# Proliferating Nodules Within a Congenital Melanocytic Nevus : Proper criteriae for surgical removal in infantile periods

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**Congenital melanocytic nevus(CMN) constitute a well-known risk factor in the development of malignant melanoma, but melanoma developing in newborn and infants are extremely rare. We describe a case of proliferating nodules within CMN at the age of 13 months. Like our case it is difficult to manage the proliferating lesions within CMN in infants. So we carefully suggest the indications of preventive excision when proliferating lesions occur within CMN as follows; 1) preventive excision is postponed until the age of two when the proliferating lesions are slowly growing, 2) surgical excision is needed when the lesions are rapidly growing exceeding 1 cm in size even though it is before the age of two.(Ann Dermatol 13(2) 120~122, 2001).**

*Key Words* : Congenital melanocytic nevus, Melanoma, Surgical indication

It is indisputable that congenital melanocytic nevi(CMN) occasionally provide the setting for the development of malignant neoplasm[1]. Although this occurrence is often considered quite remarkable, the actual frequency of a malignant tumor arising in congenital nevi is uncertain[2]. So the management of congenital nevi has been controversial.

Most CMN, regardless of size, show continuing intraepidermal melanocytic growth throughout life[3], and these CMN are raised and exhibit a variably papular, lumpy, rugose, or even verrucous surface[4]. Malignant melanoma may develop in these proliferating nodules in prepubertal periods, therefore it must be differentiated between benign proliferation and melanoma.

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However, it is difficult to predict the exact risk of malignant melanoma in patient with proliferating nodules within CMN in infantile and young childhood periods. In this report, we present a case of proliferating nodules within CMN at the age of 13 months with a review of literatures.

## CASE REPORT

A 13-month-old male patient was born with a well-defined brownish patch of CMN on the lower back. At the age of 5 months, three nodules developed within the nevus, and one of them showed rapid growth. At the time of presentation, the congenital melanocytic nevus was 4×3 cm sized, and the presence of three nodules measuring up to 1 cm sized in maximal dimension underlying the pigmented lesion(Fig.1). The largest nodule was rapidly growing with a feature of multiple grouped, deeply pigmented papules. All of the three nodules were excised. Examination of the smallest papule showed hyperplasia of basal melanocytes, but there was no atypia(Fig. 2A). The medium sized papule displayed a more organized pattern of nevus cell distribution in the dermoepidermal junction with

significant development of melanocytic nests. The excisional biopsy of the rapid growing nodule showed an extensive junctional and dermal congenital melanocytic nevus with the superficial distribution of pigment in an umbrella-like pattern(Fig 2B). Compound nests were found and most of the nests were composed of cells with bland cytologic features and rare mitotic figures, and the cells had a monomorphous aspect and lacked the pleomorphism. Infiltration of upper portion of epidermis and deeper part of dermis were not observed. He had no evidence of recurrence after 6 months of treatment.

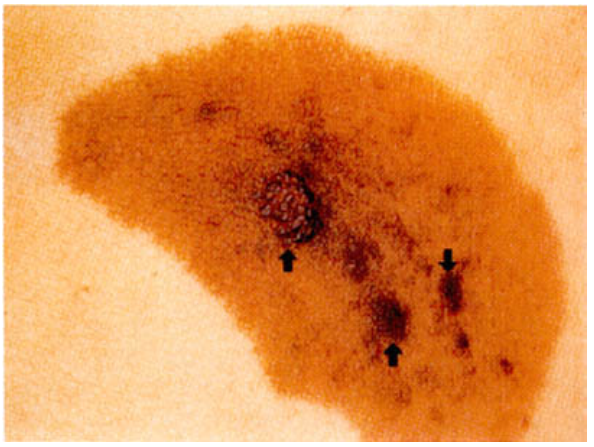

Fig. 1. A 4×3 cm sized lightly pigmented congenital nevus on the lower back, with three well-demarcated, deeply pigmented nodules(arrows).

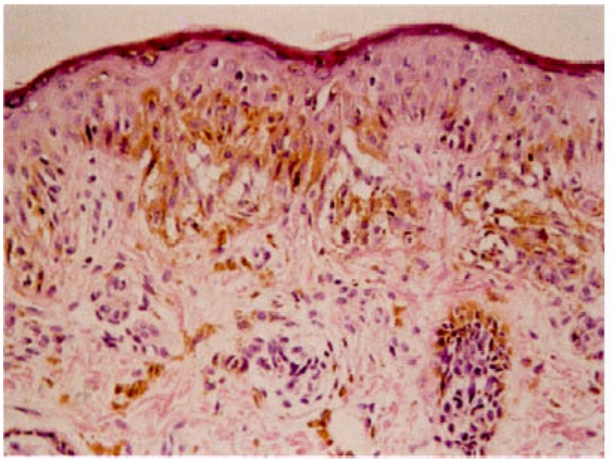

Fig. 2A. Histologic appearance of smallest papules(Fig. 1) showing the hyperplasia of basal melanocytes without atypia (Hematoxylin-eosin stain; original magnification, ×100).

## DISCUSSION

The risk of melanoma development is proportional to the size of the CMN[5-7], and many authors have arbitrarily divided CMN according to the largest diameters as small(<1.5 cm), medium(1.5 to 19.9.cm), and large(>20cm)[8]. But there is no complete satisfactory way to separate CMN as small or large. CMN may be defined as large if they cannot be excised easily and cannot be closed primarily without skin flap or grafts. And CMN has been variously defined as a giant as large as the patient' s palm when they are in the face or neck (and twice that area for other anatomic sites) and when they occupy over 30 percent of the body surface. In our patient, the lesions could be regarded as small lesions following the first criteria, but as large following the second or third criteria. So it is difficult to define the exact incidence of melanoma according to the size of CMN especially in the infantile or young childhood period when the lesion is continuously enlarging.

And the incidence of melanoma increase with age but it is exceptionally rare in prepubertal individuals (estimated incidence approximately 0.4 percent among all melanomas) and uncommon under the age of 20 years (incidence approximately 2 percent)[1,3,9]. Particularly, Handfield and Smith[9] described 24 cases of malignant melanoma in children and the youngest patient was 2 years old. So, melanoma developing in infants and young children under the age of 2 years is extremely rare and diagnosis of melanoma must always

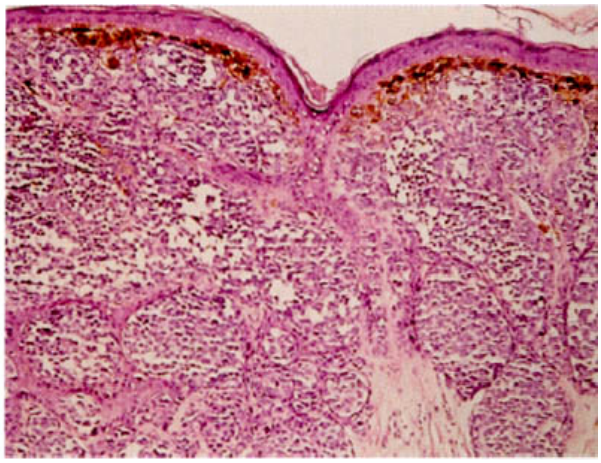

Fig. 2B. Histologic appearance of rapid growing nodule show the dermal melanocytic nevi with the superficial distribution of pigment in an umbrella-like pattern (Hematoxylin-eosin stain; original magnification, ×100).

be seriously questioned. These reports indicate the relatively good prognosis in young childhood period of patients with CMN.

CMN may give rise to intradermal nodular melanocytic proliferations[10]. But, proliferative dermal nodules within a congenital nevus are uncommon. These nodules may feature large melanocytes which can form nests and these cells have been termed melanoma simulant cells. The etiology of these proliferative cells is unclear, perhaps they are a monoclonal proliferation of melanocytic cells[11]. Such benign proliferation must be differentiated with malignant melanoma. Nodular melanocytic proliferation within a CMN differs from nodules of malignant melanoma arising in CMN in that the nodules are small (less than 1 cm), atypical melanocytes are rare, lymphocytic infiltration are absent and maturation of melanocyte is evident in the deeper cells of the nodule[4]. The nodules do not enlarge, in fact they usually gradually regress and finally disappear spontaneously[12].

Our patient had a rapid proliferating nodule within CMN starting from the age of 5 months. Histologically the proliferating nodules showed no evidence of malignant melanoma. As other authors[13], we experienced difficulty to establish strict criteria when the surgical intervention was needed. So, we carefully suggest the indication of preventive excision when proliferating lesions occur within CMN as follows; 1)Preventive excision is postponed until the age of two when the proliferating lesions are slowly growing, 2) surgical excision is needed when the lesions are rapidly growing exceeding 1 cm in size even though it is before the age of two.

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