

## Plexiform Schwannoma of the Finger: A Case Report and Literature Review

### Abstract

A 49-year-old woman with a long history of a subcutaneous mass on the dorsal side of her 4<sup>th</sup> finger of the right hand visited a dermatologist because of slight enlargement of the mass. Her past medical history was notable only for a mitral valvuloplasty performed 20 years earlier. Physical examination revealed a small, round, firm subcutaneous mass on the dorsal side of her proximal interphalangeal joint of the right 4<sup>th</sup> finger. The mass was immobile and nontender and its overlying skin was intact. An excisional biopsy was done for the patient and the specimen was sent for pathologic evaluation. On microscopic examination, the final diagnosis of plexiform schwannoma was made for the lesion. The aim of this publication is to report a rare case of plexiform schwannoma of the soft tissue and a literature review to provide a better understanding about its characteristics including epidemiologic factors and pathologic evaluation.

**Keywords:** Finger, plexiform, schwannoma, soft tissue

### Introduction

Plexiform schwannoma is a rare variant accounting for approximately 5% of all schwannomas.<sup>[1,2]</sup> It has a plexiform pattern of intraneural growth either in gross or histologic examination and is frequently accompanied by multinodularity.<sup>[2]</sup> In most solitary cases, there are no identifiable risk factors, although multiple tumors are associated with some risk factors including neurofibromatosis type II, trauma, and positive family history.<sup>[3]</sup> Plexiform schwannoma usually presents in childhood, and even congenital forms have been reported. The incidence in men and women is equal.<sup>[3]</sup> The most common sites of involvement are trunk, head, and neck and upper extremities.<sup>[1,3]</sup> These tumors are usually slow growing and asymptomatic and the average size is less than 2 cm.<sup>[1,3]</sup>

We present a rare case of plexiform schwannoma of the finger in a woman with no known risk factors for developing this tumor.

### Case Report

A 49-year-old woman with the complaint of 15-year history of a small mass on the right 4<sup>th</sup> finger was referred to the dermatology clinic. Her past medical history revealed

that a mitral valvuloplasty was performed 20 years before; she was otherwise healthy according to her statements. Family history was negative and there was no history of trauma to the site of the mass. The mass did not cause any pain or disability and there was a slight increase in its size which made the patient visit the physician.

On physical examination, there was a firm and round subcutaneous mass on the dorsal side of the proximal interphalangeal joint of the right 4<sup>th</sup> finger [Figure 1]. The mass measured approximately 1 cm in diameter and was immobile and nontender. The overlying skin had normal appearance, and the range of motion of the affected finger was intact. There were no other notable findings in her physical examination. The patient underwent an excisional biopsy and the mass was sent to our laboratory for pathologic evaluation.

### Pathologic findings

On gross examination, the specimen in formalin container was an irregular skin tissue measuring 1 × 0.6 × 0.4 cm and a tan, firm, encapsulated oval-shaped nodule with a maximum diameter of 0.7 cm attached to the deep side of the skin tissue was evident. After tissue processing and preparing glass slides, microscopic evaluation revealed a

Nafiseh Mortazavi,  
Kambiz Novin<sup>1</sup>,  
Farahnaz Bidari  
Zerehpooosh,  
Managol  
Sadatsafavi

Departments of Pathology and  
<sup>1</sup>Clinical Oncology, Shahid  
Beheshti University of Medical  
Sciences, Tehran, Iran

### Address for correspondence:

Dr. Kambiz Novin,

Department of Clinical  
Oncology, Emam Hossein  
Medical and Educational  
Center, Shahid Madani St,  
Tehran, Iran.

E-mail: dr\_novin@sbmu.ac.ir

### Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.IDOJ\_370\_16

### Quick Response Code:



**How to cite this article:** Mortazavi N, Novin K, Zerehpooosh FB, Sadatsafavi M. Plexiform schwannoma of the finger: A case report and literature review. Indian Dermatol Online J 2017;8:355-7.

**Received:** November, 2016. **Accepted:** April, 2017.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

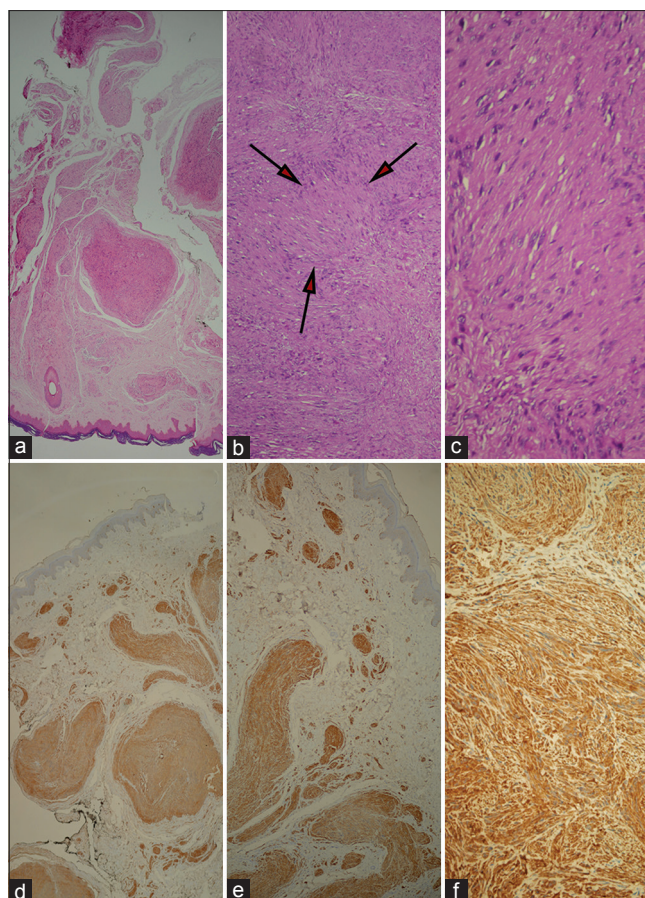


**Figure 1:** (a and b) Subcutaneous mass on the dorsal side of the proximal interphalangeal joint of the right 4<sup>th</sup> finger

dermal neoplasm consisting of nodular proliferation of neoplastic cells with round to oval and sometimes wavy nuclei, some of which were arrayed with palisading features and formed verocay bodies. On immunohistochemical study, S100 protein was strongly expressed in tumoral cells [Figure 2]. The neoplastic nodules were surrounded by a fibrous capsule in some foci. The lesion lacked necrosis, there was no marked cell crowding, or generalized nuclear atypia accompanied by significant mitotic activity seen in the malignant counterpart called malignant peripheral nerve sheath tumor (MPNST).<sup>[4]</sup> Based on histologic findings, the main differential diagnoses were plexiform schwannoma and plexiform neurofibroma. However, considering the significant correlation of the plexiform neurofibroma with neurofibromatosis type I<sup>[5,6]</sup> and the negative history of the patient as well as microscopic findings such as the presence of verocay bodies, lack of myxoid stroma and diffuse, and strong immunohistochemical staining for S100 protein, we excluded the diagnosis of plexiform neurofibroma, and the final diagnosis of plexiform schwannoma was made.

## Discussion

Schwannoma is classified as peripheral nerve sheath tumor which originates from the Schwann cells derived from neural crest.<sup>[7]</sup> It can occur as peripheral, visceral, intraspinal, or intracranial tumor. Peripheral schwannoma often manifests by an asymptomatic papule or nodule which can be yellow or light brown and cystic degeneration or hemorrhage can occur inside it.<sup>[8]</sup> Cutaneous schwannoma is not common and presents as a slow growing solitary or less frequently multiple masses.<sup>[9]</sup> There are some histologic variants – cellular, plexiform, and melanotic subtypes.<sup>[4]</sup> Among all subtypes of the uncommon cutaneous form of this neoplasm, the rare plexiform subtype accounts for approximately 5%.<sup>[1,2]</sup> The aim of this case review which presented a rare subtype of cutaneous schwannoma, the plexiform subtype in a patient without any predisposing factor was to make a better understanding for the



**Figure 2:** (a-c) Hematoxylin and eosin stained slides from the skin mass, (a) Multiple nodules in dermis with plexiform architecture (H and E,  $\times 4$ ). (b) Verocay bodies formed by alternating rows of palisading nuclei and intervening nuclei free stroma. Arrows show one prominent verocay body (H and E,  $\times 10$ ). (c) Nuclear palisading (H and E,  $\times 40$ ). (d-f), Immunohistochemical stained slides for S100 protein: purely and strongly S100 positive Schwann cell population, (d)  $\times 4$ . (e)  $\times 10$ . (f)  $\times 40$

diagnosis of such a rare neoplasm. Cutaneous form of this tumor is usually located in the hypodermal fat, however, there are some cases with dermal location,<sup>[9]</sup> similar to our patient whose lesion was a dermal neoplasm. The solitary plexiform schwannomas occur in patients with no significant predisposing factor, however, multiple tumors are more frequent in individuals with neurofibromatosis type II, schwannomatosis, Gorlin–Koutlas syndrome, and patients with positive family history or history of trauma.<sup>[3]</sup> According to these facts, an absence of risk factors in our case with a single tumor is not a reason for excluding the diagnosis of plexiform schwannoma. Trunk, head and neck, and upper extremity are the most frequent involved locations.<sup>[1,3]</sup> The location of the tumor in this case is compatible with the site of involvement of plexiform schwannoma reported in literature. This tumor routinely manifests in childhood, although it may present as a congenital lesion.<sup>[3]</sup> Our patient stated that she has had the lesion since approximately 15 years. As she mentioned during our medical interview, she could not remember the exact duration of having the lesion. No sex predilection

has been reported for plexiform schwannoma.<sup>[3]</sup> Malignant transformation of this benign neoplasm is very rare.<sup>[9,10]</sup> There are some case reports of benign solitary schwannomas with a consequent malignant transformation.<sup>[10-12]</sup> Differential diagnosis for plexiform schwannoma are plexiform neurofibroma and MPNST.<sup>[13]</sup> Plexiform neurofibroma has a tortuous mass of expanded nerve branches involved by the neurofibroma which is formed by a combined random proliferation of all the elements of a peripheral nerve (axons, Schwann cells, fibroblast, and perineural cells) in a background of loose myxoid stroma, lacking well formed verocay body.<sup>[4]</sup> Although plexiform schwannoma has a superficial resemblance to the plexiform neurofibroma characterized by multinodular growth, it is composed purely of Schwann cells with nuclear palisading and verocay bodies without myxoid changes of stroma.<sup>[13]</sup> Whereas S100 protein staining identifies a purely and strongly positive Schwann cell population in plexiform schwannoma, variable expression of this antigen is observed in plexiform neurofibroma.<sup>[4,13]</sup> To exclude the malignant counterpart, the combination of pathologic findings (cellularity, mitotic activity, and atypia), as well as immunohistochemical study for S100 protein (most MPNSTs show negative or weak staining) should be considered,<sup>[4]</sup> as noted in the present case. Finally, to consider others less probable differential diagnosis with palisading pattern, such as palisaded neuroma or palisaded leiomyoma, attention to both pathologic and immunohistochemical findings is helpful. Although focal palisading is often present in cellular nodules of palisaded neuroma, typical verocay bodies are rare and narrow clefts which separated the nodules are prominent. In immunohistochemical study, mixture of S100 positive Schwann cells and axons are seen in palisaded neuroma, in contrast to purely S100 positivity in schwannoma.<sup>[14]</sup> It should be mentioned that intense staining for S100 protein supports the neural origin of the tumor cells in schwannoma which is seldom observed in leiomyoma.<sup>[4,14]</sup>

Reviewing these rare cases can be helpful for a better identification of such neoplasms, ruling out other differential diagnoses and prevention of over or underdiagnosis in similar cases.

### *Financial support and sponsorship*

Nil.

### *Conflicts of interest*

There are no conflicts of interest.

### *References*

1. Loannou M, Papanastassiou I, Iakowidou I, Kottakis S, Demertzis N. Plexiform schwannoma of the posterior tibial nerve: A case report. *Cases J* 2009;2:8392.
2. Berg JC, Scheithauer BW, Spinner RJ, Allen CM, Koutlas IG. Plexiform schwannoma: A clinicopathologic overview with emphasis on the head and neck region. *Human Pathol* 2008;39:633-40.
3. Plexiform Schwannoma [Internet]. DoveMed [updated 2016 Aug 27; cited 2016 Oct 07]. Available from: <http://www.dovemed.com/diseases-conditions/plexiform-schwannoma/>.
4. Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: Diagnostic overview and update on selected diagnostic problems. *Acta Neuropathol* 2012;123:295-319.
5. Megahed M. Plexiform schwannoma. *Am J Dermatopathol* 1994;16:288-93.
6. Friedman JM. Neurofibromatosis 1: Clinical manifestations and diagnostic criteria. *J Child Neurol* 2002;17:548-54.
7. Neurilemmoma [Internet]. Medscape [updated 2016 Aug 18; cited 2016 Oct 07]. Available from: <http://emedicine.medscape.com/article/1256405-overview>.
8. Wollina U, Langner D, Gruner M, Schönlebe J, Haroske G. Isolated plexiform schwannoma of the hand. *J Dermatol Case Rep* 2008;2:28-30.
9. Patterson JW. Neural and neuroendocrine tumors. In: Patterson JW, editor. *Weedon's skin pathology*. 4<sup>th</sup> ed.. London: Churchill Livingstone; 2016. p. 1041-67.
10. Woodruff JM, Selig AM, Crowley K, Allen PW. Schwannoma (Neurilemmoma) with Malignant Transformation A Rare, Distinctive Peripheral Nerve Tumor. *Am J Surg Pathol* 1994;18:882-95.
11. Nayler SJ, Leiman G, Omar T, Cooper K. Malignant transformation in a schwannoma. *Histopathology* 1996;29:189-92.
12. Peng X, Chen L, Du H, Lai Y, Li F, Zou X. Malignant transformation of benign intraosseous schwannoma in the cervical spine: A case report with an immunohistochemical study. *Int Surg* 2011;96:337-44.
13. Parihar A, Verma S, Suri T, Agarwal A, Bansal K, Gupta R. Plexiform schwannoma of lumbar region. *APSP J Case Rep* 2015;6:16.
14. Koutlas IG, Scheithauer BW. Palisaded encapsulated ("solitary circumscribed") neuroma of the oral cavity: A review of 55 cases. *Head Neck Pathol* 2010;4:15-26.