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Rapidly Involuting Congenital Melanocytic Naevi in Two Children

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Congenital melanocytic naevi (CMN) are considered to be neural crest-derived hamartomas, which are visible at, or shortly after, birth as pigmented tumours (1). The incidence of any size of CMN of neonates ranges from 0.2 to 2.1% (2, 3). They are categorized according to the maximum diameter that the naevus will achieve; small (< 1.5 cm), medium (1.5–10 cm), large (> 10–20 cm) and giant (> 20 cm) (4). In addition to the cosmetic concern, CMN pose several problems, such as neurocutaneous melanosis, and risk of malignant transformation (5, 6). In a systematic review the risk of melanoma in CMN was estimated to be 0.7%, with a higher risk of malignant transformation occurring during childhood and adolescence for giant CMN (2, 7, 8).

CMN have a dynamic course and may change over time. They can increase in size during childhood, become darker in colour, become hairy, or show pigmentary regression (9). Spontaneous involution is a rare phenomenon with few cases described in the literature and, when it occurs, it is often associated with a hypo- or de-pigmented halo (6, 9, 10). We report here 2 cases of spontaneous rapid involution without the halo phenomenon, in medium to large CMN in 2 children referred to the Karolinska University Hospital.

CASE REPORTS

Case 1. A 5-month-old girl presented with a large CMN (3.5 × 1.5 cm diameter) over the dorsal aspect of the left foot, extending to the sole and the fourth and fifth toes. The lesion was light-brown with a reddish infiltrate component. No satellite lesions were present. Punch biopsy showed an atypical pigment-producing tumour, and a melanoma arising from the CMN could not be excluded. Therefore, 4 additional incisional biopsies were performed. The histopathology was difficult to interpret, and Dr Wallace H. Clark from Harvard Medical School was consulted. A final diagnosis of benign CMN with a Spitzoid differentiation was made and melanoma was ruled out. Dr Clark commented, after combining the medical history with the dermatopathology, that in his experience similar features could be seen in CMN of that age. In order not to risk impaired function of the foot, no further surgery was performed. Instead, regular examinations, including dermoscopy and palpation of the CMN and regional lymph nodes, were made every 3–6 months up to the age of 7 years, and then less often. The CMN presented a dynamic course, initially with some increase in pigmentation, but then a gradual clinical regression occurred with loss of infiltration and normalization of the skin colour at 4.5 years of age (Fig. 1).

Case 2. A 2-month-old girl presented with a medium-sized CMN (7 × 5 mm, estimated to grow by a factor of 3, i.e. increasing in size to approximately 21×15 mm), which was a dark-brown wedge-shaped macula in the umbilical area present since birth. Dermoscopy revealed irregular pigment distribution with confluence of black and brown dots. The naevus was deeply pigmented at birth, but faded by the time the patient was seen in our clinic. No biopsies were performed and, by the age of 3 years, only a minor residual brown macula (1 × 2 mm) and a greyish shadow was visible (Fig. 2).

DISCUSSION

Spontaneous involution of CMN is rare and is usually associated with the halo phenomenon or vitiligo (6, 9). Cusak et al. (10) described a case of complete regression over a 4-month period with the halo phenomenon of a medium-sized CMN. A control biopsy showed prominent lymphocytic infiltrate in the papillary dermis and aggregates of lymphocytes adjacent to the naevus in the deeper dermis, supporting an autoimmune me-

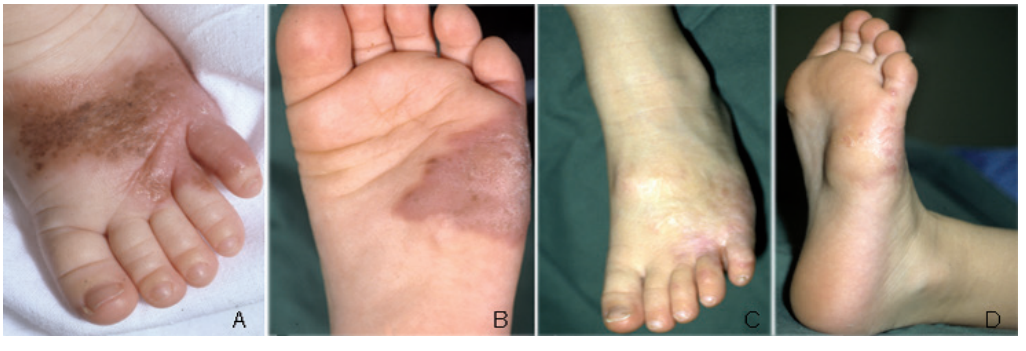

Fig. 1. Case 1. Large congenital melanocytic naevus of the foot, (A and B) at 5 months and (C and D) at 4.5 years of age. At age 4.5 years some infiltration remained in the sole, but the abnormal brown colour had disappeared.

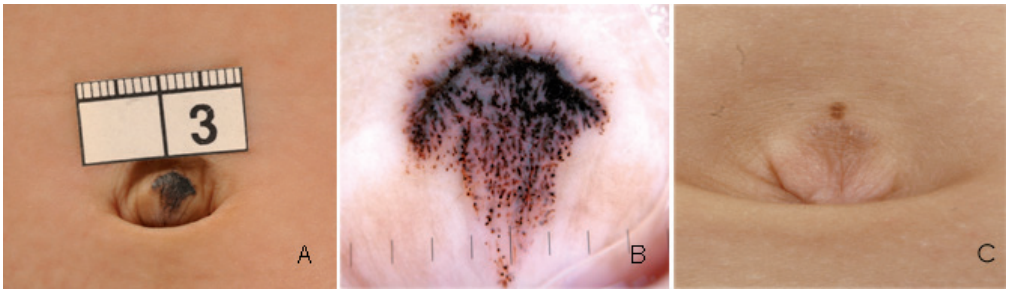

Fig. 2. Case 2. Congenital melanocytic naevus in the navel, (A) at 3 months, and (B) dermoscopy at the same age. (C) The same patient at the age of 3 years.

chanism with T-cell mediated immunity and circulating anti-naevus IgM antibodies involved in the regression.

Despite the clinical involution, pigmentary regression may be accompanied by histological persistence of naevus cells, as described previously in a case of involution of a neonatal eroded giant CMN with desmoplastic reaction (11). Vilarrasa et al. (12) reported 2 cases of medium-to-large CMN that clinically disappeared without the halo phenomenon. In both cases, skin biopsies showed that a high proportion of amelanotic naevus cells were still present in the dermis and even extended to the subcutaneous fat and pilosebaceous unit, indicating a decrease in melanin production by dermal naevus cells, rather than a reduction in their number. More recently, Nath et al. (13) described a spontaneous regression of a CMN in which histopathology demonstrated lymphocytic infiltration and loss of pigment production, and immunochemistry staining (S 100) showed the persistence of naevus cells.

Prophylactic surgery and subsequent reconstruction are often complex undertakings as well as dermabrasion, laser ablation and chemical peel for treatment of large and giant CMN (5, 14). In addition, most of these procedures often leave naevus cells behind, since these cells often extend deeply along skin appendages and even underlying skeletal muscle (5). Moreover, the risk of malignancy occurring in extracutaneous melanocytic deposits (leptomeninges, gastrointestinal tract and retroperitoneum) remains unaltered by the excision of giant CMN (5, 14). Another point to consider concerning the excision of large-to-giant CMN is that the risk of anaesthesia does not outweigh the risk of developing malignant melanoma when performed in children under 14 months old (15). Both patients and surgeons must carefully weigh the benefits of surgery and the chances of avoiding malignant transformation (14).

With these case reports, we would like to highlight that some CMN may show a rapid involution without a halo phenomenon, thereby eliminating cosmetic problems, and perhaps also decreasing the risk of malignancy. In case 1 total excision without partial amputation of the patient's foot would have been impossible, whereas it would be possible in case 2. We cannot rule out a risk of future malignant transformation originating from the CMN in these patients, so instructions have been given for self-examination to detect any signs of changes from benign to malignant.

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