Characteristics of noninvoluting congenital hemangioma: A retrospective review

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Background: Noninvoluting congenital hemangioma (NICH) is a distinct vascular tumor of infancy.

Objective: We describe the clinical characteristics, histopathology, imaging, and natural history of NICH and compare our findings with previous reports.

Methods: We conducted a retrospective review of charts and photographic databases from 2 vascular anomaly centers over a 15-year period.

Results: Thirty cases of NICH were identified. All patients had fully formed vascular lesions at birth that demonstrated a nonprogressive course. The trunk and lower extremities were preferred sites and there was a female predominance. Thirteen of 30 patients reported pain. Focal necrosis and scarring was seen in a minority. Doppler studies, when performed, confirmed high vascular flow. Microscopic evaluation of 4 excised lesions showed lobular areas of endothelial cell proliferation directly adjacent to ectatic malformed vessels. Immunohistochemical studies demonstrated absence of glucose transporter-1 protein expression in every case. Wilms tumor−1 positivity was observed in lobular areas. The larger vessels did not stain with Wilms tumor−1, but some displayed D2-40 positivity.

Limitations: Patients were referred to university-based pediatric vascular anomaly centers, with potential bias toward more severe or extensive cases.

Conclusions: This retrospective study highlights the unique clinical and histopathologic features of NICH. (J Am Acad Dermatol 2014;70:899-903.)

Key words: infantile hemangiomas; noninvoluting congenital hemangioma; vascular tumors.

Congenital hemangiomas are fully formed vascular tumors at birth and display little, if any, postnatal growth. They are further differentiated from the common infantile hemangioma (IH) by their immunohistochemical profile: glucose transporter-1 protein (Glut-1) staining is characteristic of IH, but absent in congenital hemangiomas.[1,2]

Congenital hemangiomas are divided into 2 subgroups, rapidly involuting congenital hemangioma (RICH) and noninvoluting congenital hemangioma (NICH), based on clinical characteristics and natural history: RICH typically regresses by 1 year of age, whereas NICH does not regress.[3] NICH was first comprehensively described by Enjolras et al[4] in 2001

Abbreviations used:

Glut-1:	glucose transporter-1 protein
IH:	infantile hemangioma
MRI:	magnetic resonance imaging
NICH:	noninvoluting congenital hemangioma
RICH:	rapidly involuting congenital hemangioma
WT:	Wilms tumor

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Funding sources: None.

Conflicts of interest: None declared.

Accepted for publication January 9, 2014.

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Published online March 14, 2014.

0190-9622/$36.00

http://dx.doi.org/10.1016/j.jaad.2014.01.860

in a retrospective study describing 53 patients from 3 centers. This description, together with subsequent case reports and small case series, has helped define NICH as a unique vascular tumor with a relatively distinctive clinical appearance, and histopathologic and immunohistochemical profile. Imaging characteristics emphasize its high-flow nature and the lack of expansion or involution over time.

Since the first formal description, there have been only 6 additional case series, and in 4 of these NICH was discussed together with RICH.[3,5-9] We present a series of 30 cases to highlight the clinical characteristics of NICH and emphasize differences from previous reports and the more common IH.

CAPSULE SUMMARY

- Noninvoluting congenital hemangioma differs clinically and histopathologically from the more common infantile hemangioma.
- Both patch type and plaque/nodular subtypes of noninvoluting congenital hemangioma display coarse telangiectasia, background pallor, and variable deeper ectatic vessels.
- Recognition of this distinct vascular tumor will guide appropriate clinical management and counseling.

METHODS

A retrospective review of charts and photographic databases from 2 pediatric dermatology centers, the University of California, San Francisco (15-year period: 1994-2010), and Indiana University (5-year period: 2006-2010), was undertaken to identify patients with NICH. Institutional review board approval was obtained at both sites. The diagnosis of NICH was confirmed by a pediatric dermatologist (P. W. L., I. J. F., or A. N. H.) based on clinical evaluation and, when available, histopathologic examination. Cases were excluded if the clinical aspect or natural history was inconsistent with NICH, or if there was inadequate duration of follow-up to support the diagnosis. If a child was younger than 5 years at the time of diagnosis, additional information was obtained to verify that the lesion was fully formed at birth, had not undergone involution, and did not have the clinical characteristics of other vascular tumors or malformations. Demographic, clinical, and photographic data were examined in all cases. Routine histopathology was performed on specimens from 4 patients who underwent surgical excision because of reports of pain or because the bulk of the lesion was bothersome. Immunohistochemistry, including staining for Glut-1, D2-40, and Wilms tumor (WT)-1, was performed on excised specimens.

RESULTS

Clinical features

Seventeen of 30 patients (56.7%) were female and 13 (43.3%) were male. The mean age at last follow-up was 6.5 years (Table I). All lesions were solitary, although a few had adjacent, discontinuous areas of skin involvement. The mean diameter was 6.2 cm (1.5-19 cm). The majority of lesions were located on the extremities (53%) and trunk (33.3%). Two clinical subtypes were identified: a patch type (Fig 1) and nodular/plaque type (Fig 2). Both exhibited warmth, pallor, coarse dark-red telangiectasia, and areas of bluish discoloration. The nodular/plaque type varied from being minimally indurated with more gently sloped edges to exophytic with steep peripheral margins. Deeper large draining vessels were frequently seen in the surrounding skin in both subtypes. Less commonly, focal necrosis and scarring, atrophy, and satellite lesions were noted. Subtle clinical changes were documented in 10 patients who had multiple follow-up visits: soft-tissue atrophy (1), a slight increase in prominence and size of adjacent superficial veins (1), resolution of the central "spongy" area while the rest of the lesion remained unchanged (1), a slight fading of color (1), a slightly more protuberant appearance (1), and painful recurrence after surgical excision (1).

Histopathology

Four lesions were excised and the microscopic findings in lesional tissue were strikingly similar. All included lobules of small endothelial cells accompanied by increased numbers of large malformed (dysplastic) muscular vessels and smaller, thin-walled dilated vessels (Fig 3). The lobular ("proliferative") areas included tufts of endothelium, similar to those seen in IH or tufted angioma. They consisted of slightly crowded endothelial cells arrayed in relatively compact collections; evidence of hyperproliferation or an increased mitotic index was not identified. Only rare mitotic figures could be found within vascular lobules.

The lobules were present concurrently with irregular, slightly dilated vessels with angular lumina, as can be seen commonly in vascular malformations. Some ectatic vessels had thin muscular walls. The vessels with thin muscular walls resembled normal venous structures, whereas some of the dilated thin-walled vessels had slightly protuberant endothelium and resembled lymphatic channels. No arterial or