

## PHACE Syndrome: Dramatic Response to Propranolol

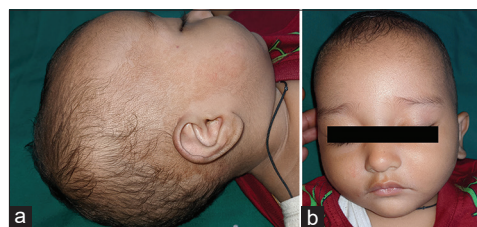
A 2-month-old pre-term (gestational age 30 weeks) singleton female baby presented with a large, soft, and lobulated dusky erythematous plaque on the right lateral aspect of the face since a week of birth, suggestive of a segmental infantile hemangioma [Figure 1a]. Similar, smaller plaques of size 1–1.5 cm were seen as satellite lesions near the nasolabial fold. This was associated with a gross right orbital swelling on the same side associated with proptosis of the globe [Figure 1b]. A clinical suspicion of PHACE(S) syndrome (posterior fossa malformations, hemangioma, arterial anomalies, cardiac

abnormalities/coarctation of the aorta, eye anomalies  $\pm$  supraumbilical raphe/sternal cleft) was considered. There was no history of cardiac or neurological symptoms and no sternal defect on physical examination. A screening magnetic resonance imaging with angiography of the head and neck showed right cerebellar hypoplasia [Figure 1c] and right orbital T1/T2 hyper-intense homogeneous soft tissue mass involving extraconal space, suggestive of orbital hemangioma [Figure 1d], fulfilling the definite diagnostic criteria (hemangioma  $>5$  cm of the head plus one major criterion in the form of posterior fossa anomalies).<sup>[1]</sup> Coarctation of the aorta was also detected, whereas the rest of the vasculature architecture of brain was normal on angiography. The baby was started on oral propranolol 0.5 mg/kg/day titrated up to 2 mg/kg/day under cardiac monitoring and standard precautions in the form of administration of drug post feed, with complete resolution over a period of 6 months. Propranolol was subsequently tapered to 1 mg/kg with a sustained response at 8 months [Figure 2a and b].

The use of beta blockers in PHACE syndrome was earlier considered to be



**Figure 1:** (a) Lateral view of the right side of the face with well-defined, dusky erythematous, soft lobulated plaque suggestive of infantile hemangioma ( $>5$  cm size). (b) Anterior view showing smaller plaques on the cheek and right orbital proptosis. (c) Right cerebellar hypoplasia seen in the coronal section on MR angiography (white arrow). (d) Right orbital T1/T2 hyper-intense homogeneous soft tissue mass involving extraconal space, suggestive of orbital hemangioma (red arrow)



**Figure 2:** (a and b) Complete resolution of hemangioma with regression of orbital proptosis at 8 months of follow-up

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**How to cite this article:** Sharma A, Parvathy S. PHACE syndrome: Dramatic response to propranolol. Indian Dermatol Online J 2023;14:443-4.

Received: 20-Sep-2022. Revised: 01-Dec-2022.

Accepted: 13-Dec-2022. Published: 27-Apr-2023.

### Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.idoj\_499\_22

### Quick Response Code:



contraindicated when vascular anomalies were present, but recent studies have demonstrated safety and good efficacy.<sup>[2,3]</sup> We demonstrate a case of PHACE syndrome with coarctation of aorta, with safe use of propranolol and dramatic response.

### ***Declaration of patient consent***

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/ have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

### ***Financial support and sponsorship***

Nil.

### ***Conflicts of interest***

There are no conflicts of interest.

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