

Excellent response of infantile hemangioma associated with PHACES syndrome to propranolol

Sir,

Facial segmental infantile hemangiomas are said to be associated with PHACES, a neurocutaneous syndrome comprising of *Posterior fossa brain malformations, cervicofacial segmental Hemangiomas, Arterial anomalies, cardiac defects or coarctation of aorta, eye anomalies and sternal defects such as sternal clefting or supraumbilical raphe.*^[1] Herewith, we report a 2.5-month-old male infant with PHACES syndrome with good response of the hemangioma to treatment with propranolol.

A 2.5-month-old male infant was brought to us with a large red-colored lesion on the right side of the face and scalp from birth. He was a term baby born of a non-consanguineous marriage with assisted vaginal delivery having a birth weight of 3 kg and normal APGAR score. On physical examination, the child was active and feeding well. The head circumference was 43 cm as against the expected 38 cm at 2 months of age. There was no neurological deficit. The child had a large segmental infantile hemangioma extending over the right side of the face measuring 14 cm x 7 cm in the maximum diameter. He also had a small hemangioma on the left eyelid [Figure 1a]. His hemogram showed a normal profile with normal platelet count. Thyroid function tests were within normal limits. Computerized axial tomography and magnetic resonance imaging of the brain revealed a cyst in the posterior fossa communicating with the fourth ventricle resulting in hydrocephalus [Figure 2]. Echocardiography of the heart revealed a small patent ductus arteriosus (PDA). Eye, ear, nose and throat examination did not reveal any abnormalities.

The infant was put on low-dose propranolol (0.17 mg/kg per dose three times daily) under careful monitoring of heart rate and blood glucose levels. The dose was gradually increased over 3 days to 2 mg/kg/day. The child was discharged and propranolol was continued for 4 months with significant regression of the hemangioma [Figure 1b]. Following this, surgery to correct the Dandy-Walker malformation was planned by neurosurgery, during the course of which propranolol was stopped. There was no rebound flare in the size of the plaques.



Figure 1: (a) Large segmental infantile hemangioma involving the right side of the face and forehead. (b) Significant regression in the size of the hemangioma following administration of propranolol after 4 months



Figure 2: Computerized tomography of the brain revealing a posterior fossa cyst communicating with the fourth ventricle and dilated lateral ventricles

It has been estimated that 2% of all patients with hemangiomas meet the diagnostic criteria for PHACES syndrome.^[2] Diagnostic criteria include the presence of hemangiomas and at least one of the extracutaneous anomalies described. Modified criteria have been proposed for definitive and possible diagnosis of PHACES syndrome.^[3] Our patient had cervicofacial hemangiomas with Dandy-Walker malformation, hydrocephalus and a small PDA.

Treatment of hemangiomas of PHACES syndrome is similar to that of infantile hemangiomas. Patients have been treated with oral steroids, intralesional steroids, vincristine, interferon- α and pulse-dye laser.^[2] Surgical repair has been advocated for the aortic arch anomalies after comprehensive assessment of the vasculature.^[4] We considered propranolol in this patient in order to avoid the long-term complications of systemic steroids. Our patient was started on

propranolol after careful work-up, monitored for side-effects and maintained on propranolol until surgery with good response. Problems commonly anticipated with propranolol are bradycardia and hypoglycemia. In the setting of PHACES, propranolol can mask signs of heart failure, worsen cardiac function or induce hypotension leading to a theoretical risk of brain infarcts in patients with anomalous cerebral vasculature.^[5] There have been few recent reports of safe use of propranolol in the setting of PHACES.^[6,7] We report this case to highlight the fact that propranolol can be used in the setting of PHACES under careful monitoring.

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