, maximum 12 months), and regression was complete within 1 year in all patients. The volume of the lesions was highly variable, ranging from 5 to 480 cm³, with 20 lesions below 50 cm³.

We retrospectively reviewed our imaging data focusing on those data that could lead us to confuse or differentiate COMMON, NICH and RICH. The data are summarized in Table 2 (RICH group) and Table 3 (NICH group).

On US, 62.5% of RICH lesions (five out of eight) exhibited both heterogeneity and visible vessels, and in 37.5% (three) a calcification was noted. In all lesions but one (87.5%), no shunt was demonstrated. For NICH, 72% of lesions (13 out of 18) exhibited heterogeneity as well, because of the visible vessels, and in approximately 17% of lesions (three) a calcification was noted. A shunt was demonstrated on US in more than a quarter of lesions (about 27%, i.e. five lesions).

T1(2) spontaneous signal on T1(2)-weighted images, *Gadolinium* pattern of enhancement after gadolinium; *isoechogenic, isodense,*

Fat stranding	MRI								Angiography
	T1	T2	Edema	Gadolinium	Flow void phenomenon	Well-defined limits	Fat stranding	Prominent veins	Description
–	Isosignal	Hypersignal	No	Homogeneous	No	No	No	–	–
–	Isosignal	Heterogeneous	No	Heterogeneous	No	No	Yes	–	–
–	Isosignal	Hypersignal	No	Homogeneous	No	Yes	No	–	–
–	Isosignal	Hypersignal	No	Homogeneous	No	Yes	No	–	–
–	–	–	–	–	–	–	–	No	Capillary blush
–	Heterogeneous	Heterogeneous	No	Heterogeneous	Yes	Yes	No	–	–
–	–	–	–	–	–	–	–	–	–
–	–	–	–	–	–	–	–	–	–
–	–	–	–	–	–	–	–	Yes	Capillary blush
No	–	–	–	–	–	–	–	Yes	Capillary blush
–	–	–	–	–	–	–	–	–	venous ectasia
–	–	–	–	–	–	–	–	–	venous ectasia
Yes	Isosignal	Hypersignal	No	Homogeneous	No	Yes	No	Yes	Venous ectasia
–	–	–	–	–	–	–	–	–	–
–	–	–	–	–	–	–	–	Yes	Capillary blush
–	Heterogeneous	Heterogeneous	No	Heterogeneous	No	No	Yes	–	–
No	Isosignal	Heterogeneous	No	Heterogeneous	No	Yes	No	–	–
–	Isosignal	Hypersignal	No	Homogeneous	No	Yes	No	–	–
–	Isosignal	Hypersignal	No	Homogeneous	No	Yes	No	–	–

The following group comparisons showed statistically significant differences in proportions: visible vessels COMMON versus NICH $P=0.010$, COMMON versus RICH $P=0.011$; and calcification COMMON versus RICH $P=0.004$. The statistical test used was a Fisher's Exact Chi-squared permutation test. Statistically significant differences were considered achieved for P values ≤ 0.05 , two-tailed.

On angiography, prominent venous vessels were noted in 80% of lesions (four out of five), as well as capillary blush. No macroscopic shunting was ever noted. In 11 lesions, all GLUT1 immunostaining examinations were negative. Nine lesions met the pathological criteria for NICH and two for RICH. Clinical/radiological suspicion and pathological diagnosis were concordant in all lesions for which pathology was available. In the absence of biopsy, RICH diagnosis is based on clinical grounds.

When compared with COMMON imaging characteristics (Table 4), NICH and RICH exhibited:

- Comparable features suggestive of hemangiomas, i.e. high vessel density and peak systolic shift on US, homogeneous enhancement and absence of peripheral edema on CT and/or MRI.
- Distinctive features that can help differentiate between those entities, i.e. heterogeneous sonographic structure, sonographically visible vessels, calcifications (specific for NICH and RICH), less well-defined limits on CT and/or MRI, and fat stranding.

We illustrate the clinical, imaging and pathological findings through two patients with RICH (patient 22, Fig. 1, and patient 2, Fig. 2) and two patients with NICH (patient 4, Fig. 3, and patient 15, Fig. 4).

Discussion

Benign vascular soft-tissue anomalies in infancy can be sonographically distinguished and include vascular tumors (hemangioendothelioma, tufted angioma, infantile myofibromatosis, etc.), hemangiomas (COMMON and CH, solid-tissue lesions) and vascular malformations present at birth (abnormal veins, arteries and veins, or

cysts on US). COMMON occur in about 10% of Caucasian infants by the age of 1 year [1] with a marked female predominance (sex ratio 3–5:1). They typically grow rapidly during the first year of life (proliferating phase), involute slowly between the ages 1 and 7 years (involuting phase) and then completely regress by 8–12 years of age [2]. Most begin as relatively inconspicuous blanched or blushed macules, in some cases evident at birth, and then dramatically expand like tumor-type masses.

The notion of congenital hemangioma was introduced in 1996 by Boon et al. [3] as a hemangioma-like lesion fully grown at birth, which seems to be at or past its proliferative peak. Among this group, two subsets were subsequently identified: NICH, which undergo proportional growth with the child but no regression [4, 5], and RICH, which regress completely within 14 months of birth [3]. It can be clinically difficult to distinguish a COMMON from a RICH when the COMMON is present and prominent at birth and undergoes a quick but moderate proliferative phase, that is, the dichotomic classification between COMMON and congenital hemangiomas is not always obvious.

In addition, NICH and RICH have overlapping clinical features [6]: they have an almost equal sex distribution, are usually solitary, and have a similar average diameter and a predilection for the same cutaneous locations (head or limbs near a joint), as confirmed in our series. Color is violaceous with multiple tiny or coarse telangiectasias, often a surrounding pale halo, sometimes a central ulceration, linear scar or central nodularity.

In previous studies, we have established vessel density estimation on Doppler US by counting the number of color Doppler signals per square centimeter in the most vascularized area of the lesion, using constant and strictly controlled technical factors [7–9]. Pulsed Doppler US served both to distinguish color Doppler artifacts from the blood-flow signal and to establish the maximal systolic Doppler shift (> 2 kHz). We chose to express this Doppler shift in kilohertz rather than in centimeters per second as a velocity measurement because it is often difficult to determine the angle between the Doppler beam and small, tortuous vessels in a highly vascularized tumor. We found that COMMON were present on US with a high vessel density estimation and a Doppler shift

Table 4 Comparative imaging findings among NICH, RICH and CIH

	COMMON	NICH	RICH
US	26 patients	18 patients	8 patients
Heterogeneous echostructure	42.3%	72%	62.5%
Visible vessels	15.4%	72% ($P=0.010$)	62.5% ($P=0.011$)
Calcification(s)	0	17%	37.5% ($P=0.004$)
Number of vessels/cm ² > 3	100%	77.8%	87.5%
Systolic shift > 2	100%	77.8%	100%
Shunts	27%	27.8%	12.5%
CT and/or MRI	26 patients	12 patients	5 patients
Well-defined limits	100%	66.6% (8)	60% (3)
Peripheral edema on T2W	0	0	0
Fat stranding	7.7%	29.4% (5)	
Homogeneous enhancement	85%	66.6% (8)	80% (4)
Flow void on MRI	34.6%	10% (1 out of 10)	50% (1 out of 2)
Angiography	–	5 patients	–
Venous ectasia(s)	–	80% (4)	–

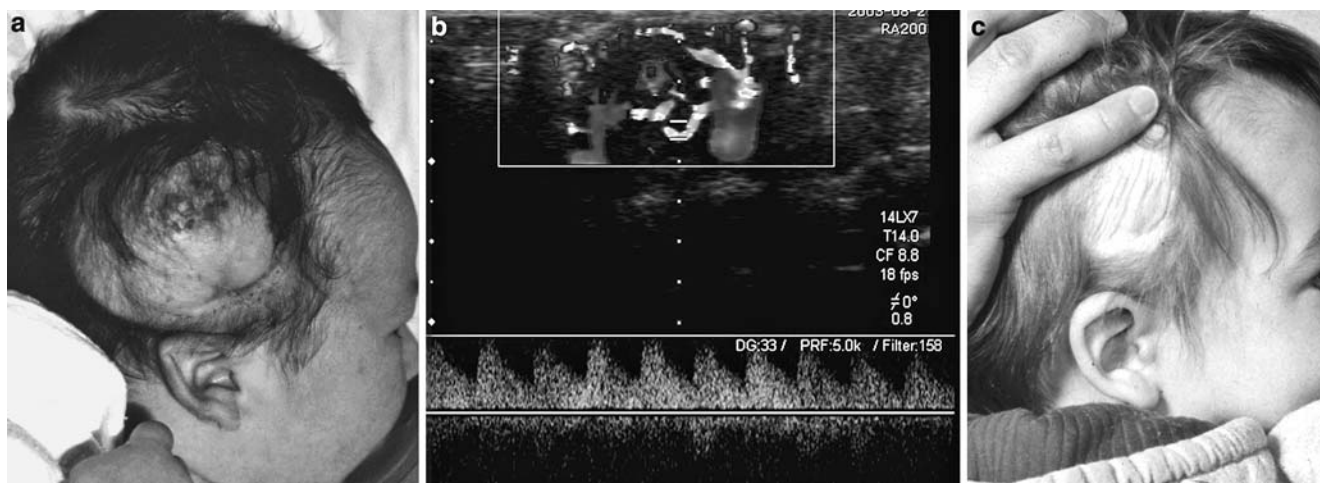
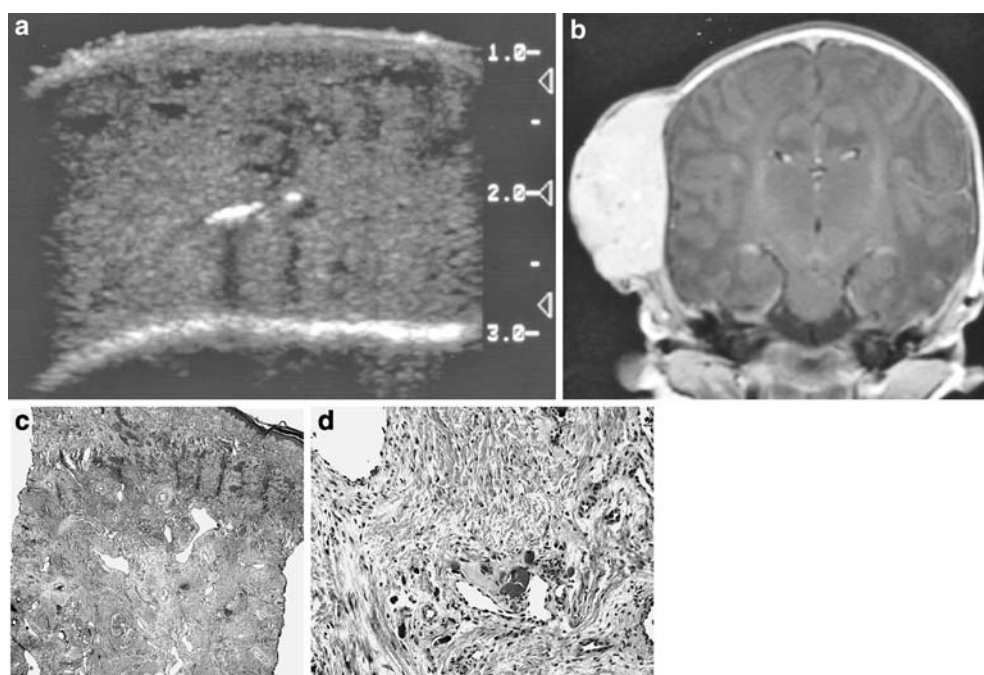


Fig. 1 Clinical and imaging findings in RICH (patient 22). **a** Photograph shortly after birth shows the large lesion localized on the right side of the head. The skin is a rather blue color without telangiectasias. **b** Transverse US Doppler scan of the lesion exhibits

the classical features of hemangiomas with high vessel density and peak systolic shift. **c** Photograph at the age of 1 year illustrates the nearly complete regression with a slight excess of skin

Fig. 2 Imaging and pathological findings in RICH (patient 2). **a** Transverse US image of the lesion at that time demonstrates a heterogeneous structure with two calcifications (posterior acoustic shadowing). **b** Coronal T1-weighted MR image after gadolinium reveals an intense and homogeneous enhancement with well-defined limits. **c** Low-power view (2.5× hps) demonstrates large central drainage veins with adjacent fibrosis and surrounding vascular lobules. **d** Medium-power (20× hps) shows regressed capillary lobules and associated dystrophic calcifications



exceeding 2 kHz. When these two criteria are met, the diagnosis of COMMON is made reliably.

In the literature, NICH and RICH have been depicted as exhibiting fast flow on US and MRI [1–3, 6, 10], but NICH remain fast-flow lesions, similar to COMMON in the proliferative phase. NICH have also been called small arteriovenous malformation (AVM) or arteriolocapillary malformation because US shows microshunts.

In this study, we found that all our NICH and RICH lesions met the US diagnostic criteria that we defined for COMMON in terms of high vessel density estimation and Doppler. Moreover, in our 13-year multidisciplinary experience of COMMON, we found none presenting with calcification on US, whereas a calcification was seen in 37.5% of our eight RICH and almost 17% of our 18 NICH. This is not to say that calcification will never be

found in COMMON, but its presence suggests CH. We also found that very frequently, i.e. in 72% of NICH and 62.5% of RICH, tubular vascular structures—both arteries and veins but mainly veins—were visible on gray-scale US, in contrast to the findings in COMMON. This visibility is a component of the atypical US heterogeneity of NICH and RICH that was found in 69% of all our patients.

On CT and/or MRI, NICH and RICH have thus far been described as very similar to COMMON, with some RICH having areas of inhomogeneity and larger flow voids [1, 2, 4–6]. A case of congenital non-progressive hemangioma was very atypical because of hemosiderin deposits [5]. In our series, we found significant phenomena of fat stranding in 29.4% of lesions examined by CT and/or MRI (5 out of 17), and none of our lesions fea-

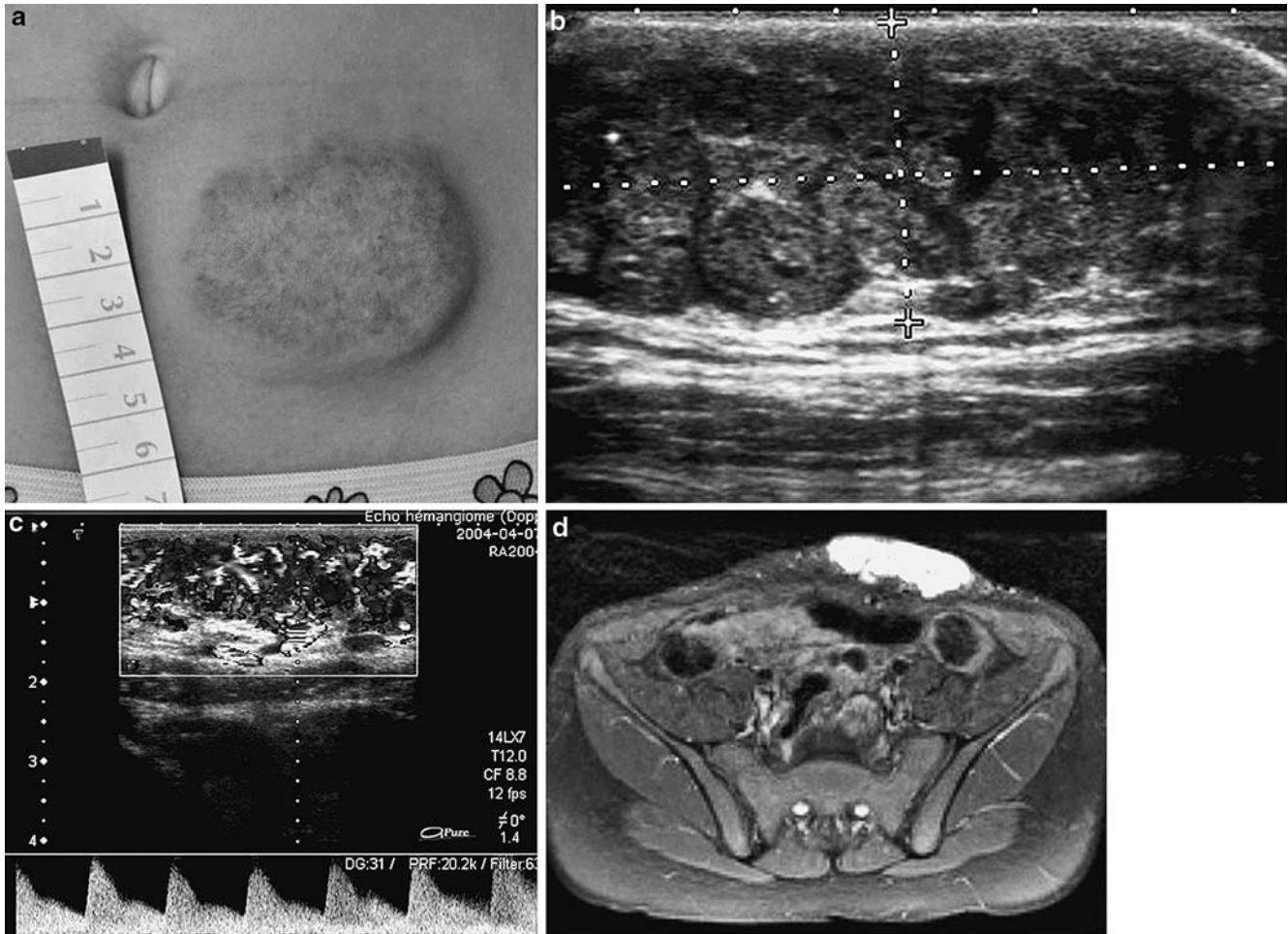


Fig. 3 Clinical and imaging findings in NICH (patient 4). **a** Photograph of patient at age 6 years shows the non-regressive lesion on the left anterior part of the abdomen. The skin is red–blue with a telangiectatic halo. **b** Transverse US scan of the lesion exhibits a very heterogeneous structure with hypoechoic areas corresponding to visible vessels. **c** Transverse US Doppler image of

the lesion demonstrates again the classical features of hemangiomas with a very high vessel density and peak systolic shift. **d** Transverse T1-weighted MR image after gadolinium and fat saturation reveals an intense and homogeneous enhancement with well-defined limits but slight fat stranding on the lateral borders of the lesion

tured peripheral edema, unlike the majority of COMMON.

None of our patients with RICH underwent angiography because regression occurred within 1 year without the clinical indication for an invasive procedure (i.e. high-output cardiac failure), so we cannot discuss previous findings [11]. Angiographically, the differential diagnosis between NICH and COMMON is possible through the venous ectasias observed in NICH (probably corresponding to the prominent vessels seen on US). The distinction between NICH and AVM can be made reliably based on the fast flow and rapid filling of the NICH and in the absence of the early venous opacification typical of AVM.

On pathology, infantile capillary hemangioma can be readily distinguished from NICH and RICH with GLUT1 antibodies. Distinguishing NICH from RICH might prove very difficult. For example, NICH removed early (2–4 years of age) is often similar or histologically indistinguishable from RICH. However, with pertinent clinical information and a skin biopsy of appropriate size, the criteria defined by Mulliken can help an experienced

physician distinguish NICH from RICH: NICH are composed of large lobules of small vessels with intervening fibrosis and dermal arteriovenous microfistulae; a large stellate vessel is often seen in the center, the endothelial cells are hobnailed, and the basement membranes are thin. RICH are composed of lobules of variable size with large extralobular vessels; the endothelium is plump; basement membranes get thicker. It is worth recalling that in the absence of biopsy in most cases, RICH diagnosis is based on clinical grounds. North et al. [12, 13] have demonstrated a major link between COMMON and the placental vasculature based on shared antigens and especially GLUT1 (glucose transporter 1). However—and this is a key diagnostic point—neither NICH nor RICH are positive for GLUT1 [5].

It is our opinion that NICH and RICH should be imaged adequately by US alone, even though the imaging findings might not be pathognomonic. A lesion that meets the criteria of hemangioma but that appears more heterogeneous, with visible vessels (especially veins) and a calcification is consistent with the diagnosis of RICH/NICH.

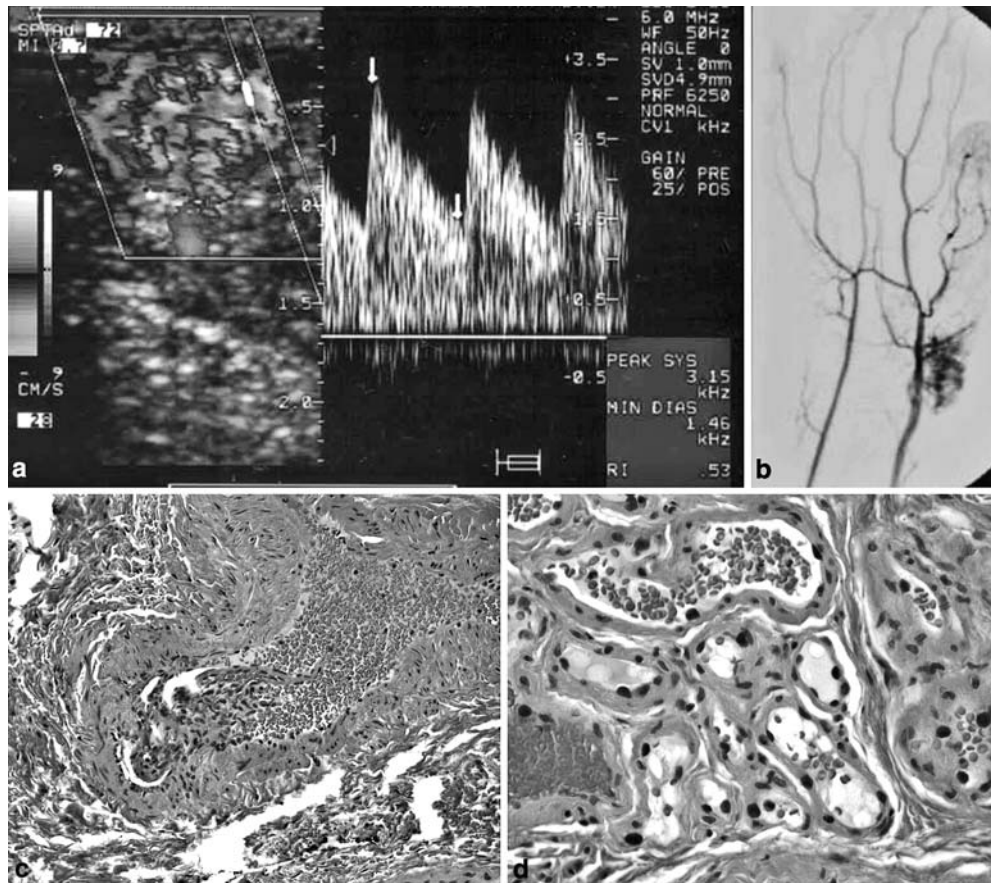


Fig. 4 Clinical, imaging and pathological findings in NICH (patient 15). **a** Coronal oblique US Doppler scan of a lesion localized on the wrist illustrates once more the classical features of hemangiomas with a very high vessel density and peak systolic

shift. **b** Selective angiography shows a capillary blush with venous ectasia. **c** Medium-power view (20× hps) demonstrates arteriovenous microfistulae. **d** High-power view (40× hps) shows hobnailed endothelial cells

The presence of microshunts could lead to a suspicion of NICH, but microshunts can also be seen initially in RICH and COMMON. Clinical follow-up will give a definite answer within 1 year. It is important to be able to inform the parents as adequately and as soon as possible about the most probable evolution of their child's lesion. To this extent, predicting the hemangiomas that will not regress is probably the most useful task we can do.

In conclusion, indicative and distinctive statistically significant imaging characteristics are observed in cases of CH distinguishing them from COMMON. These include visible vessels (vs NICH $P=0.010$, vs RICH $P=0.011$), and the presence of calcification (vs RICH $P=0.004$). US initially might be the only imaging necessary, especially when it depicts heterogeneous hemangioma-like lesions with visible vessels (especially veins), and calcification(s).

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