Quiz

Multiple hypopigmented macules on the face

A 48-year-old man presented with a 10-year history of asymptomatic, hypopigmented lesions on the face which were initially small and gradually increased in number and size. Physical examination revealed approximately twenty, similarly sized, round to angulated, hypopigmented macules on the beard area and neck [Figure 1a and b]. The clinical differential diagnoses included pityriasis versicolor, pityriasis alba, idiopathic guttate hypomelanosis, progressive macular hypomelanosis and epidermodysplasia verruciformis. Skin biopsy from one of the lesions revealed dilated follicular infundibula connected to each other and the overlying epidermis by a broad plate-like proliferation of cells that resembled isthmic epithelium with pale pink cytoplasm [Figures 2 and 3].

WHAT IS YOUR DIAGNOSIS?

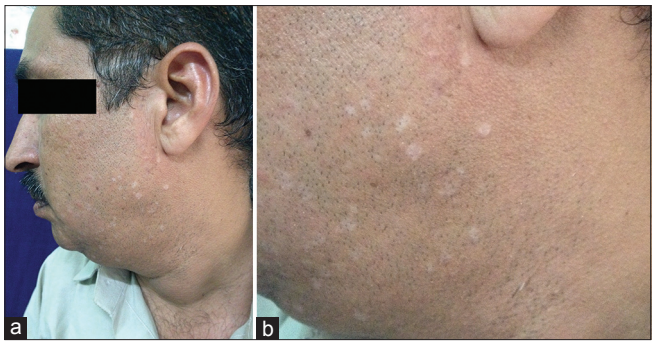

Figure 1: (a) Multiple, discrete, round to angulated, hypopigmented macules in the beard area. (b) Close up view of hypopigmented macules

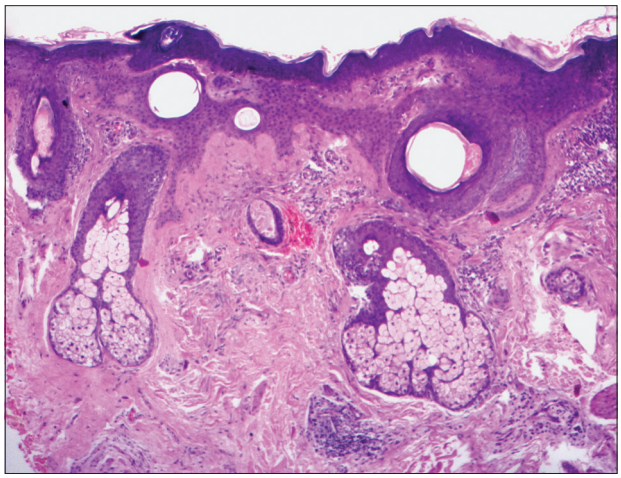

Figure 2: Dilated follicular infundibula connected to each other (H and E, ×10)

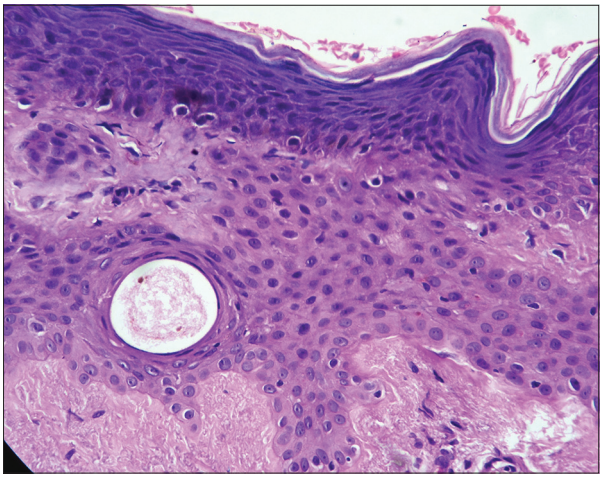

Figure 3: Broad plate-like proliferation of cells that resemble isthmic epithelium with pale pink cytoplasm (H and E, ×40)

Access this article online	
Quick Response Code:	Website: www.ijdvl.com
	DOI: 10.4103/0378-6323.184201

How to cite this article: Mittal A, Agrawal C, Balai M, Joshi R. Multiple hypopigmented macules on the face. Indian J Dermatol Venereol Leprol 2016;82:585-6.

Received: August, 2015. **Accepted:** December, 2015.

ANSWER

Follicular infundibular tumor.

DISCUSSION

Tumor of the follicular infundibulum, described by Mehregan and Butler, represents an uncommon benign adnexal tumor,.[1] It usually presents as a solitary lesion on the face, scalp, neck or upper trunk. Other variants are the eruptive variant and those associated with Cowden's disease and other tumors or hamartomas, such as nevus sebaceous.[2] Among its various presentations, only the eruptive tumors can be identified clinically. They consist of asymptomatic, hypopigmented and irregularly-shaped macules located on the face, neck and upper trunk. The lesions are usually symmetrically distributed and progressively increase in number over many years. Their number ranges from fewer than 20 to more than 100.[3] The hypopigmented macular lesions mimic vitiligo, pityriasis alba, pityriasis versicolor, post-inflammatory hypopigmentation, idiopathic guttate hypomelanosis and tuberculoid leprosy.[1,3]

The histopathological diagnostic criteria for tumor of the follicular infundibulum as listed by Ackerman are (1) a distinctive silhouette with a horizontal proliferation of keratinocytes, (2) characteristic neoplastic epithelial cells with small monomorphic nuclei and abundant pink cytoplasm and (3) thin columns and bulkier aggregations of cells, all of which are interconnected. The well-developed lesions may show peripheral palisading.[1,4] Two other distinctive features are glycogen in the cells which can be stained by periodic acid–Schiff (PAS) stain and a brush-like network of elastic fibers which are identified on van Gieson stain at the border of the tumor.[4] This can distinguish it from basal cell carcinoma and seborrheic keratosis which lack this feature.[1] Further, immunostaining with Ber-EP4 (positive in basal cell carcinoma) can be used for differentiating difficult cases.[1]

Several treatments have been suggested such as topical keratolytics, topical steroids, long-term etretinate, cryotherapy, curettage and excision; however, none gives satisfactory result.[3]

It is difficult to clinically diagnose tumor of the follicular infundibulum and a skin biopsy is essential for the diagnosis. Although these are benign proliferations, Schnitzler *et al.*, in their study of 100 patients with tumor of the follicular infundibulum, found transformation of two of these tumors to basal cell carcinoma; therefore, a regular monitoring of these patches is advisable.[5]

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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