Olgu Sunumu
Case Report

Multiple schwannoma unrelated with neurofibromatosis

NÖROFİBROMATOZİS İLİŞKİSİZ MULTIPLE SCHWANNOMA

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ABSTRACT

This is a report of a case of multiple schwannoma without any neurofibromatosis (NF) patterns. Patient's history, physical examination findings, radiological and histopathological examination findings were evaluated using the records of the hospital information system and the patient's file. The patient and his family did not have a history of NF. Excision of all symptomatic lesions was performed. Histopathological examination confirmed all lesions as schwannoma. The patient does not have a new lesion on his twenty forth postoperative month and he is satisfied with his life. The disease that is present with multiple schwannomas and that is not related with neurofibromatosis type 2 is rare. The authors recommend surgical excision for symptomatic lesions.

Keywords: neurofibromatosis, schwannomatosis, multiple primary schwannomas

ÖZ

Bu yazı, nörofibromatozis (NF) özellikleri olmayan bir multiple schwannoma olgusu raporudur. Hastane bilgi sistemi kayıtları ve hasta dosyasından hastanın öyküsü, fizik muayene bulguları, radyolojik ve histopatolojik inceleme bulguları incelendi. Hastanın ve ailesinin NF öyküsü yoktu. Semptomatik lezyonların tümüne total eksizyon yapıldı. Histopatolojik inceleme, tüm lezyonları schwannom olarak doğruladı. Hastanın post-operatif on ikinci ayında yeni lezyonu yok ve yaşamından memnundur. Nörofibromatozis tip 2 ile ilişkisiz olup, multipl lezyonlar şeklinde ortaya çıkan schwannomlar nadirdir. Yazarlar semptomatik lezyonlarda cerrahi eksizyon önermektedir.

Anahtar Sözcükler: nörofibromatozis, schwannomatozis, multiple primer schwannomalar

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Schwannomas are the encapsulated peripheral nerve tumors that have benign character and slow-growing nature. Similar to neurofibromas, they are caused stemmed from nerve sheath. They originate from the outer side of the nerve but can damage the adjacent structures (1, 2).

Schwannomas are generally solitary lesions but sometimes may affect one or more nerves (3). The presence of multiple schwannomas in the same patient reminds of a syndrome especially neurofibromatosis type 2 (4).

In this case report, we present a patient with multiple schwannomas without any NF findings.

CASE PRESENTATION

A 55-year-old female patient was admitted to the orthopedics outpatient clinic with swellings on both of the crurises. The history revealed that the swellings had been present for about 2 years, it grew very slowly and did not cause any pain at the beginning but pain had started in the last two months.

Written informed consent was obtained from the patient's legal custodian or first-degree relatives for publishing the individual medical records.

There were palpable swellings that are painful, immobile and doughy on the distal lateral of the right cruris and proximal and distal lateral of the left cruris on physical examination. They were on the course of peroneal nerve. Neurovascular examination was normal in the distal part of the lesions.

The patient was radiographed. Bone structures were normal. Soft tissue masses that were hard to differentiate from soft tissue and had more radioopacity comparing to the normal soft tissue (Figure 1).

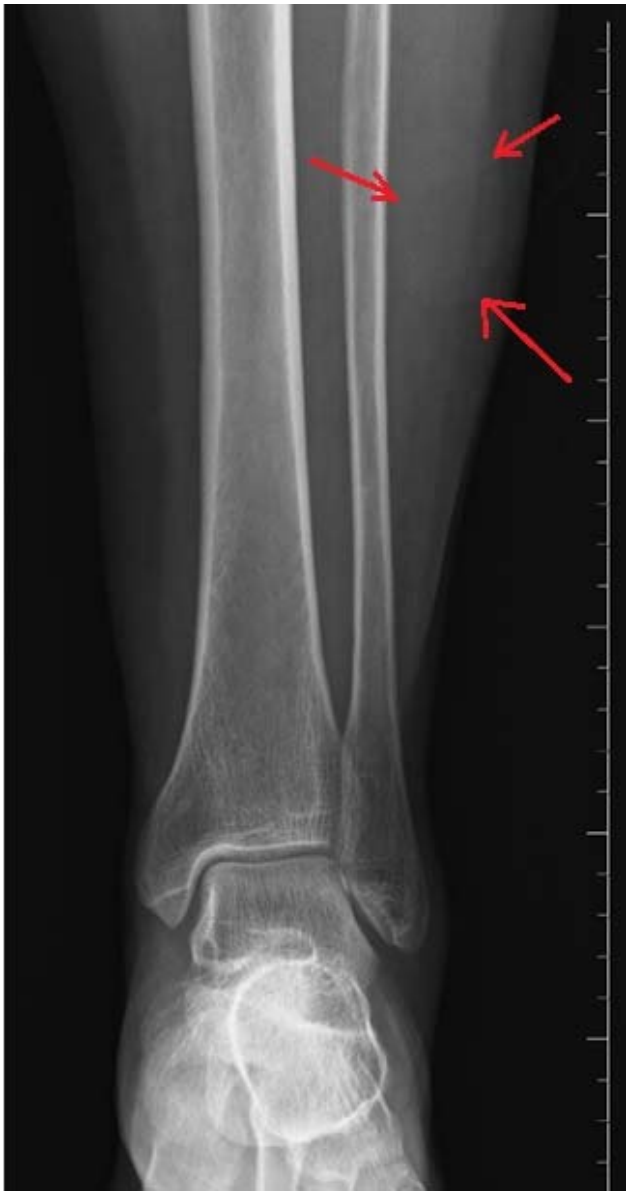

Figure 1: Radiograph shows soft tissue mass in soft tissues, showing mild radioopacity compared to normal tissues.

Magnetic resonance imaging (MRI) was performed. Soft tissue masses that were well-circumscribed, in different sizes and hypointense on T1 and T2 sequences were observed in both crurises. The localization of the lesions was correlated with peroneal nerve course (Figure 2).

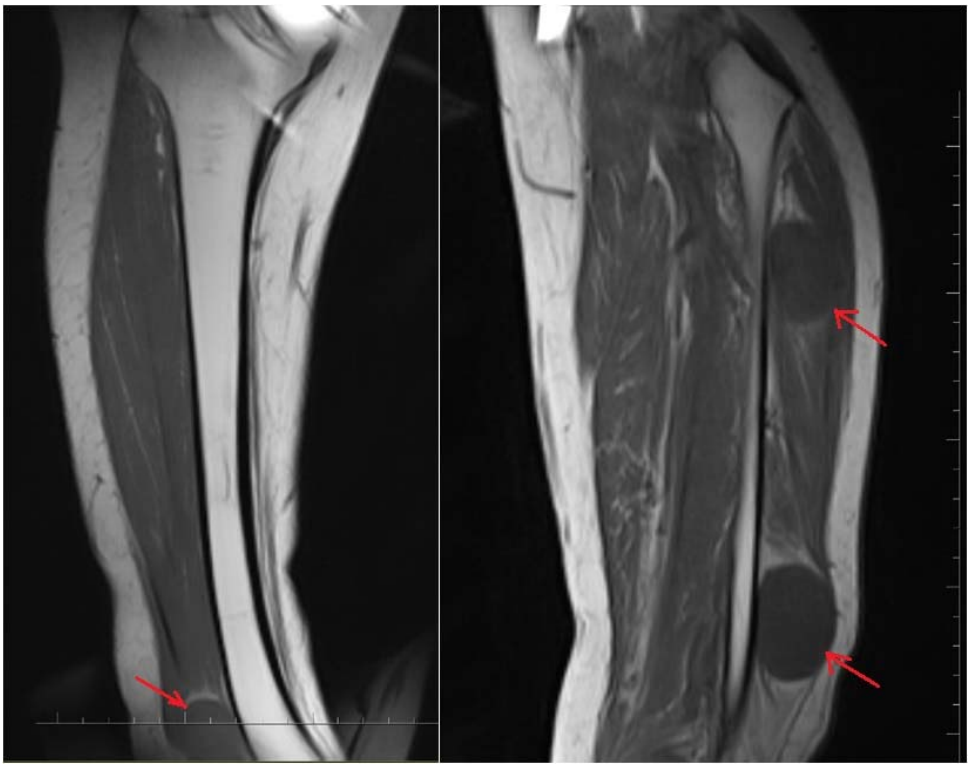

Figure 2: In MRI, in both crurises, hypointense in T1 and T2 sequences, in well-limited soft tissue masses in peroneal nerve course.

Excisional biopsy of three lesions was performed with a pre-diagnosis of schwannoma after patient's history evaluation, physical examination and radiological examination. During the operation, it was seen that the lesions were derived from the peroneal nerves, but the nerve was not damaged in the macroscopic examination (Figure 3 and Figure 4).

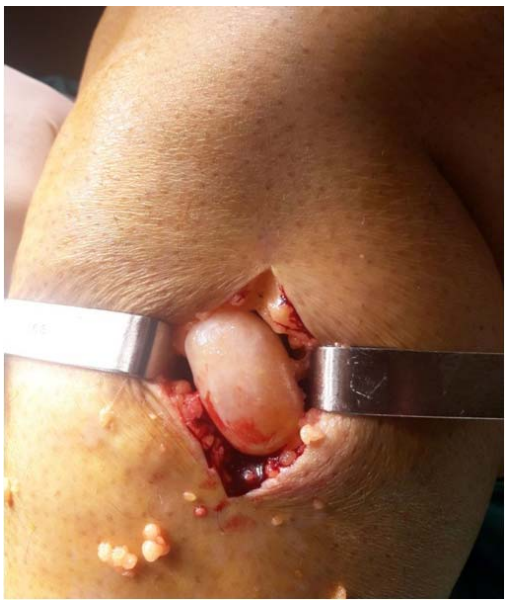

Figure 3: Intraoperative imaging, oval doughy well-limited tumor.

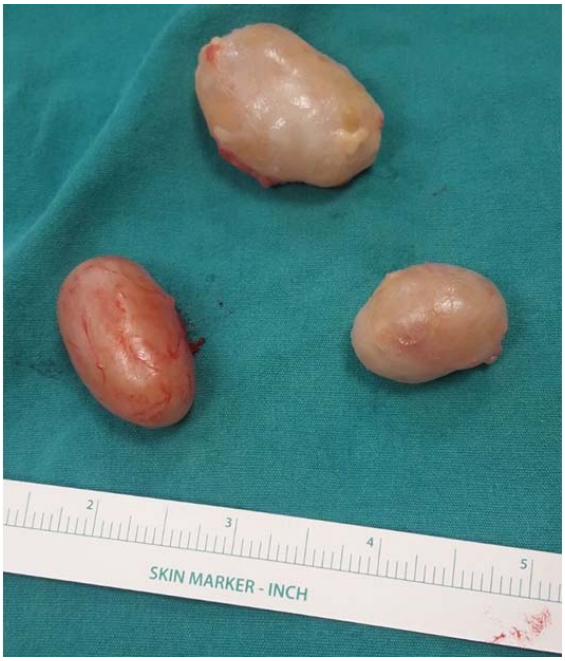

Figure 4: Macroscopic appearance of pathological materials excised.

All lesions were totally excised with maximum care for nerve structures. Histopathological examination of the lesions revealed schwannoma. The patient was evaluated on the post-operative 15th day and the sutures were removed. The pain was completely gone and there was no neurovascular symptom.

The patient was followed up periodically. There was no new lesion and the patient had no complaint at the postoperative 24th month of follow-up.

DISCUSSION

Schwannomas are very heterogeneous tumors, possibly involving a mixture of proliferative nerve sheath cells originating from perineural fibroblasts. The tumor infiltrates the nerve and takes the place of some nerve fibers. In addition, some schwannomas maybe cellular and mimic sarcoma in histological appearance (5, 6).

Multiple schwannomas in the same patient may give rise to thought neurofibromatosis type 2. In 2/3s of patients affected by NF2, schwannoma develops and can be seen before vestibular tumors (3, 4).

Reports of individuals with multiple schwannomas without evidence of other features of vestibular schwannoma or NF2 have been reported by various authors. These authors suggested that Schwannomatosis is different from other forms of neurofibromatosis (7).

Among neurofibromatosis syndromes, NF1 is the most common with an estimated incidence of 1/3000 at birth, representing more than 90% of all neurofibromatosis patients (8, 9).

Currently, there is no definitive diagnostic criteria for schwannomatosis internationally. Jacoby et al. suggested clinical criteria for the diagnosis of schwannomatosis (10). These criteria include the presence of two or more schwannomas that are pathologically proven and the absence of radiographic evidence of vestibular tumors if the patient is older than 18 years of age.

If there is no brain MRI, if the patient has two or more pathologically proven schwannomas and is more than 30 years old, a possible or hypothetical diagnosis can be made if the eighth nerve symptoms are not present. Michael et al. also revised that none of the current criteria for diagnosing neurofibromatosis were present in the patient (11).

Schwannomas can be seen in bones too very rarely (12, 13). Schwannomatosis usually occurs in the fourth year, unlike the earlier NF2 patients (3). Our patient was in the 5th decade.

A large number of schwannomas develop in the cranial, spinal and peripheral nerves in patients with Schwannomatosis but no vestibular schwannoma(VS) develops. They do not develop other tumors such as meningiomas, ependymomas or astrocytomas (1, 3, 7, 11)

The pain is the most common presenting symptom of the schwannomatosis. However NF2 patients show neurological deficit (3, 4).

Myxoid stroma, nerve edema or intraneural growth pattern in histology support the diagnosis of schwannomatosis. Molecular and genetic analyses also indicate that schwannomatosis is a prominent genetic and clinical syndrome (3).

Finally, in patients with multiple soft tissue swelling, lesions should be considered in the differential diagnosis, especially if the lesion localization in a nerve course. Schwannomatosis is a disease different from neurofibromatosis. Prognosis is satisfying after total excision by preserving neurovascular structures in symptomatic lesions.

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