SkIndia Quiz 12
A firm yellowish nodule over lip

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A 26-year-old, otherwise healthy male presented with asymptomatic yellowish mass over lower lip. The lesion started 9 years back and increased gradually to the present size [Figure 1]. There was no history of any preceding trauma.

Examination revealed a firm, non-tender, yellow-colored nodule of 7 × 8 mm size over lower lip. Overlying mucosa was normal. No other skin or systemic abnormality was found in the patient who seemed healthy. Oral cavity examination was also normal. An excision biopsy was performed. Histopathology revealed a dermal tumor with S-100 protein positive cells [Figures 2–5].

WHAT IS THE DIAGNOSIS?

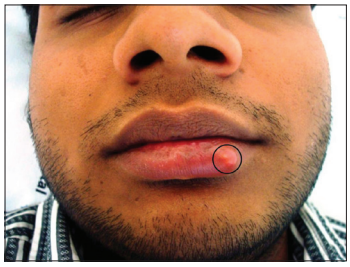

Figure 1: Yellowish firm submucosal nodule on lower lip

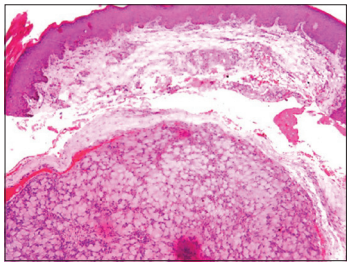

Figure 2: Biopsy showing well-encapsulated tumor situated within reticular dermis and subscutis. Most of the tumor is composed of rosetoid structures (H and E, 40×)

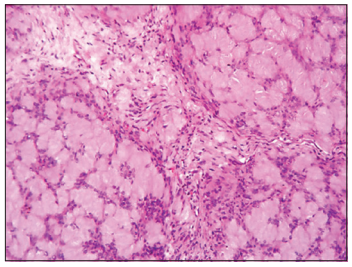

Figure 3: Striking rosette-like appearance with few spindle cells, the structures most closely resembled the Homer-Wright rosettes (H and E, 100×)

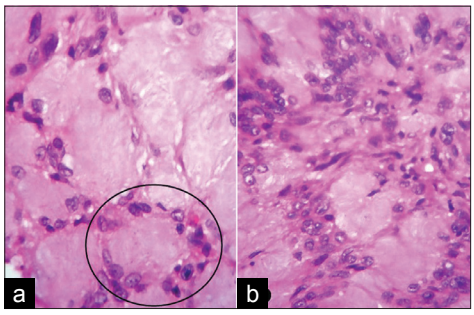

Figure 4: (a) Close-up of one of the rosettes (encircled) (H and E, 400×). (b) High-power view showing epithelioid morphology of cells (H and E, 400×)

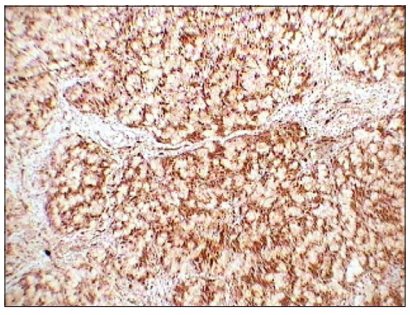

Figure 5: Immunohistochemistry showing uniform staining of cells forming rosettes with S-100 protein (immunohistochemistry, 100×)

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ANSWER

Benign epithelioid schwannoma (ES) with Homer-Wright rosette-like or neuroblastoma-like structures

Histopathology revealed normal epidermis with a well-circumscribed, symmetrical, and encapsulated mass within reticular dermis and extending to the subcutaneous tissue. The dermal mass was not connected to epidermis and there was clear grenz zone [Figure 2]. The dermal component was cellular with the predominant pattern being epithelioid cells arranged in nests to the periphery of centrally aggregated eosinophilic material, resulting in a striking rosette-like appearance [Figure 3]. No central cystic spaces, lumina, or vascular structures were present, and the structures most closely resembled the Homer-Wright rosettes [Figure 4a]. This florid rosette-like morphology was present throughout the thickness of the lesion. Cells throughout the lesions showed epithelioid morphology [Figure 4b]. No atypical mitoses or necrosis en mass was observed. All epithelioid cells showed diffuse staining for S-100 protein on immunohistochemistry [Figure 5]. Staining for smooth muscle actin was negative.

A diagnosis of benign ES of lip with Homer-Wright rosette-like or neuroblastoma-like structures was rendered.

DISCUSSION

Cutaneous schwannoma, a benign neoplasm of the peripheral nerve sheath, is classically composed of varying proportions of a highly ordered cellular component with nuclear palisading forming verocay bodies (Antoni A area) and a loose myxoid component (Antoni B area).[1] Pure epithelioid variant of benign cutaneous schwannoma is very rare.[2] Kindblom *et al*. were the first to report it in 1998.[3] Since then, very few cases have been reported. Also, many cases of neuroblastoma-like schwannoma with formation of rosettes have been described.[4] To our best knowledge, ES with Homer-Wright rosette-like or neuroblastoma-like structures has been reported only once.[5]

After the suggestion about existence of ES by Orosz *et al*. in 1993, Kindblom *et al*. reported a series of five cases with good documentation of this rare variant of common peripheral nerve sheath tumor, schwannoma.[2] Since then, very few cases of ES have been reported. Most of the reported cases are benign with female predominance, though malignant cases have also been reported.[6]

Goldblum *et al*. in 1994 reported three cases of schwannoma with perivascular rosettes and giant rosette-like structures, resembling the appearance of neuroblastoma, and termed them neuroblastoma-like neurilemmoma. Since then, very few cases of this extremely uncommon but distinctive histological variant of benign schwannoma have been reported.[7,8] Skelton *et al*. in 1994 reported a case of a schwannoma with collagenous spherulosis.[9] It was more or less similar to neuroblastoma-like schwannoma. Later, de Saint Aubain Somerhausen *et al*. described two further cases of neuroblastoma-like schwannoma,[10] and in 1998, Bhatnagar *et al*. published two new cases of schwannoma with immunohistochemical characterization showing areas that mimicked neuroblastoma/peripheral primitive neuroectodermal tumor.[11]

Many of these cases had epithelioid morphology in some foci. But pure epithelioid neuroblastoma-like schwannoma is reported only once by Fisher *et al*.[5]

It is important to recognize this distinct variant of schwannoma as both epithelioid morphology and rosette formation can cause confusion with many other tumors, especially malignant ones.[8] For example, ES may be confused with epithelial malignant peripheral nerve sheath tumor, myoepitheliomas, epithelioid fibromyxoid tumor of soft tissue, etc.,[2] and neuroblastoma-like schwannoma may be confused with neuroblastoma, fibromyxoid sarcoma, dendritic cell neurofibroma, etc.[4]

Rosette-like structures are rarely documented in melanocytic tumors also. They occur most frequently in the setting of primary and metastatic melanoma, but rarely in benign melanocytic tumors like Spitz nevus. The rosettes showing a peripheral rim of nuclei with a central aggregate of eosinophilic cytoplasm lacking a central lumen or vessel resembling most closely the Homer-Wright rosettes seen in neuroblastoma and peripheral neuroectodermal tumor are recently reported in Spitz nevus.[12]

The case presented here was remarkable for florid rosette formation affecting almost the entire lesion, including the deep aspect, resulting in a rather monotonous appearance and resembled most closely to a case of Spitz nevus with rosette-like structures.[12] Well-circumscribed, encapsulated, symmetric lesion with lack of any mitotic activity or atypia, vascular proliferation, and necrosis, as in the present case, should suggest benign nature.

Both rosette formation and epithelioid morphology are rare in schwannoma, and occurrence of both together in the same lesion as in the present case appears extremely rare.

Both dermatologist and pathologist should be aware of this rare variant of schwannoma to avoid overdiagnosis of malignant neoplasms.

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