Milia-like calcinosis cutis in a girl with Down syndrome*

Berna Solak[1]
Erdem Vargol[2]
Rabia Oztas Kara[1]

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Abstract: Milia-like idiopathic calcinosis cutis (MICC) is a very rare dermatological disorder characterized by multiple whitish to skin colored, milia-like papules, mostly found on the hands. MICC can disappear spontaneously by adulthood; therefore, its early recognition is crucial to avoiding unnecessary interventions. Herein, we present a case of MICC in a 6-year-old girl with Down syndrome.

Keywords: Calcinosis; Molluscum contagiosum; Skin diseases

INTRODUCTION

Milia-like idiopathic calcinosis cutis (MICC) is a very rare dermatological disorder characterized by multiple whitish to skin colored, firm, tiny milia-like papules, mostly on the hands and feet. MICC was first reported in 1978.[1,2] Since then, to the best of our knowledge, only about 25 cases have been reported in English language medical literature, mostly in association with Down Syndrome.[1] Herein, we present this rare case study to help the busy dermatologist or pediatrician to recognize this disorder in his/her differential diagnoses of milia-like lesions.

CASE REPORT

A 6-year-old girl was received at our medical center complaining of multiple asymptomatic, 1–2 mm in diameter, whitish papules on her hands, feet, and face. The patient reported that the papules on her hands and feet had been present for three years, while those on her face had first appeared last year, and that these lesions were increasing (Figures 1A and 1B). Her mother denied any previous trauma or wounds at the sites of the lesions. The patient presented trisomy 21 (Down syndrome), hypothyroidism, grade III vesicoureteral reflux, as well as atrial and ventricular septal defects. As treatment, she was taking levothyroxsine (Levotiron®). Dermoscopic appearance did not show typical changes associated with MICC (Figure 1C). Skin biopsies of the lesions revealed an accumulation of well-defined, dense, basophilic material surrounded by fibrous tissue in the dermis (Figure 1D). Biochemical studies, including serum calcium, phosphate, and parathyroid hormone levels, were within the normal range. Based on pathological and clinical findings, the patient was diagnosed with MICC.

DISCUSSION

MICC is a form of idiopathic calcinosis cutis, which implies an abnormal deposition of insoluble calcium salts in the dermis. MICC's exact etiology and pathogenesis is unknown; however, serum calcium and phosphorus levels do remain normal, unlike met-

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[1] Sakarya University, School of Medicine – Sakarya, Turkey.
[2] Sakarya University Training and Research Hospital – Sakarya, Turkey.

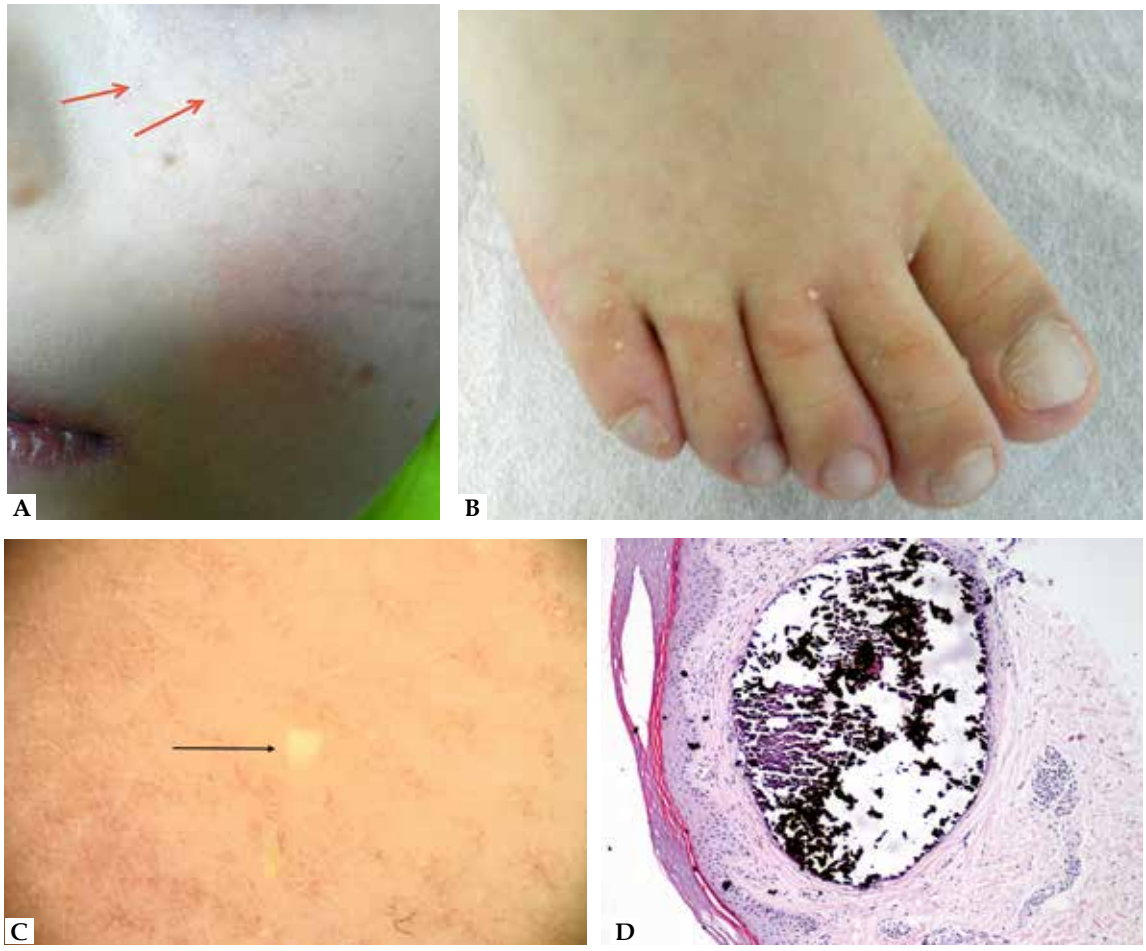

Figure 1: A. Whitish papules on the face (red arrow); **B.** Whitish papules on the foot ; **C.** Dermoscopic appearance of the lesion (black arrow); **D.** Accumulation of well-defined, dense, basophilic material surrounded by fibrous tissue in the dermis (100x H&E)

astatic or iatrogenic calcinosis.[3,4] This disorder is equally frequent in both sexes and most commonly occurs during childhood.[1] Though lesions are frequently found on the hands and feet, they may also appear on other parts of the body. MICC normally disappears spontaneously by adulthood, mostly without scarring,[4] and has mostly been reported as associated with Down syndrome.[1] Although the pathogenesis of MICC is unclear, medical studies have shown that patients with Down syndrome have ectopic calcifications, such as valvular and basal ganglial calcifications.[5,6] MICC has also been reported to occur in patients without Down syndrome. It can therefore be suggested that other unknown factors may play important roles in its pathogenesis. Although not proven clearly, some authors have suggested that sweat glands may play a role in calcium deposition by means of decreased excretion or increased sweat calcium concentrations.[7,8] Moreover, it is thought that recurrent, minor traumas may in fact trigger this disorder.[9]

Distinguishing MICC from molluscum, milium, verruca, xanthomas, or inclusion cysts is essential. Diagnosis is based on histopathological, clinical, and laboratory findings. Histopathological examination reveals foci of a well-circumscribed, round, basophilic substance, which is stained black with Von Kossa stain, in the upper dermis, surrounded by thick collagen fibers, and sometimes, epithelioid and multinucleated giant cells.[4,9]

Dermoscopy is commonly proposed as a helpful method for differential diagnosis.[10] Recently, Fox *et al.*[1], upon dermoscopic examination, described a subtle petaloid appearance, though not pathognomonic for MICC. The dermoscopic examination of our patient's lesions proved unremarkable, demonstrating a homogenous, bright white, round area which could not be distinguished from milium or molluscum.

Since this disorder can disappear spontaneously by adulthood, unnecessary aggressive interventions are not recommended. On the other hand, tretinoin cream was found useful by Choi *et al.*[4]

Because of the possibility of spontaneous recovery, watchful follow-up may be the best approach. Since diseases in the differential diagnosis of MICC, i.e. molluscum, verruca, and milium, are usually treated with traumatic methods, such as cryotherapy and cauterization, it is important to distinguish MICC from these disorders, as these therapeutic modalities may serve only to worsen the lesions and stimulate recurrence. ❑

REFERENCES

1. Fox GN, Mehregan DA, Jablonowski MN. Acral milia-like idiopathic calcinosis cutis in a child with Down syndrome: report of a case, review of the literature, and description of dermoscopic findings. Pediatr Dermatol. 2013;30:263-4.
2. Sano T TS, Ishikawa C. A case of Down's syndrome associated with syringoma, milia, and subepidermal nodule. Jpn J Dermatol. 1978;88:740.
3. Houtappel M, Leguit R, Sigurdsson V. Milia-like idiopathic calcinosis cutis in an adult without Down's syndrome. J Dermatol Case Rep. 2007;1:16-9.
4. Jang EJ, Lee JY, Yoon TY. Milia-like idiopathic calcinosis cutis occurring in a toddler born as a premature baby. Ann Dermatol. 2011;23:490-2.
5. Hamada T, Kuroda M, Miyakoshi M, Koshino Y, Murata T, Oomori M, et al. [Echocardiographic study in adult patients with Down's syndrome]. Rinsho Byori. 1993;41:807-12.
6. Takashima S, Becker LE. Basal ganglia calcification in Down's syndrome. J Neurol Neurosurg Psychiatry. 1985;48:61-4.
7. Maroon M, Tyler W, Marks VJ. Calcinosis cutis associated with syringomas: a transepidermal elimination disorder in a patient with Down syndrome. J Am Acad Dermatol. 1990;23:372-5.
8. Sais G, Jucglà A, Moreno A, Peyrí J. Milia-like idiopathic calcinosis cutis and multiple connective tissue nevi in a patient with Down syndrome. J Am Acad Dermatol. 1995;32:129-30.
9. Bécuwe C, Roth B, Villedieu MH, Chouvet B, Kanitakis J, Claudy A.. Milia-like idiopathic calcinosis cutis. Pediatr Dermatol. 2004;21:483-5.
10. Strumia R. Videodermatoscopy: a useful tool for diagnosing cutaneous dystrophic calcifications. Dermatol Online J. 2005;11:28.

Mailing address:
Berna Solak
Sakarya Universitesi,
Tıp Fakültesi,
Dermatoloji Ana Bilim Dalı
54000, Sakarya, Turkey
E-mail: bernasolakmd@gmail.com

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