

A Case of Plexiform Neurofibroma Developed under the Overlying Speckled Lentiginous Nevus

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We report a case of plexiform neurofibroma developed under the overlying speckled lentiginous nevus, which occurred in a 20 year-old man. In this patient and his family no other signs of von Recklinghausen's disease were found. Discussion is focussed on the fact that both plexiform neurofibroma and speckled lentiginous nevus, which represent a defect in the neural crest, occurred in the same area of the skin. (*Ann Dermatol* 5:(2) 109-112, 1993)

Key Words: Plexiform neurofibroma, Speckled lentiginous nevus

A neurofibroma is a benign tumor of the nerve sheath origin¹⁻³. Neurofibroma may occur as either a solitary or plexiform lesion. Plexiform neurofibroma is regarded by some as pathognomic of von Recklinghausen's disease⁴⁻⁹.

Speckled lentiginous nevus is circumscribed tan pigmentation in which more darkly pigmented, raised or speckled nevomelanocytic elements are present¹⁰⁻¹⁴.

Both plexiform neurofibroma and speckled lentiginous nevus represent a certain defect in the neural crest¹⁵⁻¹⁷.

The purpose of this report is to document a case of plexiform neurofibroma in a patient without other signs of neurofibromatosis occurring with speckled lentiginous nevus on the same site. To the best of our knowledge, no case of plexiform neurofibroma occurring with speckled lentiginous nevus has been reported.

REPORT OF A CASE

A 20 year-old man visited our clinic with an asymptomatic, ill defined, soft shiny papulonodules and speckled black macules within the brownish

patch on the abdomen present since birth (Fig. 1). The papulonodules were 0.5cm in diameter, the brownish patch was 15cm in diameter and the speckled black macules were 0.1-0.5cm in diameter.

No history of trauma, insect bites or contact dermatitis in this site could be obtained. The family history of neurofibromatosis was negative. Routine physical, neurological and laboratory examinations gave no abnormalities. No skeletal, auditory or ocular abnormalities were found.

An excision biopsy was performed. On microscopic examination there were numerous large nerve fascicles with irregular configurations embedded in cellular matrices (Fig. 2). The matrices were similar to those seen in typical neurofibroma. The tumor was well circumscribed and composed of spindle or S-shaped cells (Fig. 3). On alcian blue stain, areas of mucoid transformation demonstrated within the fascicles. The fascicular structure showed a positive S-100 protein immunoreactivity (Fig. 4). The mucoid areas and the perineurium were negative S-100 protein immunoreactivity. Histopathologic examination of the brownish pigmented patch showed slightly elongated rete ridges and increased melanin pigment in the basal layer (Fig. 5). Skin sections from the speckled black macules showed nevus cell nests in the lower epidermis and melanophages in the dermis (Fig. 6).

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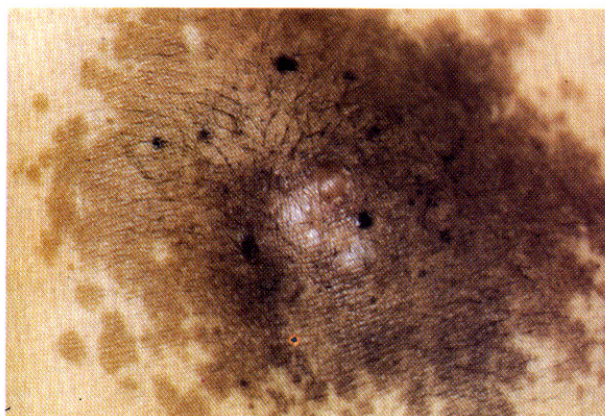


Fig. 1. The brownish patch with small darker colored macules and subcutaneous nodules on the abdomen.

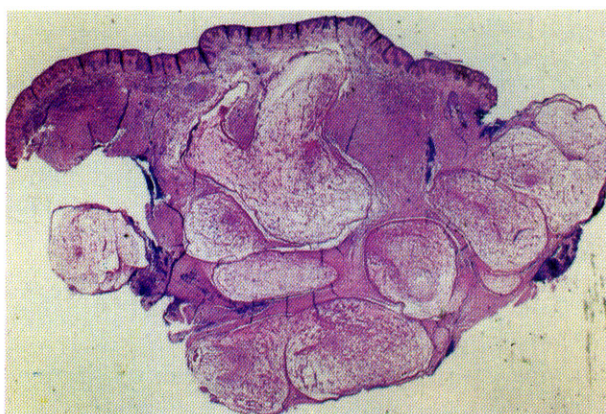


Fig. 2. Skin section from the subcutaneous nodule shows numerous expanded nerve fascicles embedded in the loose cellular matrices (H&E stain, $\times 10$).

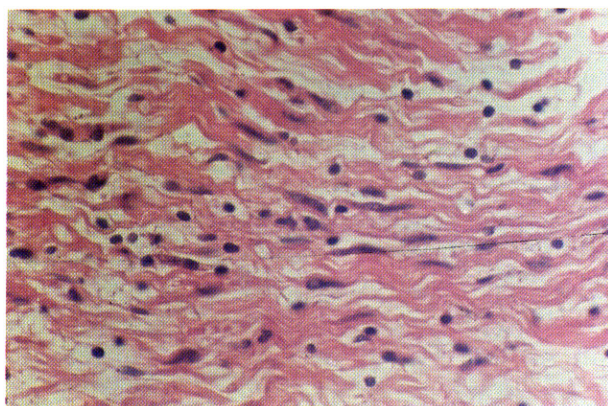


Fig. 3. A high power view of Fig. 2 shows that the tumor is composed of spindle cells (H&E stain, $\times 400$).

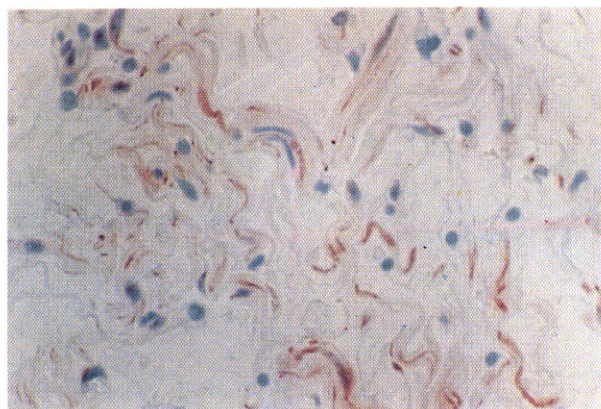


Fig. 4. Numerous cells of the fascicles present intensive positivity for S-100 protein (Anti-S-100 ABC, $\times 400$).

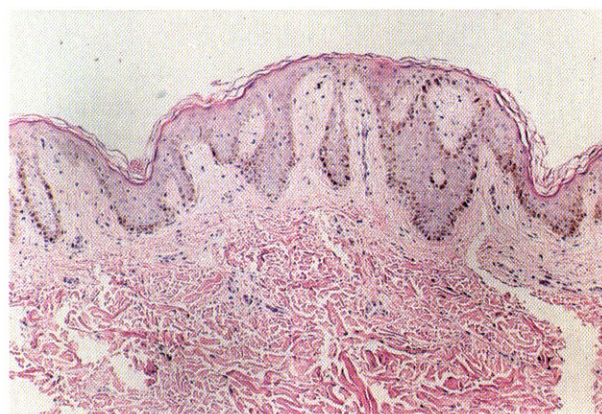


Fig. 5. Skin section from the brownish patch shows elongation of rete ridges and increase in the amount of melanin in the basal cell layer (H&E stain, $\times 100$).

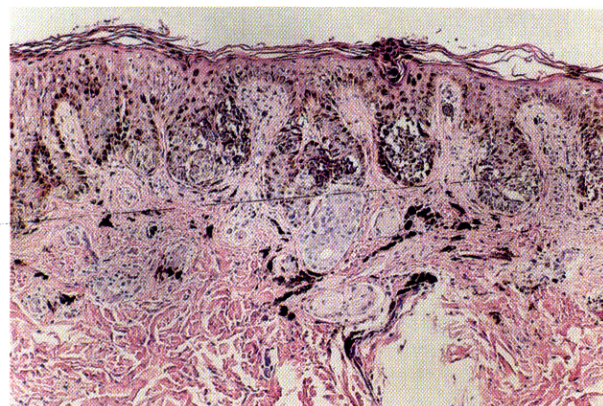


Fig. 6. Skin section from the speckled areas shows nevus cell nests in the lower epidermis and melanophages in the dermis (H&E stain, $\times 100$).

DISCUSSION

A neurofibroma is a benign tumor of nerve sheath cells, endoneurium and perineurium with little or no proliferation of axons¹⁻³. Neurofibroma is either solitary or multiple. When one or two lesions are present, no internal manifestations are combined. But when three or more lesions are present, the diagnosis of neurofibromatosis is made¹⁸. The usual patterns of neurofibroma are the common neurofibroma and plexiform neurofibroma. Several other types exist as the pacinian neurofibroma, pigmented neurofibroma, storiform neurofibroma, and myxoid neurofibroma.⁶

In plexiform neurofibroma, numerous tortuous, thickened nerves can be felt and the tumor involves nerve trunks of subcutaneous tissue or visceral organs. It is regarded by some as pathognomic of von Recklinghausen's disease^{4,9}. In our patient and his family no other signs of von Recklinghausen's disease were found. In neurofibromatosis, the age of onset is variable. In some instance the disease has first appeared in adult life¹. Based on this follow up is necessarily required in our case.

Histological diagnosis of plexiform neurofibroma is based on the recognition of mass formed by expanded and distorted nerve fascicles. Numerous large nerve fascicles showing areas of mucoid transformation are embedded in cellular matrices. The nerve fascicles are composed of spindle or S-shaped tumor cells^{19,20}. Our case histologically showed typical features of a plexiform neurofibroma.

The histopathologic differential diagnosis of plexiform neurofibroma includes true neuroma⁶. Both tumors are composed of nerve fascicles surrounded by a well developed perineurium. The key feature of the former are well developed nerve fascicles that have an axon to Schwann-cell ratio of at least 1:1, but the key feature of the latter are well developed nerve fascicles that have an axon to Schwann-cell ratio of much less than 1:1.

The pathogenesis of neurofibroma remains obscure. Cellular elements derived from neural crest, i.e., Schwann cells, melanocytes, and possibly endoneurial fibroblasts, the natural components of skin and nerves multiply excessively in multiple

foci, and the melanocytes function abnormally, but the time when this begins and the mechanism of it are unclear¹⁷. The presence of S-100 protein on immunohistochemical study of our case is evidence for the presence of neural crest origin.

Speckled lentiginous nevus is a pigmented, light brown or tan macule of varied diameter, speckled with smaller, darker colored macules or papules. It may present at birth or during childhood and frequently occurs on the trunk and lower extremities. Histologically, the light brown patch shows the histologic features of lentigo simplex. The speckled areas show nevus cell nest in the lower epidermis and melanophages in the dermis^{21,22}. The lesions of our patient showed typical clinical and histopathologic features of speckled lentiginous nevus.

Speckled lentiginous nevus represents a localized defect in neural crest melanoblast¹⁶.

Both speckled lentiginous nevus and neurofibromatosis had neural crest origin but the relationship between speckled lentiginous nevus and neurofibromatosis is not being resolved²³. Speckled lentiginous nevus can be observed in patients with multiple neurofibroma²³. In another report, a daughter had neurofibromatosis and her mother had a speckled lentiginous nevus²³. These observations suggest that both speckled lentiginous nevus and neurofibromatosis may be closely related to each other²³.

Our case, where the two distinct diseases are at the same site of the skin, strongly supports that the two diseases may be closely related.

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