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Multinodular/Plexiform Schwannoma of the Ankle: A Case Report and Literature Review

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Abstract

Background: Multinodular/plexiform schwannoma is an exceedingly rare benign peripheral nerve sheath tumor that often arises in the dermis or subcutaneous tissue, particularly of the trunk and head and neck region. The ankle is an uncommon location for this peculiar condition.

Case Report: A 50-year-old man without neurofibromatosis presented with a 10-year history of a slowly growing, occasionally painful mass in the medial aspect of the right ankle. Physical examination revealed a 5-cm, elastic-soft, poorly mobile, non-tender mass. Magnetic resonance imaging (MRI) showed multiple nodular lesions with intermediate signal intensity on T1-weighted sequences and heterogeneous high signal intensity on T2-weighted sequences. Contrast-enhanced MRI demonstrated strong and homogeneous enhancement. After core needle biopsy, the lesions were successfully treated by complete surgical enucleation. Histological examination confirmed the diagnosis of schwannoma consisting of predominantly Antoni A areas. The patient had no evidence of local recurrence and no aggravated neurological deficit at the latest follow-up.

Conclusion: This unique case provides valuable insights into the understanding and management of multinodular/plexiform schwannoma in the ankle.

Keywords: Multinodular/plexiform schwannoma, tibial nerve, ankle, MRI.

Introduction

Plexiform (multinodular) schwannoma is a unique variant of schwannoma first described in 1978 by Harkin *et al.* (1). It is characterized by a plexiform or multinodular growth pattern (2). Multinodular/plexiform schwannoma usually develops in the dermis or subcutaneous tissue (3, 4) and most frequently occurs in the trunk, head and neck (3-6). The duration of symptoms before management ranges from 1 month to 30 years, with a mean duration of 8 years (4).

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The lesion is usually less than 2 cm in greatest diameter (6). Surgical excision or enucleation is the treatment of choice for multinodular/plexiform schwannoma, although a conservative "wait and see" approach should be preferred for asymptomatic patients with stable lesions (7).

Multinodular/plexiform schwannoma involving major peripheral nerves is ultra-rare (7, 8). Herein, we describe an unusual case of multinodular/plexiform schwannoma arising from the posterior tibial nerve in a middle-aged man. We also provide a literature-based discussion of this peculiar condition. Written informed consent was obtained from the patient to publish this case report and accompanying images.

Case Report

A 50-year-old man presented with a 10-year history of a slowly growing, occasionally painful mass in the medial aspect of the right ankle. There was no history of antecedent trauma. Past medical history was unremarkable. The patient had no noteworthy family history. Physical examination revealed a 5-cm, elastic-soft, poorly mobile, non-tender mass (Figure 1). Tinel sign was negative. Neurological examination showed numbness in the lateral planter aspect of the right foot. Manual muscle testing of the right ankle and toe revealed no weakness. Laboratory data were within normal limits. Radiographs revealed soft-tissue swelling without calcification. Magnetic resonance imaging (MRI) demonstrated multiple nodular lesions with intermediate signal intensity on T1-weighted sequences (Figure 2A) and heterogeneous high signal intensity on T2-weighted sequences (Figure 2B). Contrast-enhanced fat-suppressed T1-weighted sequences showed strong and homogeneous enhancement. There was no evidence of bone involvement. Based on clinical and MRI findings, we highly suspected the possibility of multinodular/plexiform schwannoma.

Core needle biopsy was carried out, and the pathological diagnosis was schwannoma. The surgical enucleation was performed under general anesthesia with pneumatic tourniquet control and loupe magnification.

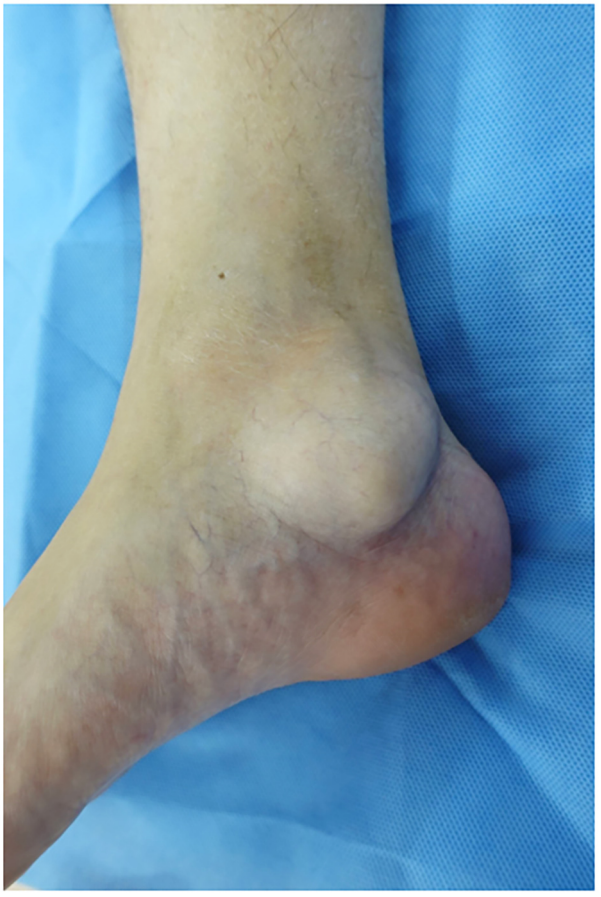

Figure 1. *Photograph displaying a soft-tissue mass in the medial aspect of the right ankle.*

The tumors were observed to cohesively follow the course of the posterior tibial nerve through the tarsal tunnel. Grossly, individual tumors were yellowish-white and ranged from 1.0 to 3.0 cm in greatest diameter (Figure 3). Microscopically, all tumors showed a proliferation of spindle-shaped cells arranged in fascicles with occasional nuclear palisade arrangement in Antoni A areas (Figure 4A). Loosely arranged reticular portions (Antoni B areas) were also focally seen (Figure 4B). Nuclear atypia and mitotic figures were absent. These findings confirmed the diagnosis of multinodular/plexiform schwannoma.

The postoperative course was uneventful. The numbness was relieved immediately after surgery. There was no evidence of local recurrence and no aggravated

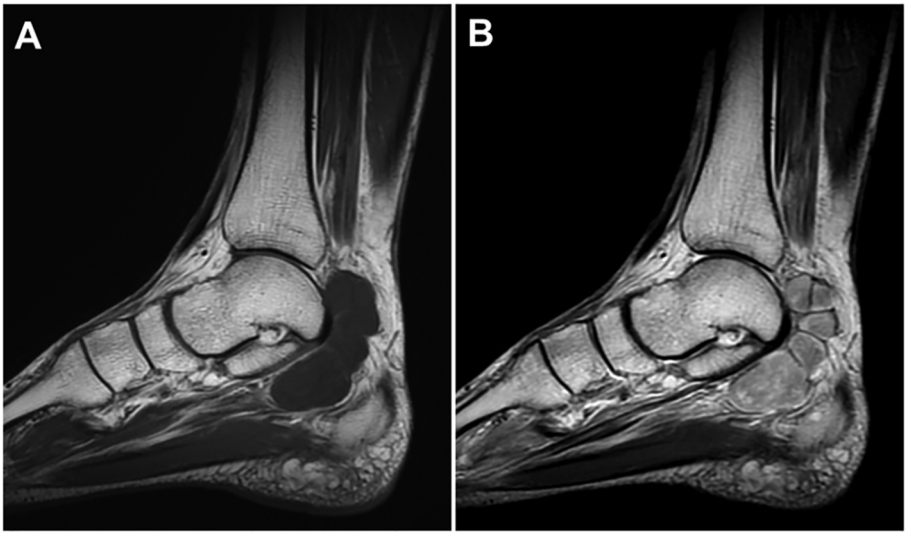

Figure 2. *Sagittal magnetic resonance images of multinodular/plexiform schwannoma in the right ankle. The multinodular lesions reveal intermediate signal intensity on T1-weighted sequences (A) and heterogeneous high signal intensity on T2-weighted sequences (B).*

neurological deficit at the 5-month follow-up. The patient was very satisfied with the outcome of the treatment.

Discussion

Multinodular/plexiform schwannoma accounts for up to 5% of all schwannomas (5, 6). It usually occurs in young and middle-aged adults, with no sex predilection (2, 4-6). Several cases have been encountered in children (9). It is generally considered that multinodular/plexiform schwannoma is not associated with neurofibromatosis or schwannomatosis.

Multinodular/plexiform schwannoma of the foot and ankle is extremely rare (10). Thus far, 17 cases of multinodular/plexiform schwannoma involving the foot and/or ankle have been reported in the English-language literature (2, 9, 11-23) (Table I). The patients ranged in age from 8 months to 66 years (mean, 27 years; median, 29 years). There was a slight male predominance. The most frequent location was found to be the planter aspect of the foot. Interestingly, 72.2% of tumors occurred on the right side of the foot and ankle. The tumor size ranged from 0.2 to 11 cm in diameter. Complete surgical excision or enucleation was typically curative.

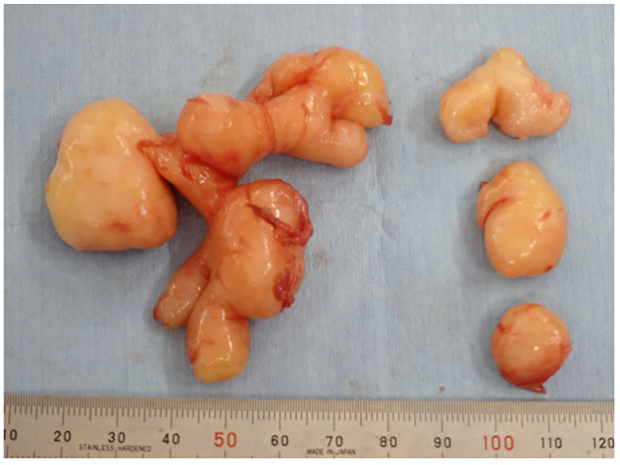

Figure 3. *Gross appearance of the well-circumscribed, yellowish-white nodules.*

On MRI, multinodular/plexiform schwannoma usually shows multiple bead-like nodular lesions (2, 24), as in our case. In general, the lesion reveals iso-signal intensity relative to skeletal muscle on T1-weighted sequences and high signal intensity on T2-weighted sequences. Like conventional schwannoma, target sign can be seen on T2-weighted sequences (8, 11, 24). Interestingly, Ikushima *et*

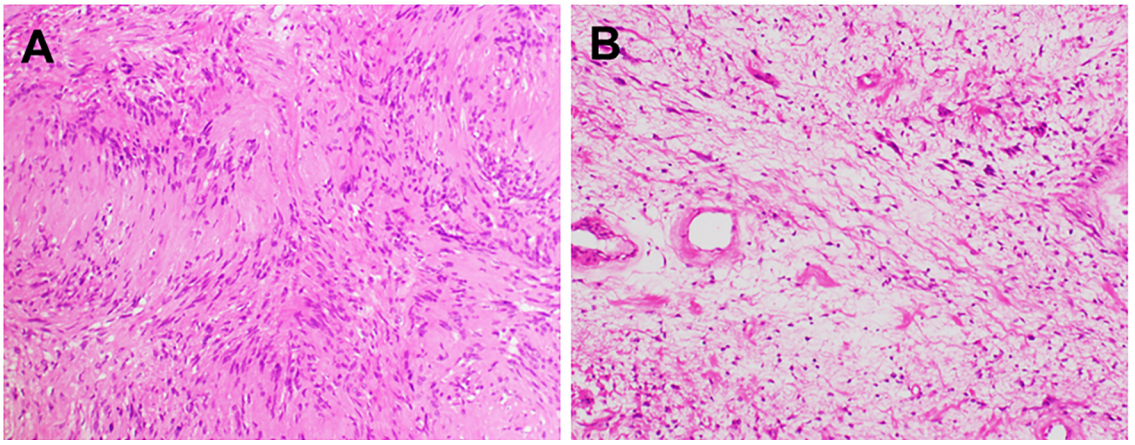

Figure 4. *Histological features of multinodular/plexiform schwannoma. A: The tumor is composed of spindle-shaped cells arranged in fascicles with occasional nuclear palisading (hematoxylin and eosin staining, original magnification ×100). B: Hypocellular Antoni B areas can be seen (hematoxylin and eosin staining, original magnification ×100).*

Table I. *Clinical characteristics of patients with multinodular/plexiform schwannoma of the foot and ankle.*

Case	Age/Sex	Site	Size (cm)	Treatment	Recurrence	FU (months)	Reference
1	32 years/F	Right ankle and foot	0.2-3.0	Enucleation	None	5	Nishio *et al.* (2)
2	8 months/M	Right foot	2.0	EB	Yes	18	Woodruff *et al.* (9)
3	8 years/M	Left foot	0.3-2.0	Excision	None	24	Ikushima *et al.* (11)
4	8 months/M	Right foot	2.0×1.9×1.8	Excision	Yes	18	Joste *et al.* (12)
5	29 years/M	Right ankle	6.0×4.0×2.8	Excision	None	27.6	Ioannou *et al.* (13)
6	65 years/M	Left forefoot	7.0×5.0×2.0	Excision	None	28	Jacobson *et al.* (14)
7	38 years/F	Right heel	1.2-4.2	Excision	NA	10	Mohammed *et al.* (15)
8	11 years/M	Right forefoot	4.5-6.0	Excision	None	36	Mohammed *et al.* (15)
9	19 years/F	Left foot	0.8-1.5	Excision	None	30	Li *et al.* (16)
10	53 years/F	Right foot	5.5×4.0	Excision	None	3	Matsuoka *et al.* (17)
11	66 years/M	Right foot	7.1×2.1×2.0	Excision	None	9	Ieong *et al.* (18)
12	33 years/M	Right second toe	3.0	Biopsy	NA	NA	Araghi *et al.* (19)
13	37 years/F	Right foot	4.7×3.5	Excision	NA	NA	Ajibowo *et al.* (20)
14	14 years/F	Right foot	11.0	Excision	None	12	Basnet *et al.* (21)
15	14 years/F	Left foot	NA	Excision	NA	NA	Agaram *et al.* (22)
16	2 years/M	Right foot	NA	Excision	NA	NA	Agaram *et al.* (22)
17	14 years/M	Left foot	1.5-3.0	Excision	NA	NA	Hammad *et al.* (23)
18	50 years/M	Right ankle	1.0-3.0	Enucleation	None	5	Present case

F: Female; M: male; EB: excisional biopsy; NA: not available; FU: follow-up.

al. (11) suggested that ambiguous curvilinear strands of low signal intensity on T1- and T2-weighted sequences might be a valuable imaging feature of this condition. The enhancement pattern of multinodular/plexiform schwannoma is variable (8).

Histologically, like conventional schwannoma, multinodular/plexiform schwannoma consists of bland spindle cells arranged in Antoni A and B patterns. Hyalinized blood vessels may be seen. Mitotic figures are generally few or absent. A cellular variant of this tumor has also been described (9, 22, 25). Plexiform cellular schwannoma most commonly occurs in infants and children and is composed entirely of Antoni A tissue characterized by spindle cells with hyperchromatic enlarged fusiform nuclei and faintly

eosinophilic cytoplasm. Mitotic figures are easily identified. Notably, it should be kept in mind that plexiform cellular schwannoma is easily mistaken as malignant peripheral nerve sheath tumor (MPNST) due to hypercellularity, hyperchromatism and high proliferative activity. On immunohistochemistry, like conventional schwannoma, multinodular/plexiform (cellular) schwannoma is diffusely positive for mature Schwann cell markers, including S-100 protein and SRY-box transcription factor 10 (SOX10) (4, 5, 9, 25, 26).

Surgery may be safely performed for symptomatic multiple/plexiform schwannoma extended along all the nerve lengths (7). In the current case, we performed the intracapsular enucleation to minimize the risk of nerve injury. However, Hébert-Blouin *et al.* (8) suggested that it might be difficult, if not impossible, to completely resect the lesion without a neurological deficit in multinodular/plexiform schwannoma. It should be noted that deep-seated multinodular/plexiform schwannoma has a higher risk of local recurrence (22). Our patient showed no evidence of local recurrence at the latest follow-up.

Complete or partial loss of chromosome 22 is the most common cytogenetic alteration in conventional schwannoma (27). In contrast, trisomy 17 has been identified as a recurrent chromosomal abnormality in plexiform cellular schwannoma (12, 28). Recently, Wali *et al.* (26) reported a case of orbital plexiform schwannoma harboring a novel SOX10 6 base pair in-frame insertion. Further investigations are required to better understand the correlation between these genomic alterations and clinicopathological features.

Conclusion

Multinodular/plexiform schwannoma is a distinctive subtype of schwannoma characterized by a plexiform or multinodular growth pattern. Careful clinicoradiological and histopathological evaluation can lead to an accurate diagnosis. Surgical enucleation is the primary treatment option with favorable outcome for symptomatic multinodular/plexiform schwannoma of the ankle.

Conflicts of Interest

The Authors declare no conflicts of interest associated with this article.

Authors' Contributions

YS was a major contributor and collected the data. JN performed the operation and drafted the article. YC reviewed the article. All Authors read and approved the final article.

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Artificial Intelligence (AI) Disclosure

No artificial intelligence (AI) tools, including large language models or machine learning software, were used in the preparation, analysis, or presentation of this article.

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