Dermatology Reports

Dermatology Reports 2026; 18:10351
doi:10.4081/dr.2026.10351

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A rare presentation of infantile hemangioma with minimal or arrested growth in PHACE(S) syndrome

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Key words: PHACE(S) syndrome; hemangioma; IHMAG; propranolol.

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Dear Editor,

PHACE(S) syndrome is a neurocutaneous disorder associated with segmental infantile hemangiomas (IHs).[1] These hemangiomas typically manifest as rapidly growing lesions on the face or upper trunk. However, this case presents an unusual phenotype – an IH with minimal or arrested growth (IHMAG).[2]

IHMAGs differ from segmental IHs in their limited growth, higher involution potential, and distinct presentation. Importantly, the growth phenotype (IHMAG) and the morphologic distribution (segmental or non-segmental) are independent features and may coexist.

The rarity of retrobulbar involvement in PHACE(S) syndrome further underscores the need for careful classification of hemangioma subtypes.

A 1-month-old male infant was referred to the pediatric clinic for the presence of a reticular-pattern vascular lesion localized on the left frontotemporal region. The lesion showed minimal external proliferation. Dermoscopy revealed a faint pink background with fine whitish reticular lines and scattered white dots, without well-formed vascular lacunae or prominent vessels. This pattern differs from classic IHs, which show homogeneous red coloration and vascular structures, and from capillary malformations, typically characterized by linear or arborizing vessels, supporting the diagnosis of IHMAG (Figure 1).

Imaging revealed a primary retrobulbar hemangioma, demonstrating high-flow characteristics on contrast-enhanced ultrasound (CEUS), differentiating it from conventional IHs. Cardiological assessment detected a minimal left-to-right shunt through an apical ventricular septal defect and the foramen ovale with normal chamber dimensions and contractility. Ophthalmological examinations showed no functional ocular impairment.

Neurological magnetic resonance imaging identified hypoplasia of the left carotid artery, hypoplasia of the left A1 segment of the anterior cerebral artery, and agenesis of the left subclavian artery. Based on the association of segmental hemangioma, arterial anomalies, and cardiac defects, a diagnosis of PHACE(S) syndrome was established. Unlike most PHACE(S)-associated hemangiomas, which are typically classic proliferative IHs, this case showed a retrobulbar hemangioma with an IHMAG growth phenotype. Although the association between IHMAG and PHACE(S) is uncommon, it is not pathognomonic and does not define a distinct vascular subtype.

The introduction of beta-blockers, particularly propranolol, has revolutionized the management of childhood hemangiomas and is the first-line systemic therapy with established safety data.[3] In PHACE(S) syndrome, cerebrovascular abnormalities necessitate careful evaluation before initiating propranolol to mitigate the risk of ischemic events. Our patient underwent prolonged propranolol therapy with progressive and complete regression of the lesion, without adverse events (Figure 1).

Dermoscopy plays an important role in the differential diagnosis between IHMAG, classic proliferative IHs, and capillary malformations. IHMAG typically shows pale reticular telangiectatic vascular patterns without dense red lacunae. Classic IHs usually exhibit closely packed red lacunae corresponding to proliferative vascular lobules, while capillary malformations display flat, homogeneous erythematous areas lacking involution and high-flow behavior. IHMAG should be regarded as an uncommon hemangioma subtype that may occur in association with PHACE(S) syndrome but does not define a distinct or pathognomonic vascular phenotype, instead reflecting a rare overlap within the broader clinical spectrum of the disease. Retrobulbar IHs in PHACE(S) syndrome have been documented, but they typically describe classic IHs rather than IHMAGs.-Differential diagnosis of retrobulbar lesions also includes venous malformations and arteriovenous fistulas. CEUS was essential in confirming the high-flow nature of the lesion and its hemangiomatous behavior.

Unlike rapidly proliferating segmental IHs, IHMAGs may not require aggressive intervention. Prolonged propranolol therapy remains the preferred treatment, but conservative management should be considered based on lesion behavior.

Given the vascular abnormalities in PHACE(S) syndrome, long-term neurovascular follow-up is imperative. Regular monitoring of heart rate, blood pressure, and signs of hypoglycemia is essential during propranolol therapy.

While no serious adverse reactions or stroke cases have been directly linked to propranolol in PHACE(S) patients,[3] the syn-

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Figure 1. a) Clinical presentation of the patient at 1 month of age, demonstrating a reticular-pattern hemangioma localized in the left frontotemporal region. The lesion exhibited minimal external proliferation, with no significant growth over time. Imaging studies identified the primary site of hemangioma proliferation as retrobulbar, distinguishing it from conventional segmental IHs typically seen in PHACE(S) syndrome. CEUS confirmed the high-flow characteristics of the retrobulbar lesion, supporting the diagnosis of IHMAG. **b**) Follow-up clinical evaluation at 2 years of age, showing significant regression of the reticular-pattern hemangioma following 18 months of propranolol therapy. The lesion appears faint and less pronounced, with no signs of residual proliferative activity, consistent with the high-involution potential of IHMAG. The patient remained asymptomatic, with no evidence of functional impairment or ocular complications. **c**) Dermoscopy shows a faint pink background with fine whitish reticular lines and scattered white dots, without well-defined vascular lacunae or prominent linear or arborizing vessels.

drome's high incidence of arteriopathy increases stroke risk. Most PHACE(S)-related strokes occur in patients with major cerebral artery anomalies. Further studies are needed to evaluate the impact of systemic therapies on ischemic risk in PHACE(S) patients with severe arteriopathy.[4]

This case presents a rare form of IHMAG in PHACE(S) syndrome, characterized by a retrobulbar proliferative component rather than the classic segmental IH. Advanced imaging techniques, particularly CEUS, are crucial for accurate diagnosis. IHMAG should be recognized as a distinct hemangioma subtype, warranting differentiation from segmental IHs. Given its involutional potential, conservative management strategies may be viable in select cases. This case contributes to a broader understanding of vascular anomalies in PHACE(S) syndrome and highlights the need for further classification studies and optimized management strategies for these rare presentations.

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Received: 12 March 2025; Accepted: 5 January 2026.

Contributions: all authors contributed equally to the study conception and manuscript preparation. All authors reviewed and approved the final version.

Conflict of interest: the authors have no conflict of interest to declare.

Ethics approval and consent to participate: no ethical committee approval was required for this case report by the Department, because this article does not contain any studies with human participants or animals. Informed consent was obtained from the patient's legally authorized representative.

Consent for publication: written informed consent was obtained from a legally authorized representative for the publication of this article and any accompanying images.

Availability of data and materials: not applicable.

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