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Case Report

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Schwannomatosis of the tibial nerve

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ABSTRACT

Schwannoma is the most common type of benign tumor arising from the sheaths of the peripheral nerves. It occurs as a solitary tumor in most cases, but when it appears in multiple forms, it is necessary to differentiate it from plexiform schwannoma, schwannomatosis, neurofibroma and malignant peripheral nerve tumors. The authors experienced schwannomatosis in the tibial nerve without the features of neurofibromatosis type 2, so here we present a case report and literature review.

Keywords: Schwannoma, Schwannomatosis, Tibial nerve

INTRODUCTION

Schwannoma is one of the most common type of benign tumors of the central and peripheral nervous systems.[1] In most cases, it occurs as solitary tumor and rarely as multiple tumors. In those cases, it should be differentiated from plexiform schwannoma, schwannomatosis, neurofibromatosis type 2 (NF 2), and malignant peripheral nerve tumors.[2,3] Among these, schwannomatosis is defined as multiple occurrences with the general characteristics of peripheral and spinal schwannoma, but is not accompanied by vestibular schwannoma of the brain, as seen in NF 2.[2-4]

The authors observed multiple schwannomas along the tibial nerve, which were not related to NF 2, so here we report the case along with a literature review.

CASE REPORT

A 47-year-old female patient presented with a palpable mass on the medial side of the ankle, which had lasted for six years. She said that the mass seemed to grow little by little, but there was no complaint of pain. On examination, a 3×2 cm-sized mass was observed in the posterior-medial part of the right ankle and Tinel's sign was positive. There was no familial history of neurofibromatosis or café-au-lait spots on the skin. Magnetic resonance imaging (MRI) of the right ankle revealed a well-defined round mass in the tarsal tunnel and multiple oval-shaped masses along the course of the tibial nerve more proximally (Figure 1). An additional ultrasound examination was performed on the peripheral nerves of both lower extremities and a 1.5 cm-sized lobular mass was found in the tibial nerve in the middle of the lower leg, which was not included in the MRI of the ankle joint (Figure 2). The cranial MRI, which was performed to rule out NF 2, was normal.

Surgical treatment was performed for biopsy and mass resection. According to the surgical findings, a mass of about 2.5×1.8×1.2 cm was observed in the tarsal tunnel. The epineurium was carefully incised and then excised. In the proximal part of the tibial nerve, multiple masses were identified in the shape of multiple nodules, and the nerve fascicle was partially excised because it was impossible to separate it from the normal nerve bundle due to adhesion.

Histological findings revealed schwannoma (Figure 3) and immunostaining showed a positive reaction for S-100 protein (Figure 4). The patient complained of severe numbness in the right foot for about two weeks after

surgery, which improved after about one month. There was no evidence of muscle weakness in the plantar side of the foot, but she complained of hypesthesia in the lateral side of the sole during walking. An electromyography test performed six months after surgery confirmed tibial neuropathy.

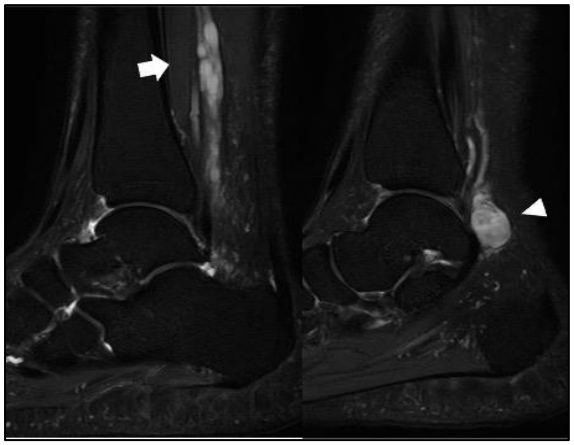

Figure 1: Sagittal T2-weighted image of the right ankle showing multinodular pattern (arrow) and round (arrow head) mass of the tibial nerve.

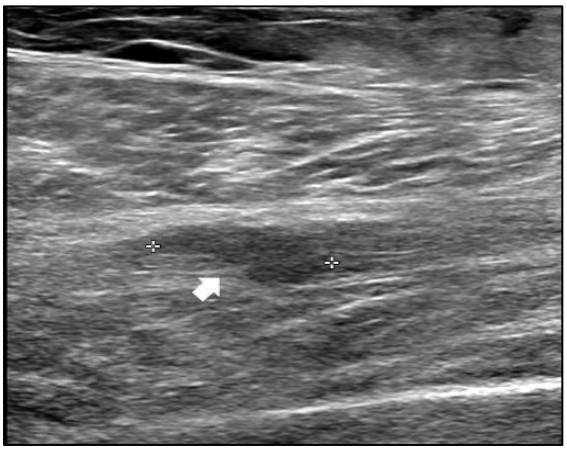

Figure 2: Ultrasound of calf area showing lobulated hypoechoic mass of the tibial nerve.

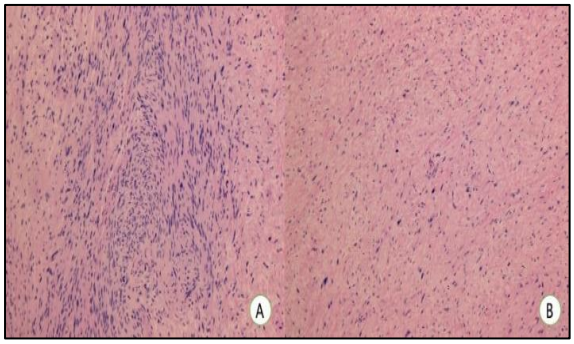

Figure 3: Photomicrograph showing schwannoma consisting of (A) Antoni A (spindle cells with nuclear palisading) and (B) B area. (H and E X100).

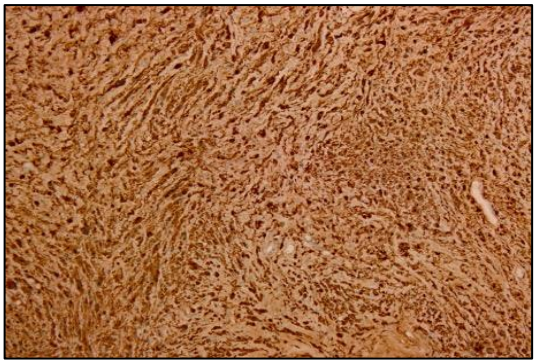

Figure 4: Diffuse positive immunostaining for S-100 protein.

DISCUSSION

Schwannomas are benign tumors originating from the peripheral nerve sheath and appear mainly as solitary lesions.3,5,6 However, they rarely occur as multiple lesions. Thus, in this case, multiple schwannomas, multiple plexiform schwannoma, schwannomatosis and neurofibromatosis should be included in the differential diagnosis- Multiple schwannomas including plexiform schwannoma and schwannomatosis are considered to be the same disease, but neurofibromatosis and malignant peripheral nerve tumors should be distinguished because confusion with those disease can lead to under or overtreatment.[1,3-8]

A plexiform schwannoma is a small, isolated mass in the skin and subcutaneous tissue, that rarely appears as multiple tumors in deep tissue.[5,6] It features multinodular lesions showing a plexiform macroscopically and histologically.[5] Yamamoto et al stated that schwannomatosis somewhat overlapped with plexiform schwannomas.[7] Hebert-Blouin et al believed that multinodular/plexiform schwannoma might be associated with NF 2 and schwannomatosis.[4] It is necessary to differentiate it from a malignant plexiform schwannoma because the cellularity and mitotic rate are often higher than those of a conventional schwannoma.[5,6]

NF 2 usually develops between the ages of 20 and 30 and is known as an autosomal dominant genetic disorder.8 It occurs in various parts of the body and appears mainly in the form of a meningioma, ependymoma, neurofibroma or schwannoma in peripheral nerves.[2,8,9] In NF 1 and NF 2, multiple nerve sheath tumors are observed which are mostly neurofibroma in NF 1 and schwannoma in NF 2.9 Schraepen et al stated that although schwannomatosis has a clinical picture somewhat similar to that of NF 2, it must be differentiated from NF 1 and especially NF 2 because the course of the disease and its prognosis may vary.[3] Therefore, in the case of multiple schwannomas in the upper and lower extremities, differentiation from NF 2 is required. The characteristic clinical finding of NF 2 is bilateral vestibular schwannoma, which can be confirmed by MRI of the brain. In this case, the patient was middle-

aged at presentation and did not have a family history of NF 1 or 2. In addition, the preoperative magnetic resonance imaging of the brain did not show a vestibular schwannoma. We believe that NF 1 and NF 2 should be included in the differential diagnosis of schwannomatosis and brain imaging studies are needed.

Due to not continuously involving entire major peripheral nerve with multiple nodules, so additional ultrasound examination or MRI of the entire nerve should be necessary. In the current case, the additional ultrasound examination revealed an additional mass in the tibial nerve in the proximal one-third of the lower leg, which was not included in the MRI of the ankle joint. Therefore, additional examinations such as an ultrasound or MRI of the proximal and distal regions as well as the lesion itself are necessary before surgery.

CONCLUSION

There is still controversy as to whether schwannomatosis is a disease with clinical and histologic features or a subtype included in NF 2. However, in the case of multiple schwannomas occurring in the peripheral nerve, ultrasound examination of the entire major peripheral nerve involved and brain imaging should be required, and the orthopedic surgeon should be familiar with the clinical manifestation of NF 1 and NF 2 such as café-au lait spots on the skin and bilateral vestibular schwannoma.

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