along Blaschko's lines and macules that converged and became bigger. Simões and Piva[2] reported that PZMPL is accompanied by itching, and that the condition remains static for a certain period of time followed by an abrupt enlargement of the lesions, which is similar to the present case.

The histological features, the extensive patches along Blaschko's lines, and the abrupt enlargement of the skin lesions suggested a diagnosis of PZMPL. Since the first case of PZMPL described by Simões and Piva[2] three more cases have been investigated, including the present one. Given that the diagnosis of PZMPL is not well established, we hope that this case report contributes towards elucidating a standard definition, the pathophysiology, and the progression of PZMPL. We recommend that dermatologists consider this disease when they encounter patients with chronic hyperpigmented dermatoses.

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# Giant Congenital Melanocytic Nevus with Proliferative Nodules Mimicking a Congenital Malignant Melanoma

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Dear Editor:

The malignant transformation of a giant congenital nevus (GCN) is extremely rare, and when it is discovered, the transformation may have already occurred during childhood. Benign proliferative nodules (PNs) in GCN may clinically and histologically mimic a malignant melanoma (MM), but clinically, PNs usually present at birth with multiple nodules, which is in contrast to MMs, and histologically, most patients with PNs do not present with

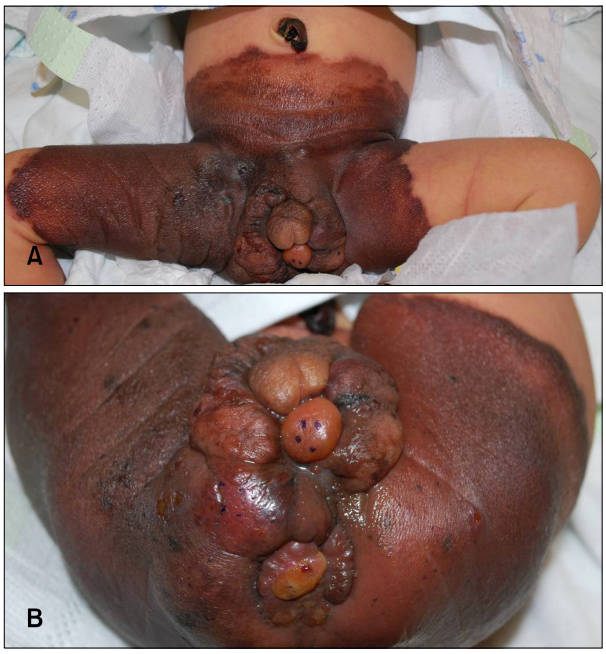


**Fig. 1.** A giant congenital nevus with proliferative nodules on lower abdomen, genitalia, anus and lower legs.

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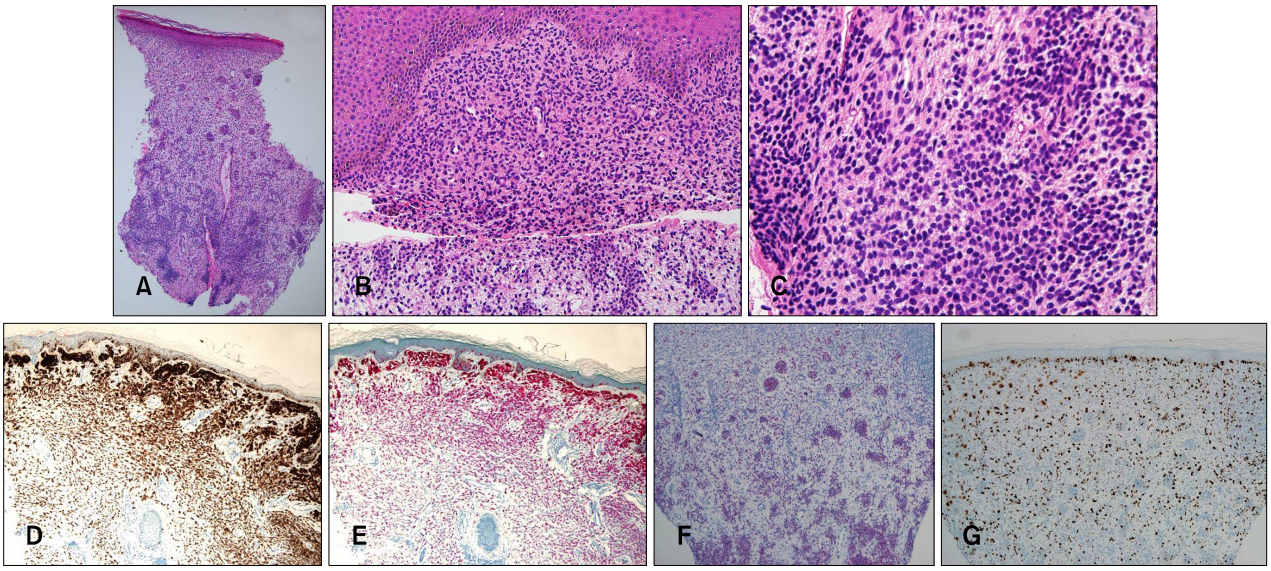


**Fig. 2.** (A) Histopathology of the proliferative nodule showed a proliferative dermal nodule in a giant congenital nevus (H&E, ×40). (B) Congenital nevus cells formed nests at the dermo-epidermal junction and upper dermis (H&E, ×200). (C) Melanocytic cells within the dermal nodule were more cellularlarger, more heavily pigmented and predominantly epithelioid cells, showing low-grade cytologic atypia and minimal pleomorphism (H&E, ×400). The immunohistochemical stains showed a positivity for (D) HMB45 (×100), (E) Melan A (×100), (F) SOX-10 (×100), and (G) Ki-67 (×100).

the characteristics that are associated with malignancy, including mitotic activity, nuclear pleomorphism, and pagetoid spreading. Furthermore, even though some cases might display these features, they soon disappear[1].

A one-day-old female child presented with GCN on her lower abdomen, genitalia, anus, and legs. Prenatal ultrasonography revealed an imperforate anus, but no genital abnormalities. Dark brown plaques and some PNs that covered the involved areas were seen during a physical examination (Fig. 1). Satellite nevi were encountered on her scalp, axillae, and ankles. A diagnostic biopsy on a PN was performed, and the histological examination presented a proliferative dermal nodule in a GCN (Fig. 2A). The nevus cells had formed nests in the dermo-epidermal junction and the upper dermis (Fig. 2A, B). They were homogenous without nuclear atypia, pleomorphisms, mitoses, or pagetoid spreading into the epidermis. The melanocytic cells in the dermal nodule showed more dense cellularity than in the background lesion (Fig. 2C), and these cells were more pigmented and larger, and most of them were epithelioid cells that showed little pleomorphism and low-grade cytologic atypia. The features of necrosis were not found. All of these histological features was consistent with a benign PN within a GCN. Immunohistochemical analysis showed that the PN was positive for HMB45 (Fig. 2D), Melan A (Fig. 2E), and SOX-10 (Fig. 2F), which detect melanocytic tumors and Ki-67 protein of less than 2% (Fig. 2G). The patient was transferred to the plastic surgery department and she underwent a staged excision.

Congenital nevi occur in 1% of newborn infants. Some authors have stated that GCN can be distinguished histologically from acquired nevi by the presence of nevoid cells in the lower two-thirds of the dermis[1]. Other authors have reported that the nevus cells subsequently migrate deeper into the dermis[2]. GCN has a 2%～42% risk of malignant transformation, and is associated with a 6%～14% lifetime risk of developing melanoma[3]. True congenital melanomas are very rare and they are supposed to develop secondary to the transplacental spread of maternal melanomas, arise *de novo in uterus*, present as prenatal growths on congenital nevi, or arise from neurocutaneous melanosis. A PN within a congenital nevus is also uncommon. Compared with congenital nevus cells, PN cells are larger and more pigmented, they have large nucleoli and a storiform pattern, and they can form nests. These cells have been called "melanoma stimulant cells"[4]. The etiology of them is unknown, and they may be a monoclonal proliferation of melanocytic cells. PNs within GCN grow rapidly, show satellitosis[5], and they often bring up diagnostic difficulty. In a study of the cells in PNs in GCN, the cells showed high malignancy, but they did not present the typical antigens expressed in MMs, and their subsequent behavior was benign[5]. But, because the possibility of malignant transformation of this cells is unclear, surgical procedure is recommended to exclude congenital

melanoma. We presented the current case to underline the development of benign PN within GCN and to suggest its differential diagnosis from congenital MM.

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