Neurofibromas are benign tumors arising from the peripheral nerve sheath. Neurofibromatosis type 1 (NF1), which is one of the most common autosomal dominant disorders, consists of multiple neurofibromas. Neurofibromas may rarely exist singly in people without neurofibromatosis type 1 at any location along the peripheral nervous system. Segmental neurofibromatosis is a rare form of neurofibromatosis in which skin lesions are limited to a region of the body. Here we report a patient of segmental neurofibromatosis who had multiple localized dermal neurofibromas on the lower limb with no other cutaneous or systemic findings. This is the first report of isolated segmental neurofibromatosis in confined to the posterior tibial nerve dermatome.

**Key Words:** Neurofibromatoses; neurofibroma

---

**ABSTRACT** Neurofibromas are benign tumors arising from the peripheral nerve sheath. Neurofibromatosis type I is one of the most commonly seen autosomal dominant disorders which consists of multiple neurofibromas. Neurofibromas may rarely exist singly in people without neurofibromatosis type I at any location along the peripheral nervous system. Segmental neurofibromatosis is a rare form of neurofibromatosis in which skin lesions are limited to a region of the body. Here we report a patient of segmental neurofibromatosis who had multiple localized dermal neurofibromas on the lower limb with no other cutaneous or systemic findings. This is the first report of isolated segmental neurofibromatosis in confined to the posterior tibial nerve dermatome.

**Key Words:** Neurofibromatoses; neurofibroma

---

**A Case of Segmental Neurofibromatosis in Confined to Posterior Tibial Nerve Dermatome**

**Posterior Tibial Sinir Dermatomuna Sınırlı Bir Segmental Nörofibromatozis Olgusu**


**Anahtar Kelimeler:** Nörofibromatozis; nörofibrom

**Türkiye Klinikleri J Dermatol 2014;24(2):90-2**
CASE REPORT

A 30-year-old female presented to dermatology outpatient department with asymptomatic swellings on her right leg of 15 years duration. The patient was complaining about an increase in the size and number of tumors in the last two years. Dermatologic examination revealed soft, multiple, flesh-colored, cutaneous nodules (ranging from 1-2 mm to 1 cm in diameter) over the right posterior tibial region in a dermatomal distribution (Figure 1). Clinically we considered these lesions as neurofibroma. However, there were no nodules or other cutaneous findings of NF1 at the rest of the body. Optic glioma or lisch nodules were not detected on ocular examination. The patient had no first degree relative with NF1. No other abnormalities were found in a detailed neurological examination. A clinical diagnosis of SN was considered. One of the nodular lesions was completely excised under local anesthesia for histopathological examination. Histopathology of the tumor revealed non-encapsulated but well-circumscribed lesion located in the dermis and subcutis (Figure 2). Proliferation of spindle cells with wavy nuclei located in fibrous stoma was seen. Atypical mitoses or nuclear pleomorphism were absent (Figure 3). These findings confirmed the clinical diagnosis of SN.

DISCUSSION

In contrast to NF1, SN is a rare disorder characterized by localized cutaneous neurofibromas and/or café-au-lait macules with no systemic involvement.\(^2\) Distribution of tumors is commonly unilateral but can be bilateral. The most common affected site of the body is the thoraco-abdominal...
region.\textsuperscript{3} Axillary freckling and family history have been reported in limited number of cases. Cutaneous neurofibromas are usually asymptomatic but especially those deep seated on peripheral nerves and spinal roots can cause neurological dysfunction. The prevalence of complications including malignancies in SN is much lower than the NF1.\textsuperscript{1}

The etiology of SN is not well-known but it is thought to arise from a postzygotic mutation in the NF1 gene of primitive neural crest cells leading to somatic mosaicism. Reguler NF-1, sarcoidosis, benign tumors (trichoepithelioma, leiomyoma), cutaneous lymphomas, xanthomas should be considered in the differential diagnosis of the disease.\textsuperscript{4}

As in our case, cutaneous lesions can be observed in a limited dermatome and can be diagnosed incidentally while the patient is being examined for a different complaint. Therefore, a thorough physical, ophthalmologic and neurologic examination should be performed in patients with SN for rule out generalized disease that may present with systemic complications.

\begin{thebibliography}{9}


2. Gabhane SK, Kotwal MN, Bobhate SK. Segmental neurofibromatosis: a report of 3 cases.


\end{thebibliography}