



HAEMANGIOMAS AND VASCULAR MALFORMATIONS

PHACE (S) SYNDROME: PROPRANOLOL USE

M Abid⁽¹⁾ - S Ben Moussa⁽²⁾ - S Amal⁽²⁾ - O Hocar⁽²⁾

Uhc Mohammed Vi, Fmpm, Uca, Department Of Dermatology And Venereology, Marrakech, Morocco⁽¹⁾ - Uhc Mohammed Vi, Fmpm, Uca, Department Of Dermatology And Venereology, Marrakech, Maroc⁽²⁾

Introduction: Hemangiomas are the most common benign tumors in childhood. In rare cases, they can be associated with dysmorphic malformations. The acronym, the PHACE (S) syndrome, includes a large facial or cervical Hemangioma, associated with one or more of the following systemic abnormalities including: Posterior fossa malformation, Arterial abnormalities, Coarctation of the aorta and/or cardiac defects, Eye abnormalities and Sternal clefts.

Case report: Five cases of PHACE syndrome were diagnosed between 2011 and 2018 in our hospital. All patients were female and presented with an infantile facial hemangioma greater than 5cm in diameter: with V1 involvement in three, V1 and V2 in one patient and bilateral V3 with cervical involvement in the fourth patient who also had dyspnea with stridor. In 3/5 of these, posterior fossa abnormalities were present and no vascular malformation. Two patients had an ophthalmological abnormality. None had cardiac, aortic or sternal involvement. All patients were treated with propranolol with good tolerance and efficacy. The neurological development of children was normal.

Discussion: Large facial or cervical hemangiomas can be associated with one or more systemic abnormalities described by the PHACE (S) acronym. Its prevalence is unknown, but it shows marked female preponderance. Neurological examination and cerebral MRI to rule out abnormality of the posterior fossa, completed by a sequence of angio-MRI in the search for cerebral artery malformations; cardiovascular exploration, completed by echocardiography in the case of doubt and examination of the eyes and sternum should be performed systematically.

Conclusion: This series is in agreement with data from the literature, particularly on the clear predominance of women and the frequent involvement of the posterior fossa. It also recalls the effectiveness of propranolol in this indication.

