

Congenital Hemangioma

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PRACTICE POINTS

- Congenital hemangiomas (CHs) are fully developed hemangiomas that are present at birth.
- In our study, CHs were more common in males, with a predilection for the limbs.
- Infantile hemangiomas are not fully developed at birth and need many years to regress.

Congenital hemangiomas (CHs) are fully developed hemangiomas that are present at birth. There are 2 possible types: rapidly involuting CHs (RICHs) and noninvoluting CHs (NICHs). We conducted a retrospective study (2008-2012) of 6 patients (2 females, 4 males) with CHs (mean age, 16 days). We analyzed the epidemiology, clinical characteristics, and clinical outcome of CHs over this 5-year period.

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Hemangiomas are the most common benign tumors of childhood. In recent years, subsets of hemangiomas that are fully formed at birth have been recognized as clinically and biologically distinct from the classic infantile hemangioma (IH). Congenital hemangiomas (CHs) are classified based on clinical course as rapidly involuting CHs (RICHs) or noninvoluting CHs (NICHs). The aim of this retrospective study was to describe the epidemiology, clinical aspects, and clinical outcome of CH over a 5-year period.

Methods

Using electronic medical records from the department of dermatology (Hedi Chaker Hospital, Sfax, Tunisia) for a 5-year period (2008-2012), we

searched for *hemangioma*. After collecting those records, we identified patients with CHs. We studied the epidemiologic (eg, sex, age), clinical course (eg, location, size, number, color, surrounding skin), and evolutionary aspects in these patients.

Results

Twenty IHs were identified, 6 (30%) of which were considered CHs. The clinical characteristics of the 6 patients are summarized in the Table. We identified 2 females and 4 males aged 2 to 60 days (mean age, 16 days). Four patients had CHs involving the limbs (knee [n=2]; ankle [n=1]; elbow [n=1]) and 2 patients had CHs involving the trunk. Congenital hemangiomas were singular, oval shaped, and surrounded by a clear halo in all 6 patients. They presented as exophytic masses (n=3) or bossed plaques (n=3). A blue hue was noted in 5 patients and a purple hue in 1 patient. In some cases, telangiectasia (n=2) or small areas of necrosis (n=1) were noted at the center of the CHs. The CHs ranged in size from 3 to 6 cm (mean, 4 cm). Doppler ultrasonography was performed in 2 patients and showed fast blood flow. It is well known that manipulating a CH when it is ulcerative may cause a fatal hemorrhage. Thus, parents/guardians should be cautious when cleaning and dressing the lesions. Regular follow-up was recommended to all patients as noted in the medical records. The lesion involuted in 4 patients after a mean period of 6 months, which allowed us to classify the lesions as RICHs (Figure, A). Two CHs were persistent after 2-year (Figure, B) and 4-year (Figure, C) follow-up, which was consistent with NICH classification.

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Clinical Characteristics of Congenital Hemangiomas

Patient No.	Age, d	Sex	Clinical Presentation	Size, cm	Location of CH	Investigation	Clinical Course
1	60	M	Exophytic mass, blue hue, clear halo	4×2	Left knee	None	Involuting (after 3 mo)
2	7	M	Bossed plaque, blue hue, clear halo	6×3	Left knee	None	Involuting (after 6 mo)
3	4	F	Bossed plaque, blue hue, clear halo, telangiectasia	4×2	Left elbow	None	Unchanged (4 y)
4	15	M	Exophytic mass, blue hue, clear halo	4×2	Right ankle	None	Involuting (after 6 mo)
5	2	F	Bossed plaque, blue hue, clear halo, telangiectasia	4×2	Trunk	Doppler ultrasonography revealed fast blood flow	Unchanged (2 y)
6	7	M	Exophytic mass, purple hue, clear halo, small areas of necrosis	3×2	Trunk	Doppler ultrasonography revealed fast blood flow	Involuting (after 6 mo)

Abbreviations: CH, congenital hemangioma; M, male; F, female.

Comment

Since 1996, vascular anomalies have been classified either as tumors or malformations.¹ Infantile hemangioma is the most common vascular tumor and presents as an endothelial cellular proliferation that develops within days after birth. Congenital hemangiomas are fully developed at birth^{2,3} and are classified as RICHs and NICHs according to their clinical outcome.

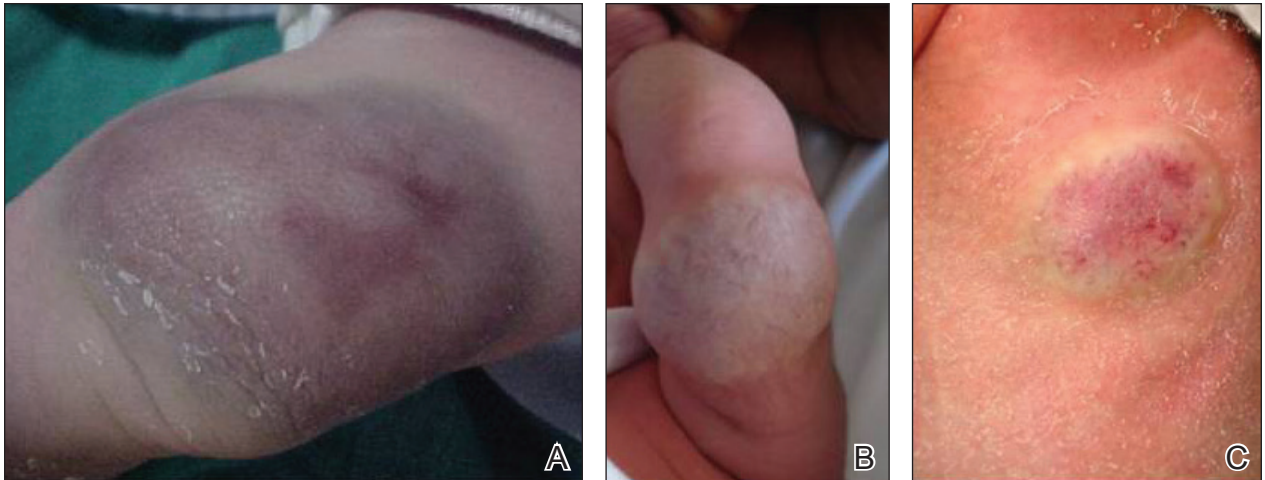
As expected, our analysis revealed that CH usually is solitary and may present as a small lesion (eg, a few millimeters) but also may be large in size.⁴ Congenital hemangioma has an equal sex distribution and a predilection for the head and limbs near a joint. In contrast, IH exhibits female predilection and can occur anywhere on the body.^{4,6} In our study, CHs were more common in males and had a predilection for the limbs. Three patients presented with exophytic masses with a clear halo and overlying telangiectasia, which are commonly described features in CH.^{4,6}

In the classification of vascular anomalies, RICHs and NICHs are fast-flow lesions that are

indistinguishable at birth.^{7,8} Untreated, RICHs usually resolve in the first 14 months of life, often resulting in an area of atrophic or excess skin.^{8,9} Noninvolving CHs persist and grow in proportion with the patient.¹⁰⁻¹²

When Doppler ultrasonography findings are inconsistent with a CH, an early biopsy from the periphery of the lesion may be performed to exclude an uncommon soft-tissue tumor such as infantile myofibromatosis or sarcoma.^{8,9,12} Because of the presence of a clear halo in all cases and mainly rapid involution of CHs, these differential diagnoses were dismissed. The histologic appearance of RICH differed from NICH and common IH, but some overlap was noted among the 3 lesions. Rapidly involuting CH was composed of small to large lobules of capillaries with moderately plump endothelial cells and pericytes; the lobules were surrounded by abundant fibrous tissue.⁹

Despite the notable differences in natural history between RICHs and NICHs, as RICHs regress within months while NICHs do



A rapidly involuting congenital hemangioma with a blue bossed plaque and a clear halo on the left knee (patient 2)(A). A noninvoluting congenital hemangioma with a blue bossed plaque surrounded by a clear halo with central telangiectasia on the left elbow (patient 3)(B). A noninvoluting congenital hemangioma with a blue bossed plaque with central telangiectasia and surrounded by a clear halo on the trunk (patient 5)(C).

not, both classes of CH share an important immunohistochemical phenotype; they do not express glucose transporter 1, the marker of IH.¹³ Tests for this marker were not performed in our study. The prognosis of CH generally is good, and special management is not required.

Conclusion

Rapidly involuting CHs and NICHs have many similarities, such as appearance, location, and sex distribution. The obvious differences in behavior serve to differentiate RICHs, NICHs, and common IHs. Infantile hemangiomas are not fully developed at birth and need many years to regress.

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