Short Communication

Neurofibromatosis-1 and zosteriform nevus spilus: A very rare clinical coexistence

Sir, neurofibromatosis-1 (NF-1), or von Recklinghausen's disease is an autosomal dominantly inherited genodermatosis that presents with multisystem involvement predominantly the skin, eyes, brain and bone. About half of the cases occur due to sporadic mutations. NF-1 is associated with several pigmented cutaneous anomalies like café-au-lait macules, axillary freckling, malignant melanoma and other melanocytic nevi.[1] The speckled and lentiginous nevus, also referred to as nevus spilus, is an uncommon congenital melanocytic disease that has a café-au-lait like hyperpigmented background, on which dark brown nevi are seen. We describe a patient of NF-1 with zosteriform nevus spilus. To the best of our knowledge, this rare coexistence has not yet been reported in literature.

A 40-year male patient presented with gradually increasing, multiple, discrete, painless nodules distributed throughout the whole body since birth. Buttonholing phenomenon was elicited on digital pressure in these nodules. The patient had multiple café-au-lait macules of variable size over the trunk and axillary freckling was present. Lisch nodules were present on ophthalmologic examination. The patient had no neurologic or musculoskeletal complaints. Systemic examination was unremarkable. Family history of similar lesions was not present. The patient was also found to have a tan coloured macule in a zosteriform distribution strictly on the right side of the lower abdomen extending upto the lower back upto the midline, which was studded with darkly pigmented lentigo-like macular lesions [**Figure 1** (**a**), (**b**)] Skin biopsy from the tan brown part of the zosteriform lesion showed elongation of the rete ridges and increased melanocytes lying in focal contiguity with one other near the tips of the rete ridges. Melanophages were found in dermis and melanin was increased in the basal layer. The biopsy from the dark specks showed junctional nests of nevus cells at the tips of rete ridges. Based on clinical and histopathological grounds, a diagnosis of neurofibromatosis-1 co-existing with zosteriform nevus spilus was made. The patient was counseled regarding the complications of his disease and regular follow up was advised.

NF-1 was first described by Friedrich von Recklinghausen in 1882. It is an inherited neuroectodermal disease that occurs due to mutations in a tumour suppressor gene located on chromosome 17 that encodes a protein called neurofibromin. Features include cutaneous and plexiform neurofibromas, tan-brown macular lesions called café-au-lait macules, pathognomonic axillary freckling, papillomatous tumors of the oral mucosa, pigmented iris hamartomas called Lisch nodules, bony abnormalities like sphenoidal dysplasia, pseudoarthrosis, thinning of long bone cortices, central nervous system abnormalities like optic nerve glioma, astrocytoma and schwannoma and endocrine disturbances. The diagnosis is chiefly clinical and is made as per the National institute of Health Consensus Development Conference, 1988 criteria. NF-1 has been associated with a number of nevi including congenital melanocytic nevi,[2] nevus anemicus[3] and Becker's nevus.[1]

Nevus spilus or speckled lentiginous nevus is an uncommon type of congenital melanocytic nevus that presents with a hyperpigmented macule initially followed by formation of the

more obvious and characteristic darkly pigmented specks later in life.[4] Lesions can be distributed in a circumscribed, block-like or linear fashion along the lines of Blaschko or in a zosteriform distribution.[4] It is distinguished from zosteriform lentiginous nevus by the macular hyperpigmentation in the background of the former. Histopathology from the background macule resembles that of lentigo simplex and the darker areas show features of compound nevi. There have been reports of malignant melanoma developing in speckled lentiginous nevi and careful follow up is advised.

The association of speckled lentiginous nevus with NF-1 has probably never been reported in literature except for a case report where bilateral segmental neurofibromatosis was found in a patient along with speckled lentiginous nevus.[5] The fact that this nevus was zosteriform in distribution further adds rarity to our case. Whether this finding was coincidental or there is an association needs further validation.

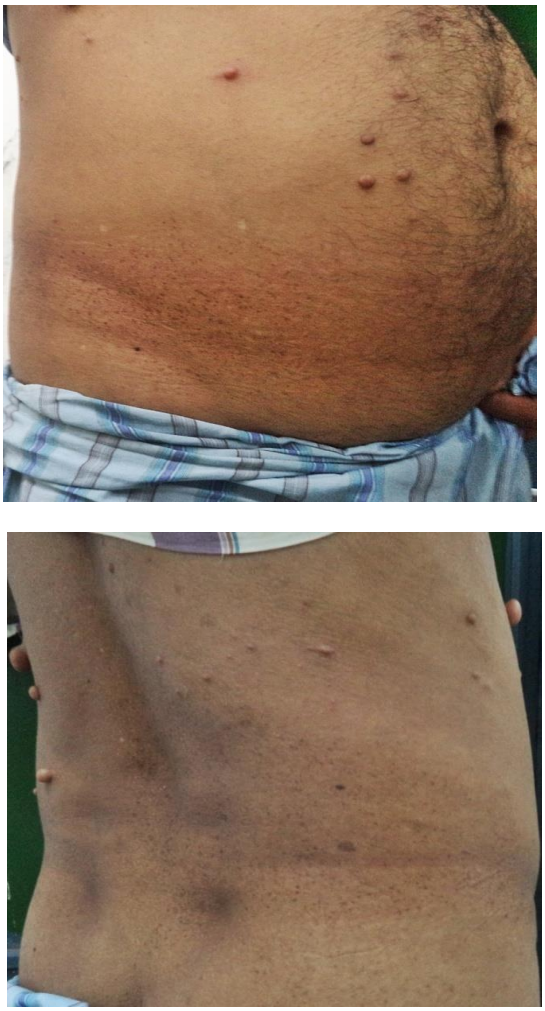

Figure 1 A strictly unilateral light brownish macular pigmentation in zosteriform distribution present on the right side of lower abdomen and back. Over this macular background, there are multiple darker brown nevi present. Multiple, discrete neurofibromas can also be seen. a) Anterior view b) Posterior view.

References

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