A Case of Isolated Plexiform Neurofibroma

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Plexiform neurofibroma is considered to be pathognomic of neurofibromatosis type 1 (NF1). Herein we report a solitary plexiform neurofibroma which is not associated with NF1.

A 61-year-old man presented with asymptomatic skin colored nodules on the medial side of his left great toe. No other abnormalities were found in his personal or family history. Clinically, the tumor was simulating the appearance of mucous cysts. Microscopically, it was a plexiform neurofibroma located in the dermis which seemed to originate from small superficial nerves. This case would seem to confirm that the superficial form of plexiform neurofibroma involving small nerves in the dermis or subcutis is not necessarily pathognomic for NF1.

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Key Words : Dermis, Nodule, Superficial form

Plexiform neurofibroma involve deeper and larger nerve trunks, and may extend to involve visceral organs. In this tumor, the nerve is converted into a convoluted mass which has been likened to a 'bag of worms'. The plexiform neurofibroma is considered to be pathognomic of NF1[1,2,3].

However, cases of isolated plexiform neurofibroma have been reported intermittently, which were not associated with NF1[4,5]. In our review, isolated plexiform neurofibromas have not been described in the Korean literature. We report a rare case of isolated plexiform neurofibroma in which the patient and his family had no other evidence of neurofibromatosis.

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CASE REPORT

A 61-year-old man presented with asymptomatic skin colored nodules on the medial side of his left great toe (Fig. 1). Incidentally he noticed the lesion two years ago. Examination disclosed two adjacent 0.3 × 0.3 cm sized rubbery transparent nodules. The medical evaluation of this patient disclosed no evidence of other skin tumors or pigmentary lesions. Family history was negative for similar skin diseases or other hereditary conditions. Initially the lesion was considered as mucous cysts. The lesion was completely excised. Excised specimen of the adjacent two nodules revealed a continuous mass of whitish-gray tangled, ropelike structures like 'bag of worms'.

Microscopic examination disclosed expanded nerve fascicles cut in various planes of sections (Fig. 2). The tumor was well circumscribed and surrounded by concentric bands of collagen displaying 'onion skin pattern'. It was composed of faintly eosinophilic, thin, collagen bundles (Fig. 3), myxoid changes and occasional blood vessels (Fig. 4). The nuclei of the cells were spindle- or S-shaped and quite uniform in size. A fairly large amount of tumor cells showed a positive S-100 protein immunoreactivity. A diagnosis of isolated

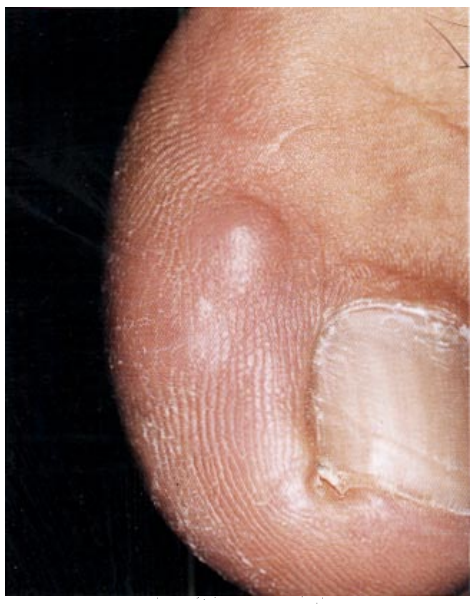

Fig. 1. Two adjacent 0.3×0.3 cm sized rubbery transparent nodules on the medial side of the great toe.

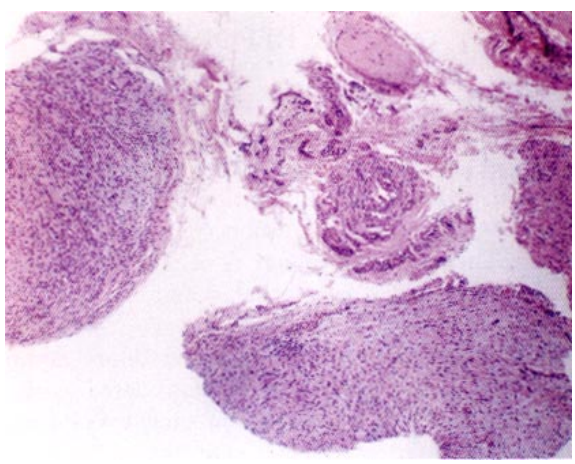

Fig. 2. Well circumscribed nerve fascicles cut in various planes of sections (H&E stain, ×40).

plexiform neurofibroma was made.

DISCUSSION

Plexiform neurofibroma had been reported to be found only in patients with von Recklinghausen's disease and considered to be pathognomic of NF1[1,2,3]. The National Institutes of Health Consensus Development Conference Statement has designated the presence of a plexiform neurofibroma

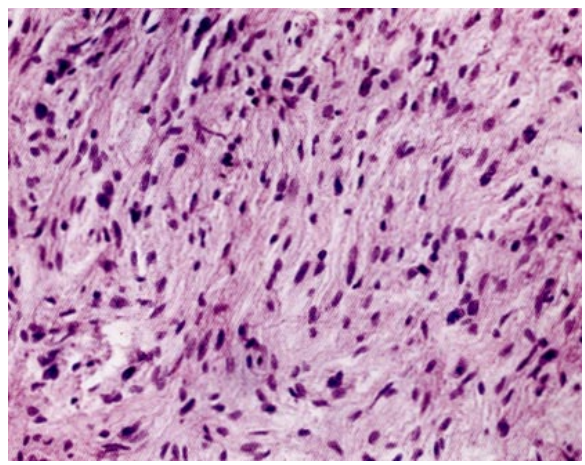

Fig. 3. The tumor was composed of faintly eosinophilic, thin, collagen bundles. The nuclei of the cells were spindle or S-shaped and quite uniform in size (H&E stain, ×400).

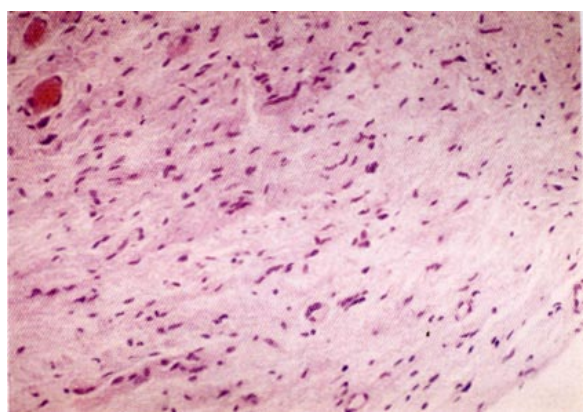

Fig. 4. Spindle shaped cells with wavy collagen fibrils in myxoid stroma (H&E stain, ×100).

or two neurofibromas as one of the two necessary findings for diagnosing generalized neurofibromatosis[6]. Typically, plexiform neurofibroma involves large nerve trunks of subcutaneous tissue or visceral organs and they also infiltrate the surrounding soft tissue. It can reach enormous size and be associated with massive soft tissue overgrowth leading to significant functional impairment. Clinically, the tumor is described as large nodular or diffuse types[1]. Histologically, numerous large nerve fascicles with irregular configurations are embedded in cellular matrices containing varying amounts of cells with spindle- and S-shaped nuclei, wavy collagen fibrils, mucin, and

mast cells[7].

Plexiform neurofibromas has to be differentiated from plexiform schwannoma[10], intraneural plexiform neuroma[11] and plexiform fibrohistiocytic tumor[12]. Plexiform schwannoma is intraneural and composed predominantly of Antoni A tissue. Plexiform schwannoma does not show axons in the tumor mass but occasionally in the capsule. A mild to moderate degree of cytologic pleomorphism is present, and mitotic figures are only rarely encountered in the plexiform schwannoma. Intraneural plexiform neuroma is intraneural tumor composed of nodules and broad cords. Intraneurally broad fascicles of axons and Schwann cells are interlaced in a scanty, clear matrix. Plexiform fibrohistiocytic tumor shows multinodular or plexiform proliferation of histiocyte-like and fibroblast-like cells associated with multinuclear giant cells and the tumor cells do not stain for S-100 protein.

Recently two cases of isolated plexiform neurofibromas were observed in otherwise healthy patients and these patients had no family history of neurofibromatosis or other anomalies. In the first case, a solitary plexiform neurofibroma was described which was presented as a linear movable cord of 0.5×10 cm size on the abdomen and involved a large nerve trunk in subcutaneous tissue[4]. Recently a superficial form of isolated plexiform neurofibroma was reported[5]. The tumor was observed as a 1.8 cm nodule on the thigh and microscopically, it was located superficially in the dermis involving small nerves. These cases suggest that presence of a plexiform neurofibroma does not always indicate the associated neurofibromatosis[4,5] or structural anomalies.

In our case, the tumor was presented as small rubbery transparent nodules on the great toe and it was located superficially in the dermis involving a small superficial nerve. The patient had no personal and family history of other abnormalities suggestive of neurofibromatosis. This case is similar to the recently described case[5] in that the isolated plexiform neurofibroma occurred as a superficial form involving small nerves in the dermis and was presented clinically as two small adjacent nodules. But the size of the nodule was much smaller in our case.

In Korean literature, a few cases of plexiform neurofibroma have been reported. They were described to have other anomalies of neurofibromatosis including multiple skeletal deformities, and giant pigmented lesions[8,9].

We described an isolated plexiform neurofibroma which was developed in a healthy man with no stigmata of von Recklinghausen's neurofibromatosis and involved small nerve fibers in the dermis. This case would seem to confirm the concept that the occurrence of superficial form of plexiform neurofibroma involving small nerves in the dermis or subcutis is not necessarily pathognomic for von Recklinghausen's disease, as in the previous cases of Aloi[4] and Fisher[5].

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