Palisaded Encapsulated Neuroma: An Unusual Presentation

Abstract

Palisaded encapsulated neuroma (PEN) is a distinctive benign neural tumor, which usually presents as a solitary skin-colored papule or a nodule. It is a benign lesion and may clinically mimic a cutaneous neurofibroma, melanocytic nevi, or cutaneous schwannoma. Here we report a rare case of multifocal cutaneous PENs and its differentiation from cutaneous schwannomas, which is possible by histopathology.

Keywords: *Neurofibroma, palisaded palisaded encapsulated neuroma, schwannoma*

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Introduction

Palisaded encapsulated neuroma (PEN) is a distinctive benign neural tumor, which usually presents as small, solitary, asymptomatic, firm, rubbery, skin-colored papule or nodules.[1] It commonly occurs in middle-aged adults with female to male ratio of 1:1, rarely it can present with multiple lesions.[2] Clinically, PEN is most commonly confused with neurofibroma, schwannoma, or skin appendageal tumor.[1,3] PEN has histological appearance between that of a neurofibroma and a schwannoma. Differentiation from neurofibromas is essential, because the latter is often associated with neurofibromatosis, and has a propensity for malignant transformation. PEN has no association with neurofibromatosis or multiple endocrine neoplasia syndrome type 2B (MEN 2B). Even after incomplete excision PEN does not recur, a fact supporting a reactive rather than a neoplastic origin. Here, we present a rare case of multiple cutaneous PEN in a dermatomal distribution on the face not associated with neurofibromatosis or MEN 2B.

Case Report

A 35-year-old female patient presented to us with asymptomatic skin lesions on the face and back of 6 to 7 years duration. These lesions were initially pinhead sized and gradually grew to the present state. On examination, lesions were multiple, discrete, lobulated, flesh colored, soft to firm, papules and nodules arranged in a segmental pattern on the face [Figure 1] and the back [Figure 2] starting from the helix of the ear and extending up to the angle of mandible. There was no accompanying history of hearing defect or other neurological disorders. None of the family members were affected. On thorough cutaneous examination, there were no café au lait macules, axillary freckling, or plexiform neurofibromas. Hair, nails, and mucous membranes were normal. Ophthalmological examination was within normal limits (WNL). Routine blood investigations, X-ray, ultrasonography of the whole abdomen were normal. Histopathological examination revealed a normal epidermis. In the dermis, there was a partially encapsulated, well-circumscribed nodule [Figure 3] composed of fascicles of small spindle cells with wavy nuclei interlaced with small slits and clefts [Figure 4]. There was no nuclear pleiomorphism or mitoses. The tumor cells were positive for S100 protein. The tumor cells were also positive for neuron specific enolase and epithelial membrane antigen.

The findings and the clinical presentation were consistent with a diagnosis of PEN. We had sent the patient to the plastic surgery outpatient department for excision of the lesion.

Discussion

PEN of the skin was first described by Reed *et al*. in 1972.[1] It is an infrequent, benign cutaneous neural tumor, clinically

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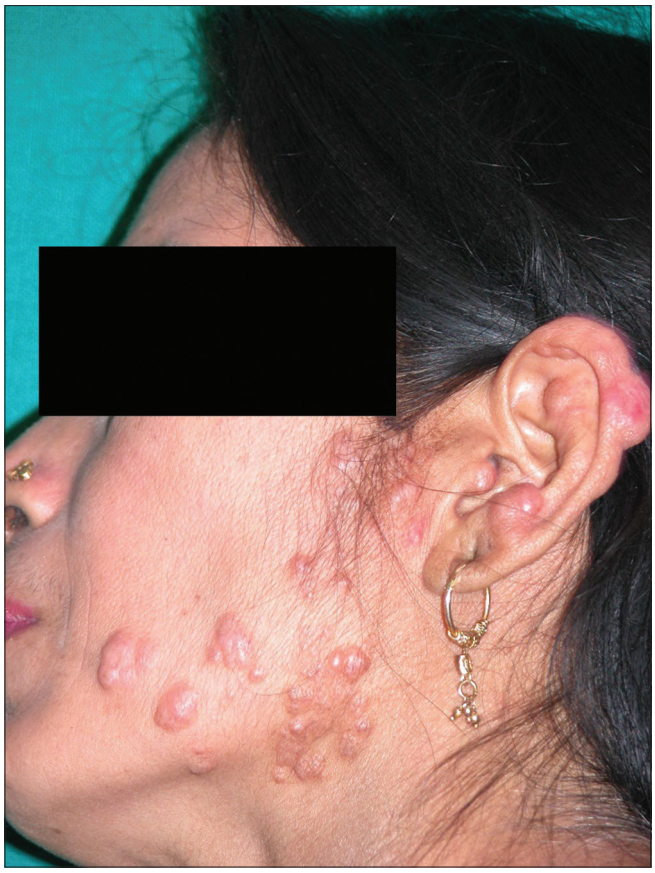

Figure 1: Multiple skin-colored discrete and coalescing papules and nodules on face extending from tip of ear to the angle of the mandible involving maxillary and mandibular divisions of the trigeminal nerve

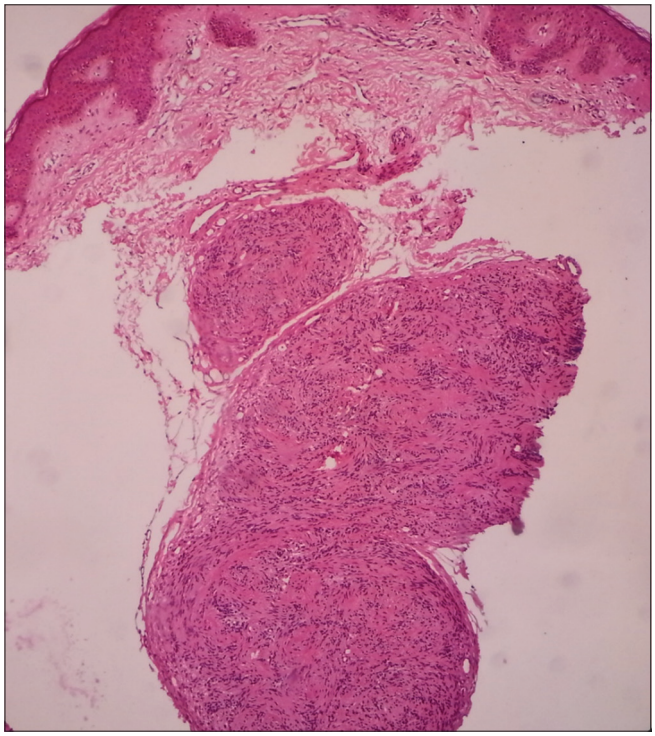

Figure 3: Partially encapsulated intradermal nodule composed of interdigitating spindle cells grouped in distinct fascicles (H and E, ×100)

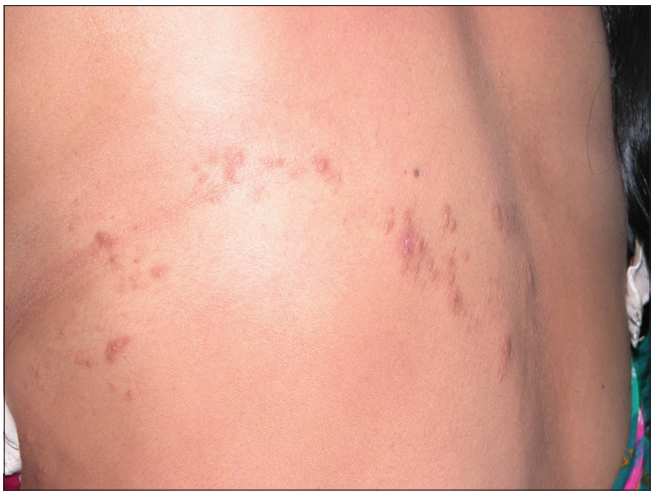

Figure 2: Similar skin-colored papules and nodules present over the back in a segmental pattern involving the T-7 dermatome

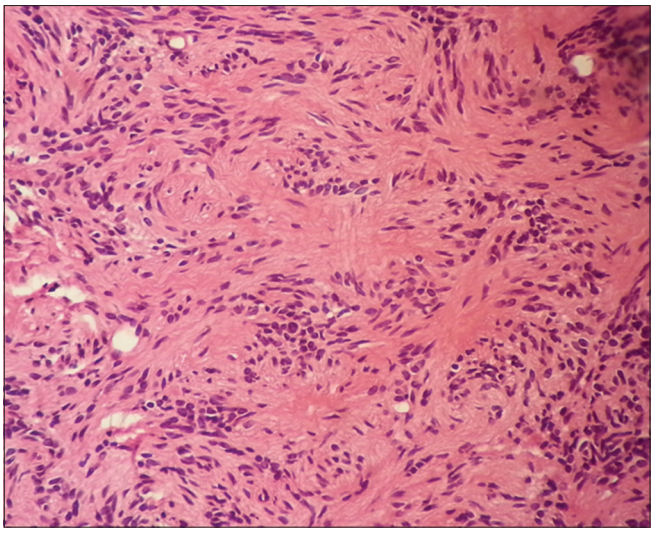

Figure 4: Spindle cells arranged in distinct fascicles. Individual cells are poorly delineated with eosinophilic cytoplasm, and pointed, wavy basophilic nuclei. There is lack of nuclear pleomorphism and mitoses (H and E, ×400)

misdiagnosed as a neurofibroma, basal cell carcinoma, melanocytic nevus, epidermal cyst, or skin appendage tumor, with a histological appearance between that of a neurofibroma and a schwannoma.

PEN clinically presents as solitary, asymptomatic, firm, sessile, immobile, rubbery, skin-colored or pink dome-shaped papule or nodule, commonly present on the face or close to a mucocutaneous junction.[1,4] The lesions have also been found on the trunk, shoulder, arm, oral mucosa, eyelid, and glans penis.[1,5] It is usually found in middle-aged adults with equal incidence in both sexes. The size of the lesion typically ranges between 2 mm and 6 mm. Rarely, it can present with multiple lesions.[5]

PEN has no association with neurofibromatosis or MEN 2B. Clinically, it is difficult to differentiate PEN from neurofibroma, though the lesions of PEN are commonly

present on the face, usually firm with absence of buttonhole sign.

Histopathologically, the tumors are well-circumscribed nodule and situated predominantly in the dermis. They are composed of a partially encapsulated mass of Schwann cells arranged in interlacing fascicles separated by small clefts, interspersed with a variable amount of tiny axons.[6] There is lack of nuclear pleiomorphism or mitoses. Although the nodular growth pattern is most frequently encountered, other types include epithelioid, plexiform, multinodular, and fungating.[7]

Immunohistochemical analyses reveal that tumor cells stain positively for S100 protein, collagen type IV, and vimentin characteristic of Schwann cell differentiation. The capsule is composed of elongated perineural cells that stain positively for epithelial membrane antigen.[8] The histological differential diagnosis for PEN includes neurofibroma, schwannoma, traumatic neuroma, and leiomyoma.[9]

Unlike PEN, neurofibromas lack a capsule, contain mucopolysaccharide ground substance, and have fewer axons with myelin sheaths.[6] Schwannomas are typically found subcutaneously, contain Antoni A and B type tissues with Verocay bodies, and do not have axons.[10] Overall, the PEN accounts for approximately 25% of all nerve sheath tumors of the dermis. True schwannomas rarely occur in the dermis. Traumatic neuromas have axonal and Schwann cell proliferation,; however, scarring and inflammatory cells are evident as well. Leiomyomas are spindle cell tumors composed of muscle cells readily differentiated by immunohistochemistry where they are consistently positive for desmin.

PEN is a benign tumor and recurs rarely. So patients can be reassured that excision is curative with a minimum possibility of recurrence, which reduces unnecessary further testing or concern to search for systemic diseases or malignancy. This case is a rare presentation of a multiple PEN that can be misdiagnosed.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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