Medium-Sized Congenital Melanocytic Nevus with Halo Phenomenon: A Report of Two Cases

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ABSTRACT
Congenital melanocytic nevi with halo phenomenon in children is a rare clinical finding. We report two cases of children who developed depigmentation within medium-size congenital melanocytic nevi. Clinicians should be suspicious when confronted with this phenomenon due to the risk of development into malignant melanoma. Herein, we review these rare cases of medium-sized halo congenital melanocytic nevi with and without associated vitiligo, with discussion on the current recommended surgical modalities in treating medium-sized CMN.

INTRODUCTION
Congenital melanocytic nevi with halo phenomenon describes the spontaneous involution of congenital nevi with a surrounding area of depigmentation. Although this phenomenon is rare and likely to be benign, there are reports of melanoma development either within the halo nevus or at a distant site.1 One systematic review of adult-onset halo nevi showed that there is a 1% risk of melanoma development in the first year following halo nevi diagnosis;2 however, there is very limited research following congenital halo nevi.

Herein, this case series describes two patients who developed a halo and complete depigmentation in pre-existing medium-sized CMN, with and without distant vitiligo. This report highlights that dermoscopy and clinical monitoring can prevent unnecessary excision in pediatric patients, but parents may still prefer surgical treatment; we review the current surgical modalities available in removal of CMN.

CASE PRESENTATION

Case 1

An 8-year-old girl presented with 2-3 years of depigmentation and surface changes on a brown birthmark involving the left medial knee. Initially, the depigmentation surrounded the lesion, but soon after affected the entire lesion. The patient was otherwise healthy and without any family history of cancer or autoimmune diseases.

Physical examination showed a 5.0 x 2.2 cm asymptomatic, depigmented patch on the left
medial knee with residual pink and brown papules and macules with poliosis (Figure 1). Dermo­scopy of the lesion (Figure 2) failed to identify any structures consistent with melanoma.

![Figure 1. A pink and brown plaque on the left medial knee with surrounding and central depigmentation and poliosis](image1)

![Figure 2. Dermoscopy of depigmented patch on left medial knee showing an absent pigment network](image2)

Case 2

An otherwise healthy 6-year-old girl presented with a black mole on the abdomen and a white spot above her left eye. The lesion on the abdomen was present at birth and evolved as the patient grew. About 8 months prior, the mole became darker and one month later a white area developed around the periphery. About 6 months later, the patient developed an asymptomatic white patch above her left eye.

Physical examination showed a 2.0 cm depigmented patch on the left upper eyelid that fluoresced on Woods lamp examination. On the abdomen, there was a 4.5 cm dark brown to black papillated plaque with a 1 cm rim of depigmentation (Figure 3) with no clinical or dermoscopic abnormalities. The patient was diagnosed with a medium-sized CMN with halo phenomenon and an associated vitiligo patch.

![Figure 3. Dark brown irregular plaque on abdomen with a rim of depigmentation](image3)

**DISCUSSION**

The patients presented in this case series demonstrate halo transformation of medium congenital melanocytic nevi. While acquired nevi may develop halo depigmentation before spontaneously regressing, this halo phenomenon is less commonly seen with CMN.³

The cause of halo nevi is thought to be caused by an immunologic response to nevus cells and melanocytes.⁴ Recent
literature has demonstrated some connection between the mainly CD8+ T-cell mediated pathogenesis of halo nevi transformation with that of vitiligo’s anti-melanocyte response. A similar reactive oxidative species pathogenesis of both halo nevi phenomenon and vitiligo has been implicated by Yang et al through measurements of H$_2$O$_2$ levels in tissues.

The similarities in pathogenesis may explain why halo nevi are up to 10 times more common in patients with vitiligo than the general population. This relationship between halo nevi and vitiligo is demonstrated by the patient presented in case two. Similarly, other cases involving CMN halo transformation have demonstrated distant vitiligo. This association may aid clinicians in educating parents of the possibility of appearance of vitiligo in patients with CMN that are undergoing the halo phenomenon.

Both of the cases presented here represent rare instances of medium-sized CMN which demonstrate halo phenomenon. While case one represents halo transformation of medium CMN without vitiligo, case two represents halo transformation of medium CMN with distant vitiligo. Just as in these two cases, the variability between nevi demonstrating halo phenomenon has been previously studied and it is recommended that clinicians note that not all nevi with this distinction are malignant, thereby, requiring excision. With thorough clinical monitoring of nevi, clinicians can avoid the use of invasive procedures while caring for patients with medium-sized CMN. However, studies have shown that parents frequently choose removal of CMN due to reasons other than concern for melanoma; specifically, a study has shown that the leading reason for treatment of CMN was to reduce psychosocial difficulties (58.7%), followed by aesthetic reasons (50.4%), and to lower melanoma risk (46.3%). Despite the side effects that surgical excision can bring, such as repigmentation and hypertrophic scarring, majority of the parents believed that a scar is more socially acceptable than a CMN.

With this in mind, there are multiple surgical modalities available for the treatment of CMN, such as complete surgical excision, dermabrasion, curettage, and laser therapy. For the treatment of large-sized CMN, fresh cultured epithelial autograft (CEA) after curettage or erbium:yttrium-aluminum-garnet (Er:YAG) ablation was found to be a novel option with high patient satisfaction and fewer side effects. On the other hand, less is known regarding effectiveness of surgical treatments for medium-sized CMN, and the patients satisfaction with such treatments. Based on a systematic review, the most acceptable surgical treatment of a medium-sized CMN would be to excise using tissue expanders, as it was shown to have few complications and high patient satisfaction. In terms of laser therapy, Q-switched ruby laser with wavelength at 694 nm was shown to be efficacious in treating medium-sized CMN as it allows for highly selective destruction of pigment-laden cells.

Parents are highly concerned about the psychosocial difficulties their child may face due to CMN, and may prefer surgical removal despite the low melanoma risk. Healthcare professionals should focus on clear communication about melanoma risk, indications for surgery, and expected outcome to best support families’ decision-making.

CONCLUSION
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