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Editorial

Bilateral nevus comedonicus of the eyelids: An unusual cause of ptosis and ectropion

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Dear editor,

In their article entitled " Bilateral nevus comedonicus of the eyelids: An unusual cause of ptosis and ectropion",[1] which was published recently in your distinguished journal, we strongly believe the authors misdiagnosed the patient. What the article and the attendant figure show is a classic textbook example of Favre-Racouchot syndrome (FRS) rather than nevus comedonicus (NC). Although FRS is a ubiquitous condition that predominantly affects the periocular region, it is unfortunate that the condition is under-appreciated in the ophthalmic literature, which may have led some authors to confuse FRS with the far less prevalent but far more popular nevus comedonicus.

With only around 200 cases of nevus comedonicus described in the literature, the condition is extremely rare, and usually manifests at birth or before the age of 10 years.[2–5] Although acquired cases of NC have been described, these lesions are typically hereditary.[2–5] The exact etiology is unknown, however, it is generally categorized as a congenital hair follicle deformity (a hamartomatous malformation caused by an imperfect development of the folliculosebaceous unit), caused by somatic mutations in the NEK9 gene (OMIM # 617025, *14q24.3*), a gene which is believed to be involved in follicular homeostasis.[5,6] Abnormalities in FGFR2 signaling (OMIM # 176943, *10q26.13*) have also been implicated in comedogenesis in NC patients.[6,7] The condition is characterized by the aggregation of dilated follicular orifices filled with keratinous material (open and closed comedones) which are usually grouped in linear streaks or distinct bands. These lesions generally follow Blaschko's lines which are the lines that are thought to map the pattern of migration of epidermal cells during fetal development (Fig. 1A and B).[2–4,8] NC is typically unilateral, although bilateral cases have been recorded but this is indeed rare.[2]

On the other hand, Favre-Racouchot syndrome (nodular elastosis with cysts and comedones) is a fairly common cutaneous disorder which is largely predisposed to by prolonged sun-exposure and ultraviolet damage (UV–B, or UV-A_1), smoking, radiation therapy, and topical or systemic steroids.[9,10] With an overall prevalence of 1.4% in adults aged between 25 and 74 years, a 6% prevalence in patients aged between 40 and 60 years, and a prevalence of 2.5% among agricultural workers,[9,11,12] Favre-Racouchot syndrome is a ubiquitous condition and is far more common than nevus comedonicus which is indeed rare with a prevalence of 1/45,000–1/100,000.[13] The onset of Favre-Racouchot syndrome is usually delayed (middle-old age), and the condition is typically bilateral, although the distribution may be asymmetric if one side of the face is unevenly exposed to the sun.[5,9,10]

Although multiple comedones are a feature of both conditions, the typical appearance in patients with FRS is the presence of bilateral discrete follicular orifices plugged by keratotic material, which usually occur in groups or clusters of variably sized nodules that have an elevated margin and an eccentric opening or pore that is predominantly filled with black material (blackheads).[9,10] The comedones in Favre-Racouchot syndrome do not follow Blaschko's lines, but they have a predilection for the lateral canthal region, and the malar eminence is usually involved as well. The surrounding skin usually shows classic unmistakable signs of severe actinic damage (yellowish skin discoloration, variable pigmentation, and severe wrinkling) which is an essential feature of the condition (Fig. 1C and D).[2,5,9,10] Signs of actinic damage are not a feature of nevus comedonicus which is a hereditary disorder.[2]

Both conditions can be easily differentiated from each other by the age of onset, by the typical arrangement of nevus comedonicus lesions in a linear streak pattern, which in its classical form in the periocular region has a striking appearance,[14] as well as by the physical appearance of the facial skin in patients with Favre-Racouchot syndrome.[2,5,9,10] The age of presentation of the patient in question strongly argues against a diagnosis of nevus comedonicus. In addition, the accompanying figure shows classic unambiguous signs of severe actinic damage, amidst which the comedo lesions are distributed, and they do not follow any particular linear pattern at all. The peculiar facial features of the Favre-Racouchot syndrome are so unique that is difficult to misdiagnose a patient with this condition,[10] yet both conditions are nearly identical histopathologically except for the presence of evidence of solar elastosis in FRS

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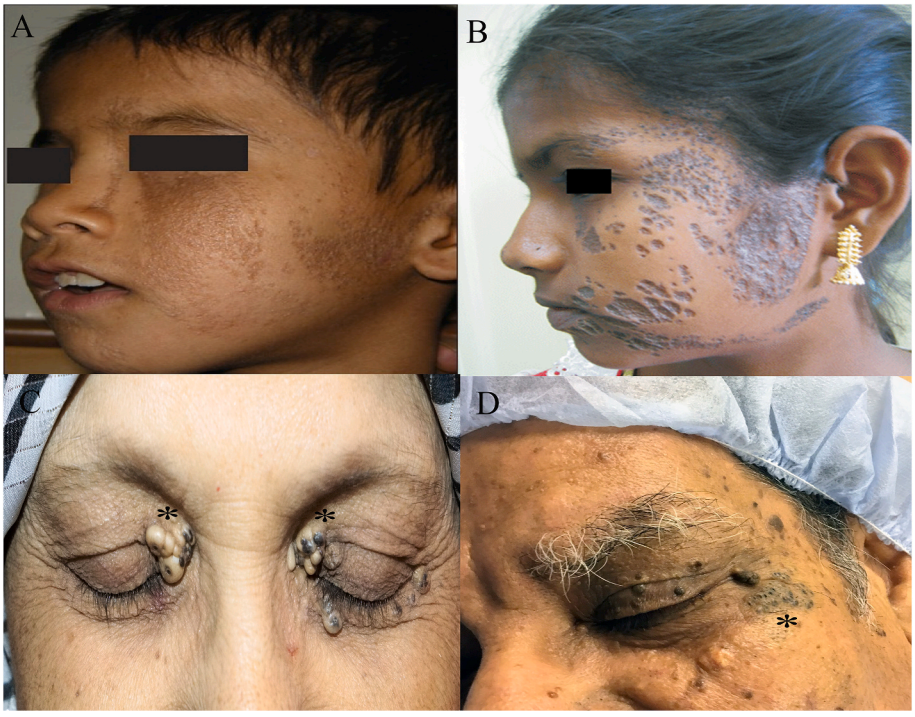

Fig. 1. A,B Nevus comedonicus syndrome. Unilateral dilated follicular orifices filled with keratinous material which are usually grouped in linear streaks or distinct bands (C,D). Nevus comedonicus may develop a progressive or inflammatory course and may result in extensive scarring but comedones may still be observed intermingled with the honeycomb-like pitted scars (D). Signs of skin ageing or solar damage are absent. **C,D. Favre-Racouchot syndrome**. Bilateral comedones (blackheads), plugged by keratotic material are typically observed in groups or clusters in the infrabrow region (C), or the lateral canthal region (D). Signs of ageing and actinic damage can also be observed (skin wrinkling, acrochordons and seborrheic keratosis). A & B are reprinted with permission from the Indian Journal of Dermatology,[14] and Indian Pediatrics.[16]

patients.[15] This histopathologic mimicry could be the reason why the authors failed to diagnose the patient correctly. It is regrettable that ophthalmologists are not very familiar with a condition as pervasive as the Favre-Racouchot syndrome despite its predilection to the periorbital region.

Financial interest

The authors have no financial interest in the study.

Declaration of competing interest

The authors of the present manuscript hereby declare they have no conflict of interest or any financial interest in this publication.

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