A Case of Multiple Plexiform Schwannomas

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Plexiform schwannoma is a relatively rare, benign peripheral nerve sheath tumor that can be located either in the deep soft tissues or in the dermis or subcutaneous tissue. This tumor may occur singly or as multiple lesions and may be localized to one anatomic site or diffusely distributed. Plexiform schwannoma should be differentiated with plexiform neurofibroma or other plexiform malignant tumors. We describe a case of a 6-year-old patient with multiple cutaneous plexiform schwannomas who had no other stigmata of neurofibromatosis 1 or family history suggesting a genetic disorder. The histopathological study revealed a tumor composed of multiple intradermal or subcutaneous interlacing and interconnecting fascicles and nodules that vary in size and shape. Characteristic Antoni A type cellular tissue showing frequent nuclear palisading and Verocay bodies were observed within well circumscribed elongated nodules. (Ann Dermatol 12(2) 130~133, 2000).

Key Words : Multiple cutaneous plexiform schwannomas

Plexiform schwannoma(PS) is a relatively rare, benign peripheral nerve sheath tumor, which usually arises in either the dermis or subcutaneous tissue[1,2]. It affects predominantly young adults and occurs most commonly in a slowly growing asymptomatic solitary nodule in the head and neck region, on the trunk, and in the upper extremities[1,3,4]. This tumor may occur singly or as multiple lesions and may be localized to one anatomic site or diffusely distributed[4-8]. The growth pattern of PS is similar to that of plexiform neurofibroma, but PS does not develop into a malignant peripheral nerve sheath tumor. We herein report a case of multiple plexiform schwannomas which occurred in a 6-year-old girl.

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CASE REPORT

A 4-year-old girl was presented to our clinic in April 1996 for the evaluation of several skin nodules which had been on the trunk for 1 year. The skin lesions showed four scattered, relatively firm and slightly tender erythematous, from pea to bean sized nodules on the chest, the abdomen and the right upper arm. The family and past medical histories were not significant. A physical examination failed to reveal any cafe-au-lait spot, Lisch nodule, or axillary freckling. The patient revisited in August 1998 because of the skin lesions gradually increasing in number and size. Examination revealed three discrete 2.0×2.0, 1.8×2.0, 1.5×1.5cm sized skin nodules on the trunk and 1.0×1.0cm sized skin nodules on the arm, all of which were previously existed(Fig. 1). There were two newly detected 0.5×0.5cm sized flesh colored nodules on the back. Brain computed tomography and ophthalmic examination showed no significant abnormality. A biopsy specimen showed encapsulated, multinodules of different sizes and shapes in the dermis. Each individual nodule was surrounded by a thin fibrous capsule and was composed of a highly cellular proliferation of closely packed spindle cells with

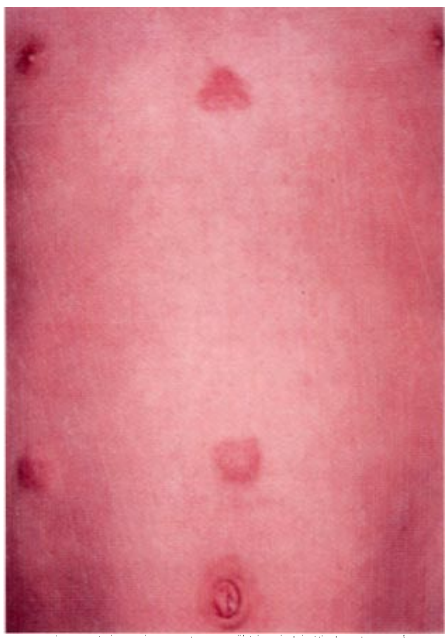

Fig. 1. Three discrete erythematous 2.0 2.0, 1.8 2.0, 1.5 1.5cm sized sessile nodules on the chest and abdomen.

elongated or slightly buckled nuclei and eosinophilic cytoplasm. Nuclear palisading and occasional Verocay bodies were present(Fig. 2). Neither necrosis nor hemorrhage was observed. Immunohistochemically, virtually all of the cells within and between the nodules were strongly positive for S100 protein(Fig. 3). However, the cells related to the tumor capsule were S100 stain negative. No axons within the tumor could be stained with Bodian stain. All these lesions were diagnosed as plexiform schwannomas. No specific treatment was given to our patient and regular follow up was recommended.

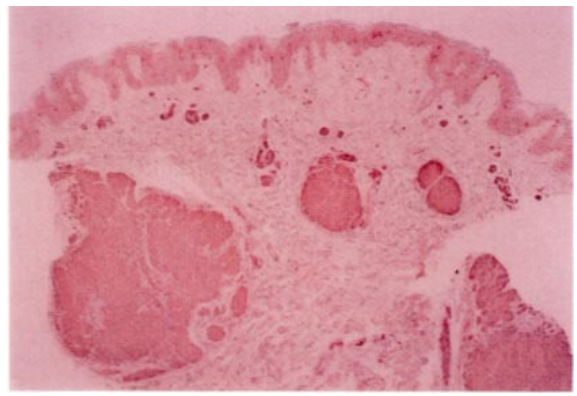

Fig. 3. Intense positivity for S100 protein in all tumoral nodules. In contrast, the cells of the fibrous capsule were S100 protein negative (× 40).

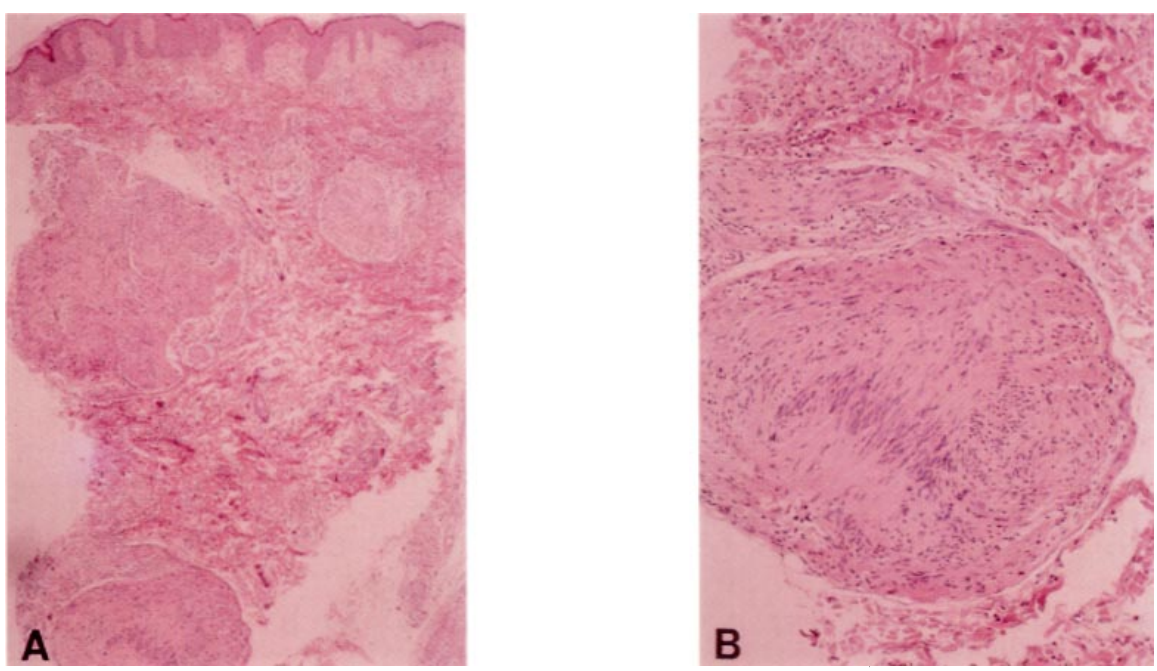

Fig. 2. A. Multiple neoplastic cell nodules and fascicles widely spaced in the dermis, with normal epidermis (H&E, × 40). B. The tumor nodule consists of Antoni A type tissue showing cells with elongated nuclei, nuclear palisades, and Verocay bodies (H&E, × 100).

DISCUSSION

Schwannoma is a benign tumor of the nervous system originating form the neural sheath. Several variants of schwannoma have been described including degenerated, cellular, glandular and neuroblastoma-like schwannoma. The term plexiform schwannoma was first introduced by Harkin et al.[9] in 1978 to describe a benign peripheral nerve sheath tumor composed exclusively of schwann cells arranged in a plexiform pattern. So far three cases of PS[10,11] have been reported in Korean literature since then.

PS usually occurs as a slowly growing asymptomatic nodule, rarely tender or painful, that has been present for months or years[1]. The tumors reported ranged from 0.5 to 15cm in diameter with an average size of 3cm[12]. It occurs most frequently in early adulthood, but may appear at any age[13]. There is no sex predominance. The lesion which the tumor arises most often are in the head and neck, on the trunk, or in the upper extremities[1,3,4]. It is rarely located in the upper lip[14], buccal mucosae[14], tongue[3,14], vulva[15], or lower extremities[7]. Histologically, PS has shown rather typical features of conventional schwannoma other than its plexiform growth pattern. The nodules of PS are composed predominantly of Antoni A type tissue showing frequent nuclear palisading and Verocay bodies[4,15]. The nodules are hypercellular, and commonly show cytologic pleomorphism. No axons can be identified within the nodules. The cells of the tumor capsule are epithelial membrane antigen-positive, as are the cells of the perineurium. A focal increase of cellularity may be found in cases of the malignant transformation of plexiform neurofibroma, and these cases must be distinguished from that of PS. Distinction from plexiform neurofibroma is important, because plexiform neurofibroma is virtually pathognomonic of neurofibromatosis 1(NF1) and carries a significant risk of malignant transformation. The presence of Verocay bodies, and the absence of mitotic figures and axons within the nodules are important findings for the differential diagnosis of these tumors[1,3,15].

Judging from the literature, most plexiform schwannomas are not associated with NF1 and NF2, especially if they are solitary[1,3,16,17]. However, some solitary and especially multiple plexiform schwannomas, are associated with NF2[16,17]. Schwannomatosis or neruilemmomatosis, initially described in Japanese literature[7], is a disease including multiple cutaneous schwannomas, and central nervous system tumors without acoustic tumors or other signs of NF1 or NF2. There appear to be a group of patients that develop multiple cutaneous or noncutaneous schwannomas who do not develop intracranial lesions, and a second group of patients with multiple cutaneous or noncutaneous schwannomas who develop intracranial lesions and, in particular, bilateral acoustic neuromas. It is this second group of patients who may or may not prove to have a subtype of NF2. In 1995 Honda, et al.[18], using DNA markers for different regions of chromosome 22, identified a mutated NF2 gene in the tumor tissue and peripheral leukocyte with in three of seven patients with multiple schwannomas. This indicated that germline mutations in the NF2 gene were the molecular mechanism of schwannomatosis. Therefore, it is evident that schwannomatosis and NF2 overlap each other clinically and pathologically. On the other hand, the association between plexiform schwannoma and NF1 appears to be much more rare and actually questionable. Only one case of multiple plexiform schwannomas reported in Korean literature was associated with cafe au lait spots reminiscent of neurofibromatosis[11].

In our case, there is no evidence of cafe-au-lait spot or intracranial neoplasm. However in the case of childhood multiple schwannomas, the association with NF2 is very difficult to be judged because the appearance of peripheral schwannoma may precede that of vestibular schwannoma[19]. When present, a positive family history is very helpful as in the case, reported by Sasaki[13], of a 5-year-old child with multiple skin schwannomas whose father had bilateral vestibular schwannomas.

Excision is the treatment of choice for schwannoma. Plexiform schwannomas may recur, but this probably reflects incomplete excision due to their multifocal nature rather than true recurrence[20].

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