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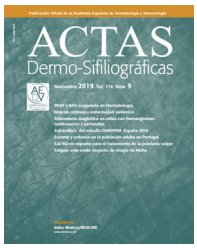

RESIDENT'S FORUM

Noninvoluting Congenital Hemangiomas That Exhibit Postnatal Growth☆

Hemangiomas congénitos no involutivos (NICH) con crecimiento postnatal

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PALABRAS CLAVE
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Congenital hemangioma (CH) is a type of vascular tumor that develops in the uterus and is therefore fully formed at birth. CH differs from infantile hemangioma (IH) both clinically and histologically. Immunohistochemistry for the marker Glut-1 (glucose transporter-1) is characteristically positive in IH and negative in CH.[1]

CH can be divided into three subgroups based on tumor course: rapidly involuting CH (RICH); non-involuting CH (NICH); and partially involutional CH (PICH). RICH is characterized by regression between 6 and 14 months, sometimes without leaving a residual lesion, and in other cases leaving progressive underlying lipoatrophy after rapid involution. NICH does not involute. PICH initially behaves like RICH but involution stops at a certain point, leaving a lesion indistinguishable from NICH. There is marked clinical overlap between the different forms of CH. PICH was described later than RICH and NICH,[2] and therefore it is highly likely that some cases of NICH described in the literature actually correspond to PICH. Distinction between PICH and NICH is difficult in the absence of images acquired during the first days of life.

NICH is slightly more common in girls and more frequently appears on the trunk and extremities. This is a vascular-like lesion of which two clinical variants have been described: a macule with a slightly atrophic surface that may exhibit mild induration or turgor on palpation; and a nodular or plaque-like lesion. In both cases a slight increase in temperature can be perceived upon palpation and Doppler ultrasound reveals an increase in flow.[1,2] Two recent publications described possible atypical postnatal growth of NICH.[3,4] Until recently there were only isolated reports of this type of hemangioma. However, Cossio et al[4] reported a series of 80 NICH, of which 9 (11.2%) exhibited growth after a period of stability of 2–10 years (mean, 5.3 years). Furthermore, the authors reported the appearance of telangiectasias, reddish, occasionally bleeding papules, and pyogenic granulomas on these tumors.[4]

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It has been suggested that persistent high flows in the vessels of these lesions may account for late growth, and that this could be much more frequent than reported, since in many cases CH is operated on before adolescence.[3,4] In both NICH and RICH somatic mutations have been described in codon 209 (Gln209) in GNAQ and GNA11, suggesting that their course is determined by other genetic, epigenetic, or environmental factors.[5]

In conclusion, CH is a group of rare vascular tumors that share a series of genetic and clinical characteristics but differ in their course, which is influenced by factors that remain to be identified.

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