

Palisaded encapsulated neuroma: A unique presentation

Anup Kumar Tiwary, SS Chaudhary, DK Mishra, FirdousJahan

Department of Dermatology, Venereology & Leprosy, Rajendra Institute of Medical Sciences, Bariatu, Ranchi

Abstract Palisaded encapsulated neuroma (PEN) is a benign intraneural neuroma presenting as a solitary, sessile, immobile, asymptomatic, skin-coloured papule or nodule, commonly affecting the butterfly area of the face of a middle-aged person. It is not associated with neurofibromatosis or multiple endocrine neoplasia. We hereby report a unique case presenting with a firm, nodular growth over the posteromedial aspect of right knee which progressively increased in size over one year. On the basis of histopathologic findings, diagnosis of palisaded encapsulated neuroma was made. It is imperative to diagnose it histopathologically, because it is more commonly misdiagnosed clinically with schwannoma and neurofibroma.

Key words

Palisaded, encapsulated, neuroma.

Introduction

Palisaded encapsulated neuroma (PEN), also known as 'solitary circumscribed neuroma' is an uncommon, benign, cutaneous, neural tumor, which usually presents as small, solitary, asymptomatic, firm, skin-coloured or pink papule or nodule, usually localized on the face or close to a mucocutaneous junction. It is a disease of middle-aged adults with equal sex preponderance. It was first described by Reed *et al.* in 1972. Rarely, it can be multiple also but never associated with neurofibromatosis or multiple endocrine neoplasia.

Case Report

A 45-year-old male presented with a single, firm, flesh coloured, nodular growth on posteromedial aspect of right knee which began as a papule 1 year back and increased in size to 2 cm over one year. Initially it was asymptomatic but in last 3 months it became

painful with mild itching. None of the family members was affected.

On general physical examination, patient was healthy and normal. On local cutaneous examination, the lesion was solitary, firm, skin coloured, pedunculated, nodular growth, neither warm nor tender (**Figure 1**). There was no history of trauma, oozing or bleeding from the lesion or other cutaneous sites. Telangiectasia, ulceration or discharge was absent. Examination of mucosa, hairs, nails and teeth was also normal. Regional lymph nodes were not enlarged. Systemic examinations including ophthalmological and neurological examinations were also normal and none of the features of neurofibromatosis were detected. All routine investigations and laboratory parameters were within normal limits and noncontributory.

Excisional biopsy was performed and histopathological study was done. Histopathology demonstrated partially encapsulated, well-circumscribed dermal neoplasm that was differentiated towards nerves (**Figure 2**). It showed several nerve

Address for correspondence

Dr. Anup Kumar Tiwary
Department of Dermatology, Venereology & Leprosy
Rajendra Institute of Medical Sciences
Bariatu, Ranchi - 834009, India
Email: anup07tunnu07@gmail.com



Figure 1 Solitary, firm, skin coloured, nodular growth on posteromedial aspect of right knee.

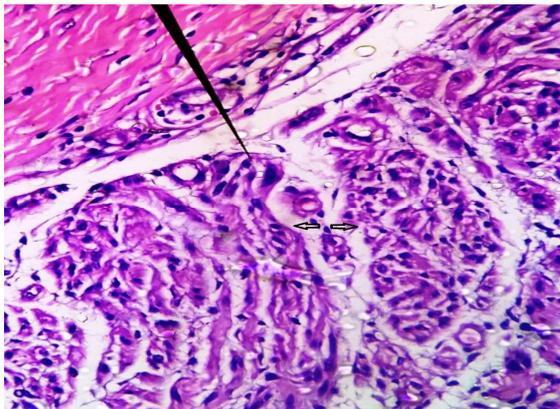


Figure 2 Partially encapsulated, well-circumscribed dermal nodules several nerve bundle like structures in the form of fascicles containing interdigitating spindle shaped schwann cells (H&E, x40).

bundles like structures or fascicles, some of which contained central acellular areas surrounded by an ill defined palisade of nuclei (Verocay bodies) [Figure 3]. Most of the cells were spindle shaped and the collagen was fibrillary. Increased number of small blood vessels was also seen in dermis.

Based on clinical features and histopathological findings, diagnosis of PEN was made. After doing excisional biopsy (Figure 4), wound was sutured with Mersilk 3-0 and patient was advised for follow up to look for any recurrence of the lesion.

Discussion

PEN or solitary circumscribed neuroma is a benign, cutaneous, dermal intraneural tumour presenting as solitary, firm, skin/pink coloured papule or nodule usually on the face in 90% of

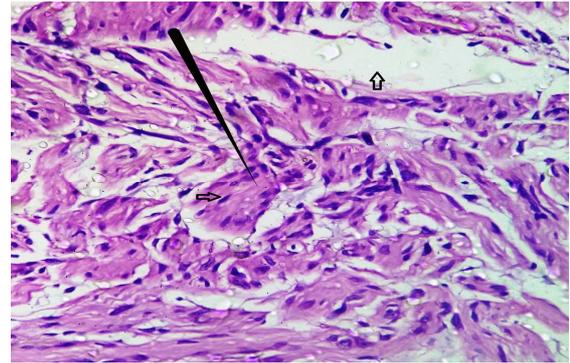


Figure 3 Spindle-shaped Schwann cells having eosinophilic cytoplasm, pointed and wavy basophilic nuclei and palisading of nuclei and acellular reticular fibres in between these nuclei forming 'verocay bodies' (H&E, X 100)



Figure 4 Wound after excisional biopsy.

cases and less commonly on trunk, shoulder, arm, hand, foot, oral mucosa, nasal fossa and glans penis. It accounts for 25% of all dermal nerve sheath tumours. There is no sex predilection and usually affects 30-60 years age group. Usually it measures between 2-6 mm in size and gradually enlarges over the time. Telangiectasia is minimal or absent. Hemorrhage and necrosis have not been reported but ulceration may be seen after trauma. No hair is seen coming out of the surface.

PEN is supposed to be caused by reactive or hamartomatous hyperplasia of Schwann cells and axons causing bulbous expansion of nerves but triggering factors are yet to be known. It is also considered as a hamartomatous growth but not neoplastic.

Clinically it can be misdiagnosed as schwannoma, neurofibroma,

appendageal tumours of skin, epidermal cyst, basal cell carcinoma and intradermal nevi. Relatively very low occurrence of telangiectasia and ulceration helps in ruling out the diagnosis of basal cell carcinoma. Neurofibroma, schwannoma and intradermal nevi simulate the clinical picture but can be ruled out histopathologically.

On histopathological demonstration, it is seen as intradermal nodule composed of encapsulated interlacing distinct fascicles containing interdigitating spindle cells. These fascicles are separated by artifactual clefts. The tumour has fine capsule or is partially encapsulated because it does not include epidermis. The capsule contains epithelial membrane antigen (EMA) positive flattened, elongated perineural cells. The tumour cells are composed of Schwann cells and axons. Schwann cells have eosinophilic cytoplasm, pointed and wavy basophilic nuclei and palisading of nuclei may also be seen in few fascicles. Palisaded nuclei and acellular reticular fibres in between these nuclei form Verocay bodies but are ill defined usually. Verocay bodies are more distinct and consistent finding in schwannoma rather than in PEN. Blood vessels can be seen occasionally. Apart from the nodular pattern of intradermal fascicles, other less common patterns have also been reported e.g. plexiform, epithelioid, multinodular, fungating and vascular.

On immunohistochemical analysis, capsule stains for EMA, and Schwann cells stain for S-100, collagen type IV and vimentin. Axons stain with neuron specific enolase and neurofilament proteins. Histopathologically the close differential diagnoses are schwannoma, traumatic neuroma, neurofibroma, leiomyoma. Usually schwannoma is located subcutaneously and has Antoni A and Antoni B areas. It is rich in

Verocay bodies and lack axons. Neurofibroma lacks capsule, rich in mucopolysaccharide ground substance and has fewer axons. Traumatic neuroma closely mimics the histological picture but can be ruled out by the presence of inflammatory cells and scarring. The presence of smooth muscle cells and positive staining for desmin, alpha smooth muscle actin and vimentin clinches the diagnosis of leiomyoma.

Treatment of choice is complete excision and chance of recurrence is very less even after incomplete excision.

References

1. Reed R, Fine R, Meltzer H. Palisaded encapsulated neuromas of the skin. *Arch Dermatol*. 1972;**106**:865-70.
2. Newman MD, Milgraum S. Palisaded Encapsulated Neuroma (PEN): An often misdiagnosed neural tumor. *Dermatol Online J*. 2008;**14 (7)**:12.
3. Golod O, Soriano T, Craft N. Palisaded encapsulated neuroma-A classic presentation of a commonly misdiagnosed neural tumor. *J Drugs Dermatol*. 2005;**4**:92-4.
4. Halder C, Sen S, Gangopadhyay A, Bala S. Zosteriform palisaded encapsulated neuroma: An unusual presentation. *Indian J Dermatol*. 2013;**58**:492.
5. Argenyi ZB. Immunohistochemical characterization of palisaded, encapsulated neuroma. *J Cutan Pathol*. 1990;**17**:329-35.
6. Dover J, From L, Lewis A. Palisaded encapsulated neuromas. A clinicopathologic study. *Arch Dermatol*. 1989;**125**:386-9.
7. Dubovy SR, Clark BJ. Palisaded encapsulated neuroma (solitary circumscribed neuroma of skin) of the eyelid: Report of two cases and review of the literature. *Br J Ophthalmol*. 2001;**85**:949-51.
8. Koutlas IG, Scheithauer BW. Palisaded encapsulated ("solitary circumscribed") neuroma of the oral cavity: A review of 55 cases. *Head Neck Pathol*. 2010;**4**:15-22.