

A Newborn with Facial Hemangioma and Sternal Defect

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ABSTRACT

Large facial segmental hemangiomas tend to be associated with extracutaneous abnormalities. We report a case of large progressive segmental facial hemangioma, sternal cleft and talipes equinovarus deformity.

Key words:

Infantile hemangioma, neurocutaneous syndrome, PHACE association, segmental hemangioma, sternal defect

INTRODUCTION

We report a full-term singleton male neonate with large progressive segmental facial hemangioma, which had a



Figure 1: U-shaped midline sternal cleft which is covered by a raphe in its lower half, together with a well-demarcated, raised, erythematous, plaques over the cheeks, chin, lower lips (upper lips is spared), ear pinna, tragus, neck and upper chest



Figure 2: A surgically repaired sternal cleft with more evident segmental hemangioma

mandibular pattern of distribution with sparing of the upper lips. In addition, the newborn was noted to have a midline sternal cleft whose lower half was covered by raphe [Figure 1] and talipes equinovarus deformity. Furthermore, ophthalmologic examination, laryngoscopy, abdominal sonogram, echocardiography and magnetic resonance imaging/angiography of the head, neck and chest revealed normal results.

A study published in 1996, Frieden *et al.* were the first to coin the acronym of PHACE syndrome to describe a neurocutaneous syndrome, which includes Posterior fossa malformations, Hemangiomas, Arterial anomalies, Cardiac defects/Coarctation of the aorta and Eye abnormalities^[1] and since then several cases have been reported.

Infantile hemangiomas usually follow a benign course. However, segmental hemangiomas, which exhibit a linear involvement of a specific cutaneous territory, usually have more aggressive behavior than localized lesions. Furthermore, the risk of detection of PHACE syndrome in patients with large facial segmental hemangioma was estimated to be 31%.^[2]

In the year 2008, an expert panel meeting was conducted to establish consensus criteria for PHACE syndrome.

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Diagnosis of definite PHACE is fulfilled by co-existence of a characteristic segmental facial hemangioma, in addition to one major criterion or two minor criteria.^[3] Our patient had a large segmental hemangioma on the face, together with sternal cleft (major criterion). Therefore, these findings are consistent with the diagnosis of PHACE.

Our patient underwent a plastic surgery to repair the sternal defect by pectoralis major muscle flap after proper closure of the pericardium [Figure 2]. Patient was maintained upon oral prednisolone at a dose of 2-3 mg/kg/day for 1 month, before pulsed dye laser was performed.

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