

CASE AND RESEARCH LETTER

[Translated article] Infantile Hemangiomas of the Vulvar Region: A Therapeutic Challenge

Hemangiomas infantiles de localización vulvar: un reto terapéutico

To the Editor,

Infantile hemangioma (IH) is the most common benign tumor of infancy, affecting 1% up to 10% of children younger than 1 year.^{1–3} IHs can occur anywhere on the body but are most frequently found on the head and neck.¹ Approximately 1% of cases develop in the genital area.⁴

When managing vulvar IHs, several factors should be considered, including their natural history, location, involvement of functionally significant structures, ulceration, symptoms, and the potential for long-term sequelae.^{1–3} Most IHs are small and tend to regress spontaneously, allowing for an expectant management approach.^{1–3} However, 5% up to 10% require early active therapy to prevent anatomical distortion.^{2,3,5,6}

The objective of this study is to describe the clinical and evolutionary characteristics and treatment of vulvar IHs, proposing a diagnostic-therapeutic algorithm. We conducted a retrospective review of vulvar IHs seen at a tertiary referral center dermatology department from 2016 to 2023, including cases with clinical images and a 6-month minimum follow-up.

A total of 10 patients were included, whose clinical characteristics are shown in [table 1](#). All had appropriate birth weights for their gestational age, with a median weight of 2982 g (range, 2600–4000 g). A total of 90% were born at term, with a median gestational age of 39 weeks, except for 1 preterm birth on week 36. Pregnancies were uneventful, except for 1 uncontrolled pregnancy requiring C-section delivery due to the risk of neonatal infection by *Streptococcus agalactiae*.



Table 1 Characteristics of the patients, hemangiomas, and treatment received.

Vulvar infantile hemangiomas (n = 10)	
<i>Gestational age at birth (weeks), median</i>	39
<i>Birth weight (grams), median</i>	2982
<i>Maternal conditions (number, %)</i>	
Diabetes gestational	1 (10%)
Hypothyroidism	1 (10%)
None	8 (80%)
<i>Type of delivery (number, %)</i>	
Vaginal	6 (60%)
Cesarean	4 (40%)
<i>Location of IH (number, %)</i>	
Labia majora	5 (50%)
Labia minora	3 (30%)
Clitoris	2 (20%)
<i>Morphology of IH (number, %)</i>	
Focal	7 (70%)
Segmental	1 (10%)
Indeterminate	2 (20%)
<i>Depth of IH (number, %)</i>	
Superficial	8 (80%)
Deep	0 (0%)
Mixed	2 (20%)
<i>Largest diameter (mm)</i>	
Range	3–50
Mean	15.22
<i>Ulceration</i>	
Yes	3 (30%)
No	7 (70%)
<i>Treatment</i>	
Oral propranolol	4 (40%)
Topical timolol 0.5%	8 (80%)
PDL laser	3 (30%)
Surgery	1 (10%)

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Figure 1 Clinical images of vulvar IHs. Mixed ulcerated IH in a 2-month-old girl (A). Three months into propranolol 3 mg/kg/day (B) and at the end of treatment (9 months) (C). Segmental vulvar and perianal IH in a 7-month-old girl (D). Five months into propranolol 3 mg/kg/day (E) and at the end of treatment (7 months) (F). Superficial focal IH in the clitoris of a 6-month-old girl (G). At 12 months of age, on timolol 0.5% gel and PDL (H). One month into propranolol with decreased volume and erythematous component (I) – lost to follow-up.

All IHs were found on external genitalia (5 on the labia majora, 3 on the labia minora, and 2 on the clitoris), with a median diameter of 8 mm (range, 3–50 mm). A precursor lesion was noted at birth in 50% of the cases. Most IHs were focal (70%), with a smaller proportion being indeterminate (20%) or segmental (10%) (Fig. 1). A total of 8 lesions were superficial, and 2 were mixed. In 1 segmental IH, magnetic resonance imaging (MRI) of the lumbosacral spine and pelvis ruled out the presence of any associated malformations. The IH of 1 of the patients occurred in the context of benign neonatal hemangiomatosis, with 4 additional lesions on the left leg, trunk (2), and face. Visceral IHs were ruled

out via abdominal ultrasound. Three cases (30%) developed ulceration at the follow-up.

All patients received treatment, either monotherapy ($n=7$) or combination therapy ($n=3$), including oral propranolol ($n=4$), topical timolol ($n=8$), pulsed dye laser (PDL) ($n=3$), and excision with electrocautery for one pedunculated IH. Patients treated with oral propranolol started at a median age of 5 months and were dosed at 3 mg/kg/day for a median duration of 7 months. At the follow-up, 50% achieved complete resolution, 20% showed partial regression, and 30% had stable lesions, with a median follow-up of 12 months.

Anogenital IHs are considered high risk due to their greater tendency to ulcerate and associate with various congenital anomalies.⁵

The most common complication is ulceration, which can affect 53% of cases vs 11.54% globally for IHs.⁵ Predictors of ulceration include segmental or indeterminate morphology, mixed IHs, location on buttocks or perianal area, and ≥ 5 cm diameters.^{5,7} In these cases, early treatment with oral propranolol should be considered, before the 5th month of life, with a therapeutic dose of 3 mg/kg/day for, at least, 6 months.^{1–3}

Perineal or lumbosacral IHs, especially if large and segmental, may be associated with congenital anomalies (pelvic or sacral or lumbar syndrome).^{1,2} In these cases, an MRI of the lumbosacral spine and pelvis is recommended to rule them out. In children under 6 months, lumbar canal and abdominal-pelvic ultrasound can be considered as part of the initial screening.² In the largest series of anogenital IHs, congenital anomalies were found in 6.4% of cases, with the most common ones being urogenital anomalies and myelopathy.⁵ These associations were more common in penile, sacral, and perianal IHs vs vulvar IHs.⁵

The location of the clitoris is especially sensitive due to its functionality and the risk of permanent deformity. In our series, it affected 20% of patients, representing a significant therapeutic challenge. Initially, they were treated with timolol and PDL with little response, so in 1 case, propranolol was initiated. It is recommended to consider propranolol treatment at this location from the beginning.

Alternatively, topical timolol 0.5% gel applied twice daily could be used for fine and superficial non-ulcerated IHs, with an adequate safety and efficacy profile.⁸ PDL is also useful for improving the texture of residual lesions and treating telangiectasias.⁹

The above-mentioned description is illustrated in Fig. 2 where we propose the management algorithm for vulvar IHs.

In conclusion, vulvar IHs, although rare, can present a high complication rate. They should be considered high risk and referred to a specialist early. Ulcerated, segmental, mixed IHs, those with diameters ≥ 5 cm, and those located in the clitoris require treatment with propranolol to minimize complications.

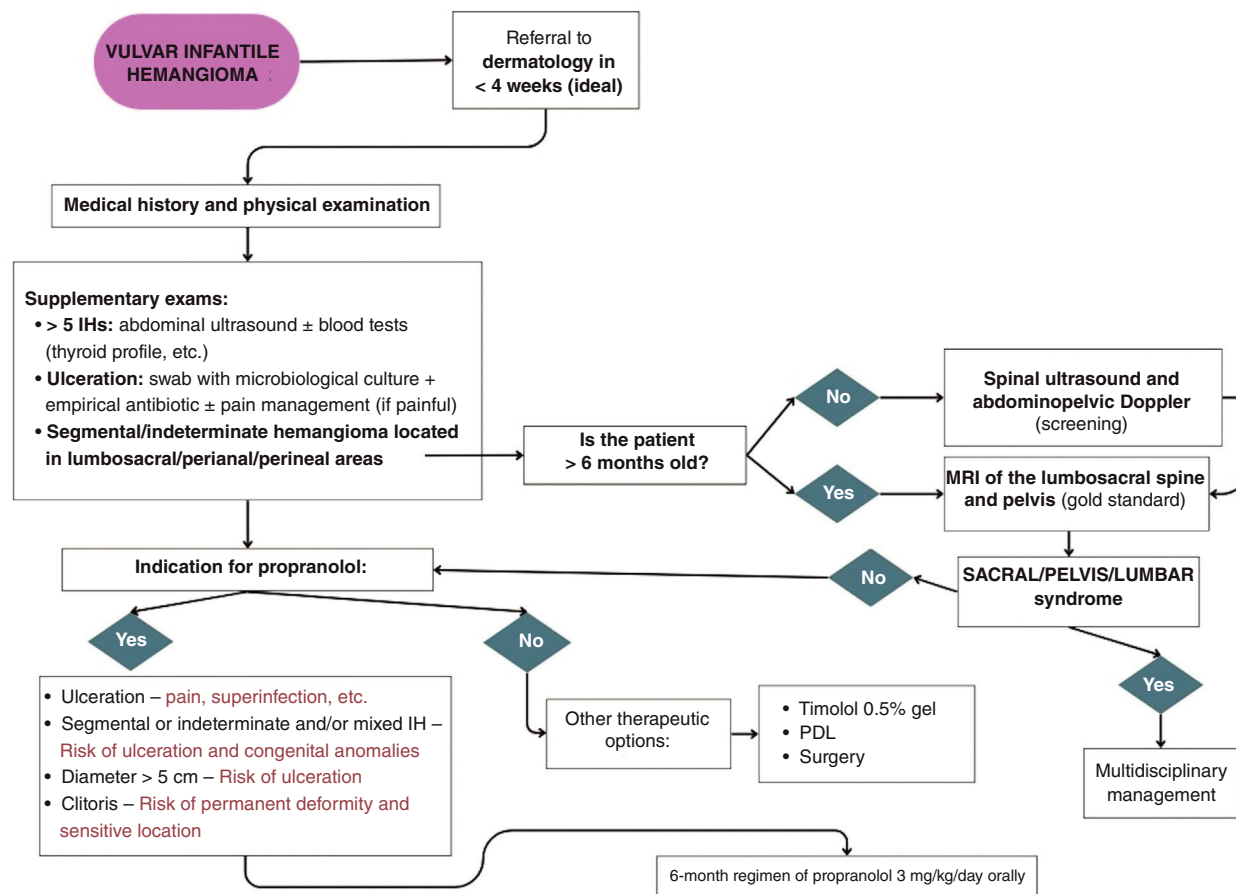


Figure 2 Diagnostic and therapeutic algorithm for vulvar IHs. IH: infantile hemangiomas; PDL: pulsed dye laser; MRI: magnetic resonance imaging.

Informed consent

All patients provided informed consent for the publication of their case details.

Conflicts of interest

None declared.

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