

## CASE REPORT

# Milia-like Idiopathic Calcinosis Cutis Occurring in a Toddler Born as a Premature Baby

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Milia-like idiopathic calcinosis cutis (MICC) is characterized by smooth, firm, whitish papules resembling milia. Histologically, it appears as a well-defined, round, basophilic nodule within the upper dermis. Although the etiology and treatment remain unclear, it may resolve spontaneously. Some cases have been associated with Down syndrome, and the mean age of MICC patients was 9.9 years old. Herein, we report a rare case of MICC that was not associated with Down syndrome. Noticeably, the patient, a toddler, was born as a premature baby and had an ischemic injury on the right foot at birth. However, the lesions appeared on both feet, including the non-injured left foot. Otherwise he was healthy. After a 21-month follow-up period, the lesions had almost disappeared without any treatment. (**Ann Dermatol 23(4) 490~492, 2011**)

**-Keywords-**

Calcinosis cutis, Prematurity

## INTRODUCTION

Milia-like idiopathic calcinosis cutis (MICC) is a peculiar subtype of idiopathic calcinosis cutis. The lesions appear as multiple, round, whitish papules resembling milia.

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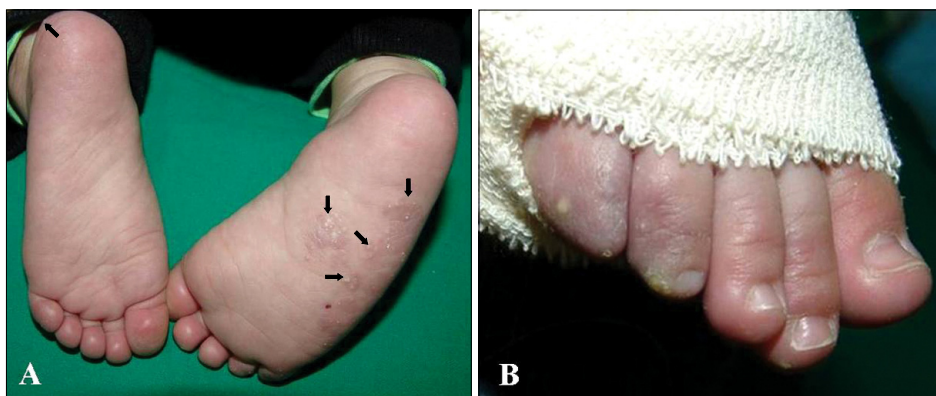
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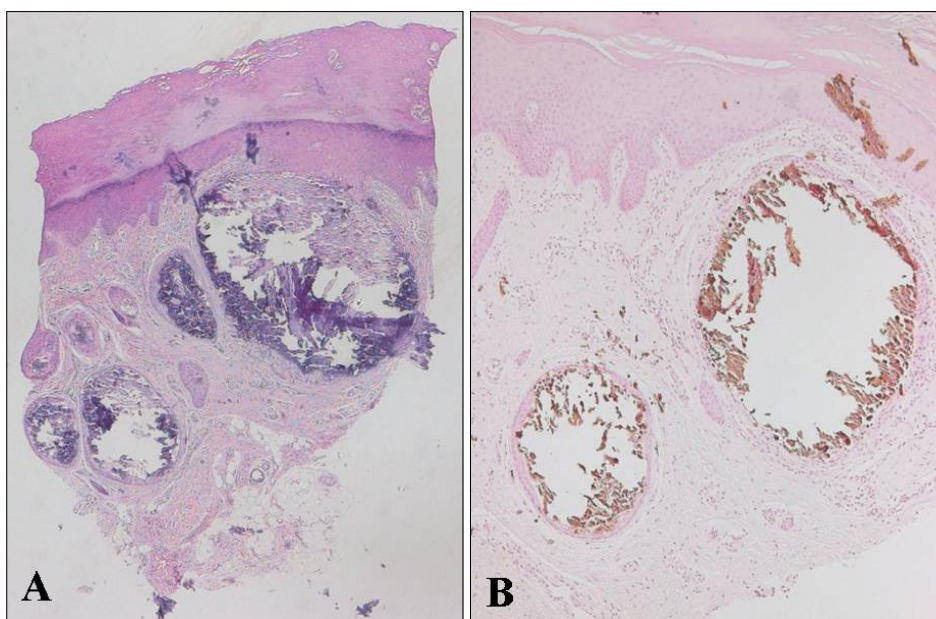
Since its first description by Sano et al.<sup>1</sup> in 1978, about 23 cases have been reported in the English and Korean literature so far, and the mean age of the patients was 9.9 years old<sup>2,4</sup>. In some cases, MICC was associated with Down syndrome. We report a rare case of MICC that occurred in a toddler who did not have any evidence of Down syndrome. He was born as a premature baby and had an ischemic injury on the right foot at birth. However, the lesions occurred on both the injured right foot as well as the non-injured left foot.

## CASE REPORT

A 19-month-old toddler presented with a 3-month history of multiple whitish papules on both feet. At the time of the patient's arrival to the clinic, the lesions had gradually increased in number. He was born at 26 weeks' gestation as a premature baby. His parents noted a history of ischemic injury on the right foot at birth. Otherwise, the patient had no problems at birth. Physical examination revealed multiple, firm, 1 to 4 mm sized, whitish papules on the right sole, 5th toe, and left heel (Fig. 1). Histopathologic examination of the papules from both feet revealed small nodular foci of basophilic materials surrounded by epithelioid structures within the upper dermis resembling a pseudocyst and focal transepidermal elimination of Basophilic materials (Fig. 2A). Von Kossa stain confirmed that the materials were calcium (Fig. 2B). Immuno-Histochemical staining for carcinoembryonic antigen (CEA) of epithelioid structures around the deposited materials was negative. Laboratory findings, including serum calcium and Phosphorus Levels, were within normal limits. With all those findings, the diagnosis of MICC was made. After a 21-month follow-up period, the lesions had almost disappeared without any treatment.



**Fig. 1.** (A) Multiple, firm, 1~4 mm sized, whitish papules on the right sole, left heel (arrows), and (B) right 5th toe. Morphologic changes on the right 4th and 5th toes may be caused by ischemic injury at birth.



**Fig. 2.** (A) Several focal amorphous basophilic materials in the upper dermis and focal transepidermal elimination of the basophilic materials (H&E,  $\times 40$ ). (B) Von Kossa staining shows reaction of the basophilic materials, which were identified as calcium (Von Kossa stain,  $\times 40$ ).

## DISCUSSION

Calcinosis cutis is characterized by the deposition of insoluble calcium salts in cutaneous tissue. Based on the pathophysiologic mechanisms, it has been classified into dystrophic, metastatic, idiopathic, and iatrogenic forms<sup>5,6</sup>. MICC is a subtype of idiopathic calcinosis cutis that occurs in the absence of identifiable underlying tissue abnormalities or systemic metabolic disorder.

MICC appears as smooth, round, firm, whitish papules resembling milia. They are sometimes surrounded by erythema. The predilection sites are the hands and feet, but it can occur on the any part of the body. Clinically, MICC may resemble warts, epidermal cysts, molluscum contagiosum, and syringomas. In our case, milium, verruca plana, and molluscum contagiosum were diagnostic considerations at the time of biopsy. Histopathologically, there are well-defined, round, basophilic materials that are

stained black with Von Kossa stain. The materials may be surrounded by thick collagen fibers, epithelioid cells, and multinucleated giant cells. As was observed in our case, transepidermal elimination of basophilic materials may be present. MICC occurs in childhood, and the average age of patients is 9.9 years old, ranging from 4 months to 21 years<sup>2,4</sup>. In some cases, MICC has been associated with Down syndrome. To our knowledge, two cases of MICC without Down syndrome have been reported in Korea (Table 1)<sup>3,4</sup>. The patients were children, and their clinicopathologic findings were similar. In one case, immunohistochemical stainings for CEA and epithelial membrane antigen were positive in the structures around the calcium deposition<sup>3</sup>.

Although the pathogenesis of MICC is not clear, several theories have been suggested. One is that sweat ducts may play a role in calcium deposition, and calcified sweat ducts have been described in some patients<sup>3,7,8</sup>. Another

theory is that the lesions represent microepidermal cysts that secondarily generate an inflammatory reaction and calcium deposition<sup>9,10</sup>. In the present case, immunohistochemical staining for CEA was performed to find sweat ducts around calcium deposits, but the result was negative. Moreover, there was neither considerable inflammatory cell infiltration around calcium deposits nor evidence of microepidermal cysts on histopathologic examination. In Down syndrome, a premature aging process has been suggested as a pathogenesis of MICC<sup>1,11</sup>. However, MICC may occur in healthy children, and our patient was not affected by Down syndrome, although he was born premature.

There have been a few reports of calcinosis cutis associated with prematurity. Most of them were iatrogenic forms that were caused by extravasation of calcium solution. Cambiaghi et al.<sup>12</sup> reported another kind of calcinosis cutis in a premature baby who was affected by periodic transepidermal elimination of calcified nodules in the soft tissue of the fingertips until the age of three years. The authors suggested that the fingertip calcinosis cutis was probably caused by ischemic damage due to venous obstruction.

Our patient was born premature at 26 weeks' gestation and was managed in the intensive care unit during the neonatal period. Though there was no problem such as extravasation of calcium solution during that time, deformities on the right 4th and 5th toes were observed. Deformities on the right foot reminded us of the possibility of occurrence of MICC following ischemic injury, as in the report of Cambiaghi et al.<sup>12</sup>. However, the MICC lesions also occurred on the normal appearing left foot. As a result, other factors besides ischemic damage were likely to play roles in the pathogenesis. Further studies are warranted to elucidate the complete pathogenesis of MICC.

MICC may heal spontaneously, with or without scarring<sup>7</sup>, and mostly disappears before adulthood<sup>13</sup>. Choi et al.<sup>3</sup> reported successful treatment with tretinoin cream in one patient who had showed long-lasting lesions and repeated occurrence of new lesions<sup>3</sup>. In the present case, during the 21-month follow-up period, new lesions occurred within the first 1~2 months. Thereafter, new lesions were not observed, and existing lesions regressed gradually and

almost disappeared at the age of 40 months old.

In conclusion, we describe a peculiar case of MICC occurring on both the injured and the non-injured skin in a toddler born prematurely.

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