Asymptomatic Spontaneous Segmental Neuroma: Case Report

Asemptomatik Spontan Segmental Nöroma

Handan KELEKÇİ,[a]
Şemsettin KARACA,[a]
Ali KARAKUZU,[a]
Selçuk ÖZYURT,[a]
Aylin ORGEN ÇALLI[b]

Clinics of
[a]Dermatology,
[b]Pathology,
İzmir Katip Çelebi University
Atatürk Training and Research Hospital,
İzmir

Geliş Tarihi/*Received:* 12.03.2014
Kabul Tarihi/*Accepted:* 05.07.2014

Yazışma Adresi/*Correspondence:*
Selçuk ÖZYURT
İzmir Katip Çelebi University
Atatürk Training and Research Hospital,
Clinic of Dermatolgy, İzmir,
TÜRKİYE/TURKEY
ozyurtselcuk@yahoo.com

ABSTRACT Peripheral nerve neuromas are benign hyperplastic proliferative lesions. They are most commonly seen as clinically painful papules or nodules in adults. Neuromas may develop following trauma, infection, scarring and amputation or may be spontaneous, but sometimes they may be an early sign of multiple endocrine neoplasia syndrome type 2B (MEN) 2B. Also, Phosphatase and tensin homolog gen (PTEN) hamartoma-tumor syndromes such as Cowden, Banayan- Riley- Ruvalcaba and Proteus like syndromes can be associated with neuromas. Histopathologic examination of neuromas reveal fibrous scar tissue embedded in irregular bundles of cross-linked nerve fascicles and S-100 protein positive nerve fascicles. An 18 year-old patient with spontaneous painless neuroma is presented to remind that neuroma may occur without pain and previous trauma, and possible associating syndromes should be kept in mind.

Key Words: Neuroma; skin

ÖZET Periferik sinir nöromaları selim hiperplastik proliferatif lezyonlardır. Daha çok erişkinlerde, klinik olarak ağrılı papül ya da nodüller olarak görülürler. Nöromalar travma, enfeksiyon, yaralanma ve amputasyonu takip edebileceği gibi spontan olarak da gelişebilirler. Ancak bazen multipl endokrin neoplazi sendromu tip 2B (MEN) 2B'nin erken bulgusu olabilirler. Ayrıca, Cowden, Banayan- Riley- Ruvalcaba ve Proteus benzeri sendromlar gibi Fosfataz ve tensin homolog gen (PTEN) hamartoma-tümör sendromları nöroma ile birlikte olabilirler. Nöromanın histopatolojik incelemesinde düzensiz demetler halinde çaprazlama sinir fasikülleri arasına yerleşmiş fibröz skar dokusu ve S-100 protein pozitif sinir fasikülleri görülebilir. Nöromaların travma öyküsü olmadan ve ağrısız olarak ortaya çıkabileceğine dikkat çekmek ve sendromlarla ilişkili olabileceğini hatırlatmak amacıyla 18 yaşında, spontan ağrısız nöromaları olan bir hasta sunulmaktadır.

Anahtar Kelimeler: Nöroma; cilt

Turkiye Klinikleri J Case Rep 2015;23(3):299-301

Peripheral nerve neuromas are benign hyperplastic proliferative lesions. Clinically painful papules or nodules occur in adults. A single nodule or papule on the back develops generally in response to minor trauma.[1] The diagnosis of mucosal neuromas is important as they may be part of some syndromes, such as MEN 2B and Cowden Syndrome.[2] We present a patient with rarely seen asymptomatic and segmental neuromas.

CASE REPORT

A 18 year-old woman was admitted to our clinic with multiple skin colored nodules and papules on the medial part of her left foot, distal portion of

doi: 10.5336/caserep.2014-39741

2nd, 3rd and 4th toes and the proximal portion of 5th left toe (Figure 1). Her complaints started four years earlier and increased in number gradually. She had no other diseases and routine tests were normal. A biopsy from the back of her foot revealed, randomly scattered nerve sections in the papillary dermis and perineural fibrosis. S-100 protein was considered positive in the immunohistochemical staining (Figure 2, 3). The painless, segmental lesions were diagnosed as neuroma with these clinical and histopathological findings. Lesions were managed by cryotherapy.

DISCUSSION

Neuromas are rare neurogenic tumors of skin, subcutaneous tissues and mucous membranes. Traumatic neuromas are benign formations due to partially or completely interrupted neural pathways. Other than trauma, infection, scarring and amputation can cause these formations. Small, painfull, oval lesions can be clinically seen.[3] Cutaneous neuroma case reports are few in the literature and mostly solitary lesions secondary to surgery or trauma have been reported. Neurotouch sensitive dysestetic nodules are observed below the incision in amputation neuromas.[3,4] Development of multiple painless nodules and papules throughout several months was remarkable in our patient without a history of trauma and surgical procedures.

Multiple mucosal neuromas may be an early sign of multiple endocrine neoplasia syndrome type 2B (MEN2B). MEN 2B is charecterized by medullary thyroid carcinoma, pheochramocytoma and mucosal neuromas often localized to tongue, lip and buccal mucous membranes. Other symptoms are skeletal and ocular abnormalities.[4,5] Skin and neural tissue anomalies can often be observed in PTEN hamartoma-tumor syndromes associated with PTEN tumor suppressor gene. PTEN hamartoma-tumor syndromes such as Cowden, Banayan-Riley-Ruvalcaba and Proteus like syndrome can be associated with neuromas. In Cowden syndrome tricholemmomas, oral papillomatosis,

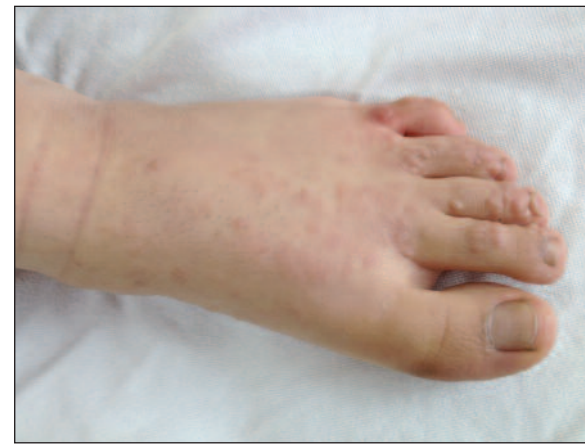

FIGURE 1: Skin colored nodules and papules on the medial side of left foot and distal portion of 2nd, 3rd and 4th toes and the proximal portion of 5th toe.

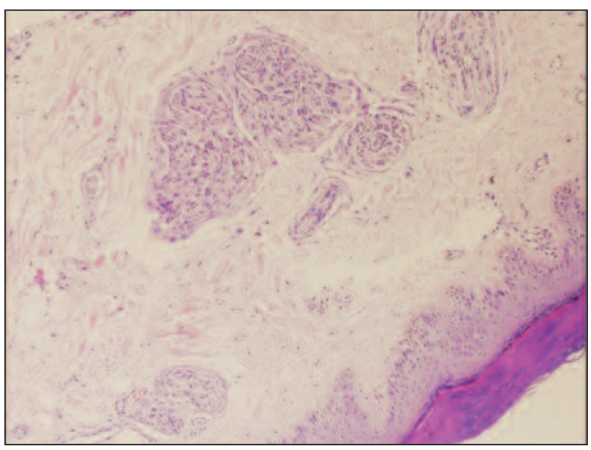

FIGURE 2: Mild perineural fibrosis and scattered nerve sections in papillary dermis (H&E, 400x).

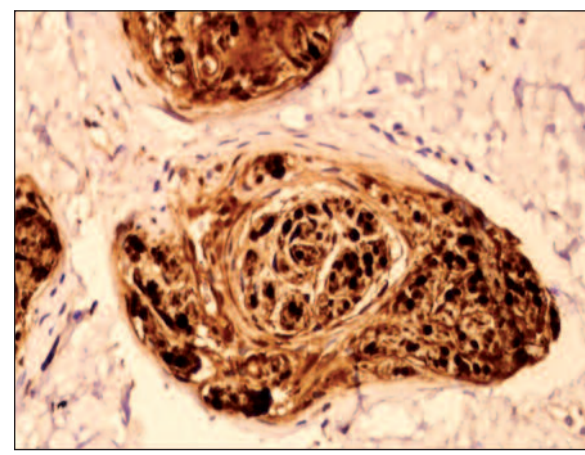

FIGURE 3: Immunohistochemical staining: S-100 protein positive nerve fascicles.

acral keratoses, sclerotic fibroma and gastrointestinal polyps are observed. Bannayan-Riley-Ruvalcaba syndrome is characterized by acrochordon, acanthosis nigricans, cafe au lait spots, macular pigmentation in the genital area and gastrointestinal polyps.[6]

The histopathologic examination of neuroma reveals; fibrous scar tissue embedded in irregular bundles of cross-linked nerve fascicles. S-100 protein positive nerve fascicles containing schwann cells are surrounded by epithelial membrane antigen positive perineural cells. In our case, nerve sections randomly scattered in the papillary dermis and mild perineural fibrosis were detected. Immunohistochemical examination revealed S-100 protein positivity. Histopathological differential diagnosis contains: Localized interdigital neuritis (Morton's neuroma), encapsulated palisade neuroma, neurofibroma and schwannoma. Interdigital neuritis and schwannoma can be distinguished from neuromas by the absence of reduction in axons.[1]

Treatment is often simple surgical excision.[3] Our patient underwent cryotherapy for papular lesions on the foot. Post inflammatory hyperpigmentation was observed after cryotherapy.

Neuromas may develop spontaneously without trauma and it should be kept in mind that pain symptom may not always occur in patients.

REFERENCES

1. Rodríguez-Peralto JL, Riveiro-Falkenbach E, Carrillo R. Benign cutaneous neural tumors. Semin Diagn Pathol 2013;30(1):45-57.
2. Qualia CM, Brown MR, Ryan CK, Rossi TM. Oral Mucosal Neuromas Leading to the Diagnosis of Multiple Endocrine Neoplasia Type 2B in a Child with Intestinal Pseudo-obstruction. Gastroenterol Hepatol (N Y) 2007;3(3):208-11.
3. Serrano Falcón C, Serrano Falcón Mdel M, Ruiz Villaverde R, Linares Solano J, Serrano Ortega S. Amputation neuromas after neck surgery. Dermatol Online J 2005;11(2):24.
4. Li Q, Gao EL, Yang YL, Hu HY, Hu XQ. Traumatic neuroma in a patient with breast cancer after mastectomy: a case report and review of the literature. World J Surg Oncol 2012;10:35.
5. Truchot F, Gréazard P, Wolf F, Balme B, Perrot H. Multiple idiopathic mucocutaneous neuromas: a new entity? Br J Dermatol 2001;145(5):826-9.
6. Schaffer JV, Kamino H, Witkiewicz A, McNiff JM, Orlow SJ. Mucocutaneous neuromas: an under recognized manifestation of PTEN hamartoma-tumor syndrome. Arch Dermatol 2006;142(5):625-32.