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More on Speckled Lentiginous Nevus Syndrome

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Key Words

Speckled lentiginous nevus • Speckled lentiginous nevus syndrome • Nevus spilus • Mosaicism • Hyperhidrosis • Neurocutaneous syndrome

Speckled lentiginous nevus (SLN) syndrome is a condition characterized by a segmental papular SLN associated with ipsilateral neurological or musculoskeletal disturbances. First reported retrospectively in 2002 by Happle [1] and Vidaurre-de la Cruz [2] on the basis of a handful of cases published for other reasons [3–8], only further 2 affected individuals have been reported ever since [9, 10].

I wish to propose herein an additional striking case of SLN syndrome, published in the literature >30 years ago but remained unrecognized so far. Heine and Poppele [11] described a 14-year-old boy who presented since the age of 2 a checkerboard-arranged papular SLN, mainly involving the upper trunk and right arm, and eventually developed homolateral hyperhidrosis, dysesthesia and musculoskeletal abnormalities (table 1).

Table 1. Main features of speckled lentiginous nevus syndrome cases described so far

R	A	S	Side	Speckled lentiginous nevus		Associated manifestations	
				A'	location	A''	other
3	27	M	right	0	chest, arm	NS	pectoral muscle hypertrophy
							gingival hyperplasia, focal epilepsy
4, 5	15	M	left	6	abdomen, flank	NS	leg sensorimotor neuropathy
6	43	F	right	0	leg	14	popliteal nerve palsy and thinning
					blue nevi		right bundle branch block
7	41	F	left	C	trunk, leg	NS	lower limb asymmetry, L5 vertebra malformation
							melanoma (right chest)
8	30	M	right	0	face, neck, trunk, limbs	25	upper limb spinal muscular atrophy
					blue nevi, Ota nevus-like changes, alopecia		
9	42	M	left	0	abdomen	C	hyperhidrosis, dysesthesia, hyperalgesia
							dyslexia
10	9	M	right	NS	arm, trunk, extending to the contralateral arm	NS	asymmetry of arms with muscular atrophy, scoliosis
							developmental delay, obsessive-compulsive disorder, Tourette's syndrome
11	14	M	right	2.5	arm, chest, extending to the contralateral arm	2.5	hyperhidrosis, dysesthesia, asymmetry of arms with muscular atrophy, scoliosis, extrapyramidal symptoms

All patients were white. R = Reference; A = age at diagnosis (years); S = sex; A' = age at onset of speckled lentiginous nevus; A'' = age at onset of extracutaneous symptoms; M = male; F = female; C = childhood; NS = not specified.

Moreover, I retrieved other cases featuring a lateralized SLN and a significant ipsilateral abnormality. Cabrera et al. [12] reported an 11-year-old boy with developmental delay, mental retardation, dyskinesias, ear malformations and hypertonia of the left leg, who presented pigmented macules over the left abdomen and limbs, among which an oval-shaped SLN with terminal hairs on the left thigh was described. Garrido-Arredondo et al. [13] documented a white patient affected by a large hairy SLN on the left supraclavicular/deltoideal region and idiopathic scoliosis. However, the lack of important data as well as some unfitting features prevent one from labeling the overmentioned patients with sine dubio SLN syndrome.

The impression is corroborated that SLN syndrome may be rather various as of clinical presentation and severity, being characterized not only by peripheral nerve and neuromuscular disturbances, but also by skeletal and central nervous system anomalies (table 1). Moreover, beside hyperhidrosis and dysesthesia (also present in phacomatosis pigmentokeratotic in which SLN syndrome is associated with Schimmelpenning syndrome), other ipsilateral skin manifestations, such as blue nevi and hair growth abnormalities, may represent significant findings as well as a further clue to the understanding of papular SLN-associated syndromes.

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