



IMAGES IN PAEDIATRICS

Capillary malformation of the lower lip: CLAPO syndrome



Malformación capilar del labio inferior: síndrome CLAPO

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Vascular malformations constitute a broad spectrum of disorders caused by the abnormal development of blood vessels. Capillary malformations (CMs) are the most frequent type of vascular anomalies and affect up to 0.3% of the population.

We present the case of a boy aged 8 months referred to us on account of a CM of the lower lip, lymphatic malformation (LM), asymmetry in the right side of the face (Fig. 1A) and overgrowth of the first toe in both feet with bilateral sandal gap deformity (Fig. 1B and C). A PIK3CA-related overgrowth spectrum (PROS) syndrome was suspected, prompting performance of a brain magnetic resonance scan that ruled out central nervous system involvement and leading to diagnosis of CLAPO syndrome.

CLAPO syndrome is characterised by capillary vascular malformation of the lower lip, lymphatic malformations

of the head and neck, asymmetry and partial/generalized overgrowth¹. It is a sporadic disorder associated with somatic changes in the *PIK3CA* gene and other PROS syndromes, such as megalencephaly-capillary malformation syndrome (MCAP), which explains the overlap of the phenotypes of these two syndromes². The CM of the lower lip is characteristic of CLAPO syndrome, so in the case of a CM in the gums or lower lip, it is important to monitor the patient for the development of a LM or overgrowth, while in the case of a CM of the upper lip, MCAP syndrome should be suspected and neurological involvement must be ruled out. In this sense, CMs are important findings that, based on their location, may guide the differential diagnosis with other underlying anomalies or syndromes.

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Figure 1 A) Capillary malformation between the brows and in the lower lip (arrow) with right facial asymmetry (arrowhead). Overgrowth of the first toe of the feet (arrowhead) with bilateral sandal gap (double-headed arrow): B) right foot, C) left foot. Phenotype compatible with CLAPO syndrome.

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Conflicts of interest

The authors have no conflicts of interest to declare.

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