

Wagner-Meissner neurilemmoma of the lip occurring in a patient with neurofibromatosis type 1: A case report

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Abstract. Wagner-Meissner corpuscles are specialized mechanoreceptors located in the dermal papillae. Wagner-Meissner corpuscle-like structures are occasionally a component of certain types of tumors, such as melanocytic nevus and neurofibroma. Benign tumorous lesion entirely composed of Wagner-Meissner corpuscles are described as Wagner-Meissner neurilemmoma, and only four such cases have been reported. Here, we report the first case of Wagner-Meissner neurilemmoma in a patient with neurofibromatosis type 1. A 16-year-old Japanese male with neurofibromatosis type 1 presented with a tumorous lesion on the upper lip. Resection of the tumor was performed under a clinical diagnosis of neurofibroma. Histopathological examination revealed an unencapsulated, poorly-circumscribed tumor, comprised of abundant Wagner-Meissner corpuscle-like structures, which were composed of 5-15 lamellated Schwann cells containing eosinophilic cytoplasm and peripherally located nuclei. No spindle-shaped neoplastic cell proliferation, as seen in conventional neurofibroma, was observed. Accordingly, a diagnosis of Wagner-Meissner neurilemmoma was made. The pathogenesis of Wagner-Meissner neurilemmoma remains unclear. The hamartomatous or reactive proliferative nature has been proposed. In addition, this lesion may represent an extreme form of diffuse neurofibroma with abundant Wagner-Meissner corpuscles associated with neurofibromatosis type 1, even though the previously reported four patients did not have neurofibromatosis. Therefore, further studies are needed to clarify the pathogenesis of this extremely rare tumor, including its association with neurofibromatosis.

Introduction

Wagner-Meissner corpuscles are specialized mechanoreceptors located in the dermal papillae that directly connect with the basal layer of the epidermis, and are prominent in the palms and soles (1). They show characteristic histological features: An encapsulated round to oval structure with central lamellation and peripherally located nuclei of Schwann cells (1). Wagner-Meissner corpuscles or Wagner-Meissner corpuscle-like structures (pseudo-Meissner corpuscles) are occasionally a component of some types of cutaneous and neurogenic tumors, including melanocytic nevus and neurofibroma (2,3). Benign tumorous lesions entirely composed of Wagner-Meissner corpuscles were first described by Kaiserling and Geerts (4). They named these lesions Wagner-Meissner neurilemmoma, and to date, only four such cases have been reported in the English literature (4-6).

Neurofibromatosis type 1 is a relatively common autosomal dominant disorder, characterized clinically by presence of café-au-lait spots (7). It is well recognized that various types of tumors, including nervous and non-nervous systems, develop in patients with neurofibromatosis type 1. Multiple cutaneous neurofibromas are the most frequent tumor in patients with neurofibromatosis type 1, and patients with this disorder have a risk of development of malignant peripheral nerve sheath tumor (7). However, to the best of our knowledge, occurrence of Wagner-Meissner neurilemmoma in patients with neurofibromatosis type 1 has not been described. Here, we report the first case of this lesion in a patient with neurofibromatosis type 1 and discuss the clinicopathological features.

Case report

A 16-year-old Japanese male with neurofibromatosis type 1 presented with a tumorous lesion on the upper lip. He had multiple café-au-lait spots in the entire body and ephelides in the face. Moreover, he had undergone surgical resection of the congenital melanocytic nevi of the back and thigh. Resection of the lip tumor was performed under a clinical diagnosis of neurofibroma. No recurrence has been observed during medical follow-up.

Formalin-fixed and paraffin-embedded specimens of the resected tumor were processed for routine histological examination and immunohistochemical analyses.

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Table I. Clinicopathological features of Wagner-Meissner neurilemmoma.

Case no.	Age (years)	Sex	Neurofibromatosis	Location	Capsulization	(Refs.)
1	80	Male	-	Thigh	Encapsulated	(4)
2	33	Male	-	Leg	Encapsulated	(4)
3	24	Female	-	Vulva	Encapsulated	(5)
4	10	Male	-	Cheek	Unencapsulated	(6)
Present case	16	Male	Type 1	Upper lip	Unencapsulated	

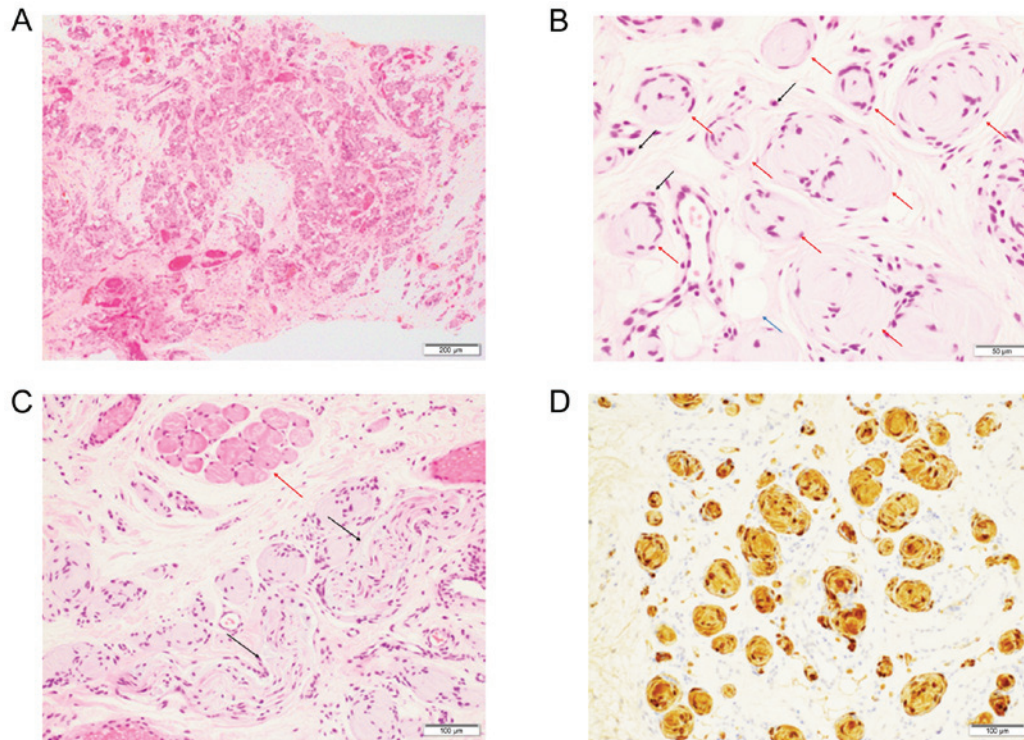


Figure 1. Histopathological and immunohistochemical findings of the upper lip tumor. (A) Unencapsulated poorly-circumscribed tumor composed of abundant Wagner-Meissner corpuscle-like structures (H&E, x100). (B) Wagner-Meissner corpuscle-like structures are composed of 5-15 lamellated Schwann cells containing eosinophilic cytoplasm and peripherally located nuclei (red arrows). A few mast cells are observed (black arrows). Moreover, a few fatty cells are also present within the lesion (blue arrow) (H&E, x400). (C) Peripheral nerve with myxoid changes is seen within the tumor (black arrows). Striated muscles are also present (red arrow) (H&E, x200). (D) Immunohistochemically, S-100 protein is diffusely expressed (x200).

In this report, immunohistochemical analysis was performed using an autostainer (Autostainer link 48; Dako Cytomation). The primary antibody used in this report was a rabbit polyclonal antibody against S-100 protein (Dako Cytomation).

Histopathological examination revealed an unencapsulated, poorly-circumscribed tumor located in the fatty tissue. The tumor was comprised of abundant Wagner-Meissner corpuscle-like structures, which were composed of 5-15 lamellated Schwann cells containing eosinophilic cytoplasm and peripherally located nuclei (Fig. 1A and B). These structures were packed in several portions; however, they were intermingled with fatty tissue and striated muscles in the periphery of the tumor (Fig. 1A). No mitotic figures were noted. Additionally, no spindle-shaped neoplastic cell proliferation, as seen in conventional neurofibroma, was observed (Fig. 1A and B). Peripheral nerves with myxoid changes (Fig. 1C) and a few mast cells were observed within

the tumor (Fig. 1B). The tumor extended to the margin of the resected specimen, however, no additional resection was not performed.

Immunohistochemical analysis clearly demonstrated that these corpuscles were diffusely positive for S-100 protein (Fig. 1D), but S-100 protein-positive spindle cells were absent (Fig. 1D).

Based on these features, a final diagnosis of Wagner-Meissner neurilemmoma was made.

Discussion

There has been no previous report of Wagner-Meissner neurilemmoma occurring in a patient with neurofibromatosis type 1. The clinicopathological features of the previously reported four cases of Wagner-Meissner neurilemmoma, in addition to our case, are summarized (Table I). Based on these reports, males are preferentially affected (female:male, 1:4),

and the tumor mainly occurred in adolescents or young adults, except for one patient (case 1). The predilection sites were the leg and oral mucosa.

The characteristic histopathological feature of the present tumor was the presence of abundant Wagner-Meissner corpuscle-like structures and a lack of neoplastic spindle cell nests, as seen in conventional neurofibroma (4-6). Therefore, we diagnosed this lesion as Wagner-Meissner neurilemmoma, although neurofibroma with abundant Wagner-Meissner corpuscles or pseudo-Meissner corpuscles must be differentiated. The unique feature of the present tumor was the poor circumscription and lack of encapsulation. The previously reported three cases (two cases occurring in the leg and one case in the vulva) were well-circumscribed and encapsulated (4,5); however, a case with a poorly circumscribed and unencapsulated tumor, percolating into the surrounding fatty tissue, resembling the present case, occurring in the cheek of a patient has been reported (6). The reason for this difference remains unclear; however, it might be a reflection of the location of the tumor (skin or mucosa). Further studies are needed to clarify the detailed clinicopathological features of this extremely rare tumor.

The pathogenesis of Wagner-Meissner neurilemmoma remains unclear. The hamartomatous nature of the tumor has been proposed to be related to its occurrence mainly in adolescents and young adults (6). Wagner-Meissner corpuscles are also present in the oral mucosa, where they are termed the end-bulbs of Krause (7). Moreover, pseudo-Meissner corpuscles are also found in cases of traumatic neuroma (4). Interestingly, myxoid changes in the peripheral nerves, suggestive of nerve degeneration, were observed within the present patient's tumor, and may be indicative of reactive proliferative changes. Moreover, our patient was also diagnosed with neurofibromatosis type 1. Thus, this lesion might represent an extreme form of diffuse neurofibroma with abundant Wagner-Meissner corpuscles associated with neurofibromatosis type 1, even though the previously reported four patients did not have neurofibromatosis (Table I). Further studies are needed to clarify the pathogenesis of this extremely rare tumor, including its association with neurofibromatosis.

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Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Authors' contributions

CM and MI undertook study conception and design. The acquisition and analysis of data was performed by CM, MI, YK, NM, KK, HO, and KT. CM and MI drafted the manuscript and prepared the figure. The final version of the manuscript has been read and approved by all authors.

Ethics approval and consent to participate

This study was performed according to Kansai Medical University Hospital's ethical policy.

Patient consent for publication

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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