A Rare Association in Down Syndrome: Milialike Idiopathic Calcinosis Cutis and Palpebral Syringoma

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PRACTICE **POINTS**

- Down syndrome is associated with rare dermatological disorders and an increased prevalence of common dermatoses.
- It is important to differentiate milialike idiopathic calcinosis cutis from other dermatological diseases using histopathology and dermoscopy.

To the Editor:

Down syndrome (DS) is associated with rare dermatological disorders, and the prevalence of some common dermatoses is greater in patients with DS. We report a case of milialike idiopathic calcinosis cutis (MICC) associated with syringomas in a patient with DS. We emphasize that MICC is one of the rare dermatoses associated with DS.

A 4-year-old girl with DS presented with a 4-mm, flesh-colored, regular-bordered, exophytic papular lesion on the left upper eyelid of 8 months' duration (Figure 1). It was clinically recognized as syringoma. On dermatologic examination of the patient, there also were 1- to 3-mm, round, whitish, painless, milialike papules on the dorsal surface of the hands and wrists (Figure 2). Some of these papules were surrounded by erythema. There was no sign of perforation. Her personal and family history were unremarkable.

Histopathologic examination of a biopsy from a milialike lesion on the hand showed a hyperkeratotic epidermis. In the dermis there was a roundish calcific nodule surrounded by a fibrovascular rim. The patient's guardians refused a biopsy from the lesion on the eyelid.

Laboratory tests including serum vitamin D, thyroid and parathyroid hormone, calcium, phosphorus, and urinary calcium levels, as well as renal function tests, were within reference range. On the basis of these clinical and histopathological

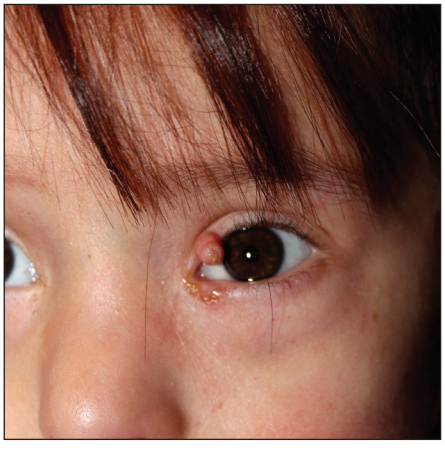

Figure 1. A 4-mm exophytic papular lesion on the left upper eyelid.

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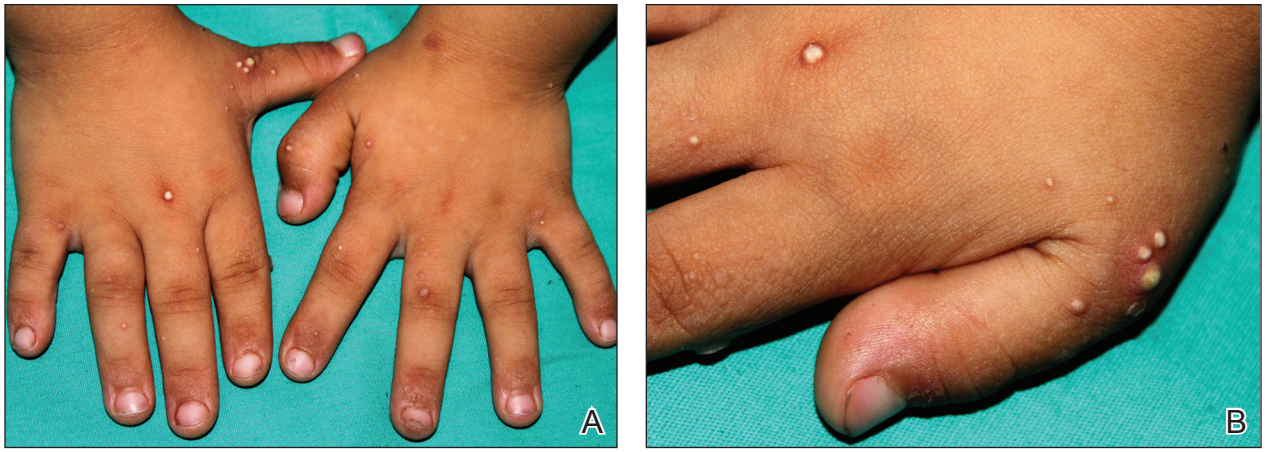

Figure 2. Round whitish papules on the dorsal aspects of the hands and wrists (A) and milialike papules on the dorsal aspect of the hand (B).

findings, the patient was diagnosed with MICC and palpebral syringoma.

Many dermatoses associated with DS have been reported including elastosis perforans serpiginosa, alopecia areata, and syringomas.[1-3] Sano et al[4] first described MICC and syringomas in a patient with DS in 1978. Milialike idiopathic calcinosis cutis is characterized by asymptomatic, millimetric, firm, round, whitish papules that are sometimes surrounded by erythema. These papules may show perforation leading to transepidermal elimination of calcium, similar to the transdermal elimination of elastic fibrils in elastosis perforans serpiginosa. Although MICC usually is described in acral sites of children with DS, it also is reported in adults without DS and on other parts of the body.[5-7]

The cause of MICC is unknown. One hypothesis of the development of MICC is an increase of the calcium content in the sweat leading to calcification of the acrosyringium.[8] Milia are small keratin cysts that usually develop by occlusion of the hair follicle, sweat duct, or sebaceous duct. However, milia also can occur from occlusion of the eccrine ducts where syringomas originate.[9] Therefore, syringomas can be seen in association with milia and calcium deposits.[5,9-11]

We believe that MICC in DS may be more common than usually recognized, as the lesions often are asymptomatic. It is important to differentiate MICC from other dermatological diseases such as molluscum contagiosum, verruca plana, milia, and inclusion cysts. Histopathology and dermoscopy could aid in the accurate diagnosis of MICC.

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