

Segmental Neurofibromatosis versus Giant Nevus Spilus

Sir,

Segmental neurofibromatosis (NF-5) is characterized by café-au-lait (CAL) spots, and/or cutaneous neurofibromas limited to a circumscribed body segment (1). However, in the absence of clear diagnostic criteria for NF-5 it is difficult to establish the diagnosis in many cases. The recent reports by Selvaag et al. (2) and Menni et al. (3) in this journal demonstrate the difficulty in diagnosis of NF-5. No unique histological feature defines a lesion as a CAL spot (3). Some immunohistochemical (4) and ultrastructural (5) features may provide a clue for CAL but are not diagnostic.

The diagnosis of NF-5 was occasionally applied to patients with giant nevus spilus (6), despite insufficient clinical and histological evidence (7). Likewise, in Selvaag's case, the lentiginous hyperplasia and the presence of nevus cell nests are characteristic features of speckled lentiginous nevus (8) and are not diagnostic of CAL. Therefore, this case may represent unilateral lentiginosis rather than NF-5.

NF-5 is regarded as a mosaic form of other types of neurofibromatoses (mainly NF-1). Such mosaicism may also involve the gonads (9), resulting in transmission of full-blown NF-1 to an offspring of a parent with NF-5 (10). Consequently, patients with NF-5 may not be reassured that there is no risk of NF-1 in their offspring. Considering this, we may better serve our patients by restricting the diagnosis of NF-5 to cases that comply with Riccardi's criteria. For the patient this may prevent unjustified stress and, possibly, unnecessary investigations.

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