SkIndia Quiz 4

Asymptomatic sessile skin colored nodule over columella of nose

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A 38-year-old male presented with an asymptomatic, skin-colored nodule over the columella of his nose since one year which had gradually increased to the present size. There was no history of trauma, oozing, crusting or similar lesions elsewhere, nor was there any family history of similar lesions. Examination revealed a single, skin-colored, soft, nontender, sessile nodule, measuring 1.5 × 1 cm in size over the left side of the nasal columella [Figure 1]. The rest of the cutaneous and systemic examination was within normal limits. An excision biopsy was performed [Figures 2 and 3].

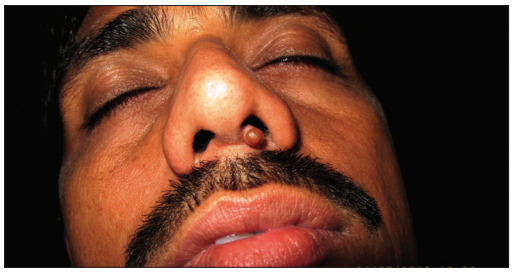

Figure 1: Soft, sessile, skin colored nodule over columella of nose

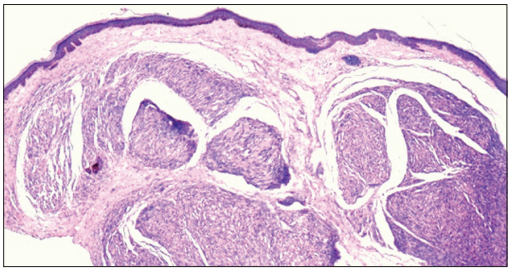

Figure 2: Well circumscribed poorly encapsulated multiple dermal nodules of spindled cells with prominent clefts (H and E, High Power)

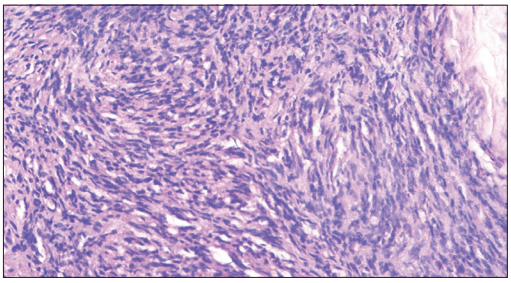

Figure 3: Spindled cells arranged in interlacing fascicles and nuclear palisades (H and E, Low Power)

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ANSWER

Diagnosis

Palisaded and Encapsulated Neuroma (PEN) — a multinodular variant.

DISCUSSION

Histological examination of the H and E-stained specimen revealed a normal epidermis and multiple partially encapsulated, well-circumscribed, dermal nodular proliferations of the cells, with delicate spindled nuclei, with prominent clefts at places and with partial fibrous capsules of flattened elongated cells [Figure 2]. On higher magnification the neoplasm was made up of proliferating Schwann's cells arranged in interlacing fascicles, with nuclear palisades characteristic of PEN [Figure 3]. No atypia or abnormal mitoses was seen. The biopsy wound had healed completely and no recurrence was reported within six months of follow-up.

Palisaded and encapsulated neuroma, also known as solitary circumscribed neuroma, is a benign proliferation of the peripheral nerve, usually solitary, although multiple lesions have been described. No malignant transformation or recurrence after complete removal has been reported.[1] Clinically it presents as non-characteristic, solitary or multiple nontender, skin-colored papule, mostly over the face and neck areas, rarely on the mucosa (oral, nasal or genital), trunk or proximal extremities, and more rarely on the acral parts.[1,2]

Palisaded and encapsulated neuroma is most commonly seen in adults, but can occur at any age and affects both sexes equally.[1] Regarding the origin of PEN, regeneration after local trauma has been suggested, but not found in many cases reported, including our case.[3]

It should be differentiated from dermal melanocytic nevus, basal cell carcinoma, and adnexal neoplasm clinically, and from traumatic neuroma, schwannoma, neurofibroma, and dermal melanocytic nevus with neural differentiation, as also from angioleiomyoma histologically.[3,4]

Histologically, PEN shows dermal nodules of Schwann cells with slender nuclei and fine chromatin, arranged in intersecting fascicles and separated by clefts with or without distinctive encapsulation and separated from the overlying epidermis.[1-3]

Correct diagnosis and differentiation from other neural neoplasms like schwannomas is important, as multiple schawannomas may be a manifestation of neurofibromatosis type 2.[1] Multiple mucosal neuromas and rarely multiple cutaneous neuromas may be associated with MEN type 2B (mutation in exon 16 in RET-proto-oncogene) or Cowden and Bannayan-Riley-Ruvalcaba syndromes (mutation in tumor suppressor gene PTEN).[2,1]

Apart from the classic nodular form, PEN has many unusual histopathological variants, such as, plexifrom, epitheloid, vascular, subcutaneous, multinodular, and fungating growth pattern, as also unusual features like fibrosis, myxoid change, chronic inflammation and acne-like changes.[1,3,5]

Immunohistochemically PEN shows strong S-100 protein, collagen type IV staining and variable staining for the myelin basic protein, Leu-7, neuron-specific enolase, while the capsule shows staining for the epithelial membrane antigen.[6] No immunohistochemistry was done in our case, due to nonavailability and the high cost to the patient.

Simple complete surgical excision is curative for a solitary lesion without recurrence,[1] while patients with multiple lesions should be investigated for the neurocutaneous syndrome, especially MEN type 2B.[2]

Our case highlights a multinodular variant, with partial encapsulation and prominent clefting, as also the importance of histopathology in correct diagnosis.

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