Sir,
A 33-year old female presented with a history of painless swelling on her left cheek for 5 years. The lesion was excised twice but recurred on both occasions. There was no history of ulceration or discharge from the lesion or history of trauma at the site preceding the onset of the lesion. On examination, there were two erythematous nodules over the left cheek. The larger nodule measuring about 3 × 3 cm was present around 3 cm lateral to the left angle of the mouth [Figure 1]. The nodules were firm and non-tender. Lupus vulgaris, sarcoidosis, pseudolymphoma, Jessner’s lymphocytic infiltrate and lymphomatoid papulosis were the differentials considered.

Incisional biopsy was done and specimen was sent for histopathological examination and mycobacterial and fungal culture. Mantoux test, sputum for acid-fast bacilli and chest X-ray were non-contributory. Histopathological examination showed islands of epithelial cells covered with a basement membrane molding onto each other and separated by a thin stroma [Figures 2 and 3]. Periodic acid–Schiff (PAS) stain was done, which highlighted the acellular basement like material surrounding the islands [Figure 4].

Question
What is your diagnosis?

Figure 1: Erythematous firm non-tender nodules on the cheek

Figure 2: Mosaic-like masses separated by fibrous stroma (H and E 10X). Inset: Small basaloid cells in the periphery and large cells with pale nuclei in the center (H and E 40X)
Answer

Cylindroma.

Cylindromas are uncommon tumors arising from cutaneous adnexal structures. Female preponderance has been noted and may present clinically with single or multiple nodules. It is seen commonly over the scalp, other reported sites include face, trunk and upper limbs, notably only over the hairy areas and never over the palms and soles. They are also known as “turban tumors” as multiple lesions can cover large portions of the scalp like a turban. The term “cylindroma” was first used by Billroth in 1856 in accordance with the cylindrical outline seen in its transverse section. Three categories of cylindromas identified are:
1. Benign cylindroma, which may be single or multiple
2. Malignant cylindroma as part of Brooke–Spiegler syndrome (BSS), which is usually multiple in number with an earlier age of onset
3. Malignant salivary cylindroma, otherwise known as cystic adenoid carcinoma.

Clinically, cylindromas present as slow-growing, pink to red, firm nodules of size ranging from 0.5 to 6 cm. The surface can be smooth or ulcerated, mimicking malignancy. The origin of cylindroma is still a matter of debate as it has not been proven beyond doubt whether its origin is from the eccrine glands, apocrine glands, or follicular epithelium. Thus, the cell of origin has often been described as pluripotent stem cells in the folliculo-sebaceous-apocrine unit.

BSS results from a mutation in tumor suppressor gene CYLD located on chromosome 16q12-q13 and is characterized by the presence of different skin tumors like multiple cylindromas, trichoepitheliomas and eccrine spiradenomas. It has an autosomal dominant inheritance and a wide phenotypic variability.

Histopathology of cylindroma is characteristic with a “jigsaw puzzle” appearance comprising of irregular islands of basaloid cells surrounded by hyaline eosinophilic sheath, which is PAS-positive. The isles have two types of cells: undifferentiated epithelial cells with small and dark nucleus in periphery in palisading fashion, and more differentiated cells with larger and paler nucleus showing ductal differentiation. Immunohistochemistry has consistently shown p63 positivity and CD15 negativity. The thickened basal membrane in cylindroma has multiple ultrastructural alterations, including absence of mature hemidesmosomes, defects in laminin 5, reduced expression of integrin α6β4 expression, and increased expression of integrin α2β1. In addition, defects in expression of α11 and α5 chains of collagen IV have also been documented. Malignant cylindromas differ histopathologically as they lack the typical pattern, biphasic cellular distribution, and surrounding hyaline sheath. Cylindromas can be differentiated histopathologically from other hair follicular tumors like trichoepitheliomas with their characteristic morphology and presence of surrounding hyaline eosinophilic sheath.

Malignant transformation is uncommon with solitary, sporadic cylindromas. The treatment of choice is surgical excision of the tumor, and recurrence may rarely occur after radiofrequency excision, while it is unlikely after excision with a scalpel. Recurrence in the present case might be because of incomplete excisions initially.

Although cylindromas are classically seen over the scalp, it should be included in the list of differential diagnoses of painless slow-growing nodules over the face.

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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Nil.
Conflicts of interest

There are no conflicts of interest.

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