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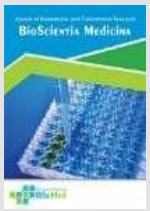

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A Rare Case of Giant Congenital Melanocytic Nevi: A Case Report

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A B S T R A C T

Background: Giant congenital melanocytic nevi are often characterised as melanocytic lesions present at birth. It is thought that it affects 1 in 20,000 births. The lesion is significant because it may be associated with serious consequences, such as malignant melanoma, and may also result in neurological deficits, such as neurocutaneous melanocytosis. This study aimed to present the case of giant congenital melanocytic nevus as a rare case. **Case presentation:** This study reported a 14-year-old female reported with large blackish rough spots on the trunk that were neither itchy or painful since birth. The patient had no previous history of convulsions, headaches, or vomiting. Plaque hyperpigmentation, macula hyperpigmentation, and hypertrichosis were found on the trunk. Dermoscopy show reveals brown-black homogeneity. Histopathology examination reveals brown-pigmented nevus cells distributed diffusely, with some nevus cells surrounding adnexa skin. **Conclusion:** Regular examination is essential to monitor the possibility of neurocutaneous melanosis and malignant melanoma development.

1. Introduction

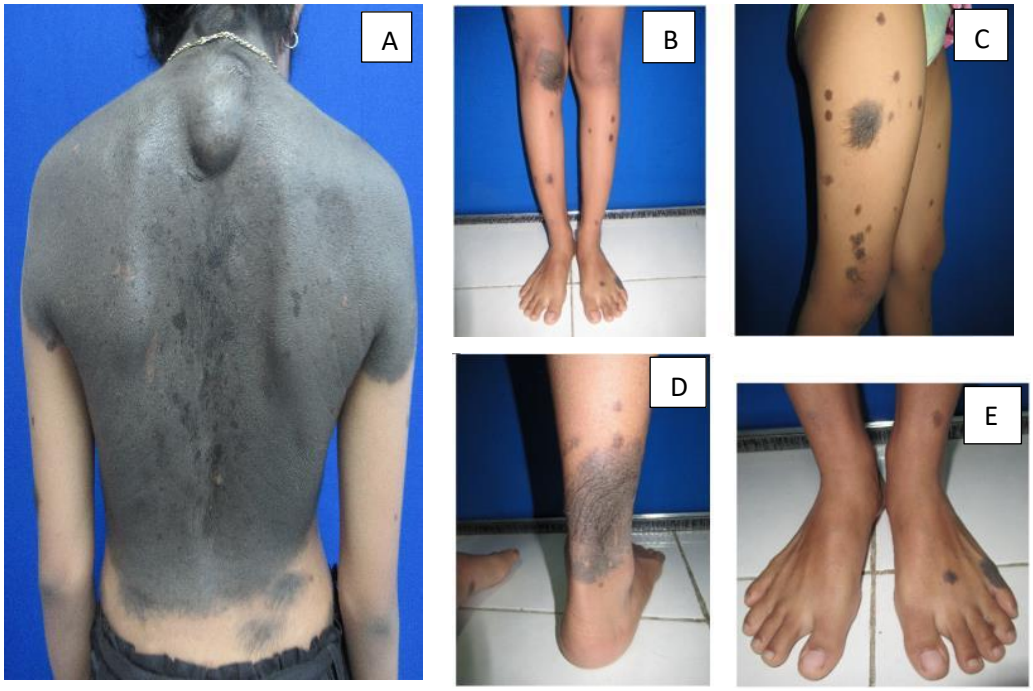
A giant congenital melanocytic nevus (GCMN) is an adult melanocytic lesion reaching at least 20 centimeters in diameter.[1,2] Approximately 1% of births present with a CMN. GCMN is expected to affect fewer than 1 in 20,000 neonates. The "garment-like" variant of GCMN is much more uncommon: 1:500,000.[3,4] GCMN is important despite its rarity due to its association with severe consequences such as malignant melanoma and central nervous system (CNS) involvement, as well as its unpleasant appearance, which has a substantial psychosocial impact on the patient and his family. Due to the disagreement surrounding the treatment of these lesions, which stems mostly from the unpredictability of the hazards involved, the medical team is also agitated when determining the optimal therapeutic approach in these circumstances.[2,5] The GCMN would then be classified into three categories: G1 (21 to 30 cm), G2 (31 to 40 cm), and G3 (more than 40 cm). Patients who were previously classified into a category based only on the nevus size would increase if they had giant nevi and more than 50 satellite lesions.[6-8] This study aimed to present the case of giant congenital melanocytic nevus as a rare case.

2. Case Presentation

A 14 years old girl was admitted to the dermatology and venereology outpatient department of Dr. M. Djamil General Hospital Padang with a chief complaint

of blackish patches at the trunk that did not feel itchy or painful. The patches have existed on the back, neck, shoulders, left breast, left arm, lower leg, or buttock since birth. There was a family history of the patient's father, uncle, and grandmother having blackish, painless patches from birth. The patient had no history of seizures, headaches, and vomiting.

Physical examination revealed dermatological state 1, location on the back, neck, shoulders, left arm, and lower limbs with hyperpigmentation, hypertrichosis, and efflorescence plaques (Figure 1). Dermatological condition 2 is located on the middle to upper back and is a hyperpigmented tumor. Histology indicates brown-pigmented nevus cells scattered diffusely, with some nevus cells surrounding the adnexa skin. A massive congenital melanocytic nevus was observed on the patient. The dermatology life quality index score evaluation was 10.



Figures 1. (A-E) Plaque hyperpigmentation.

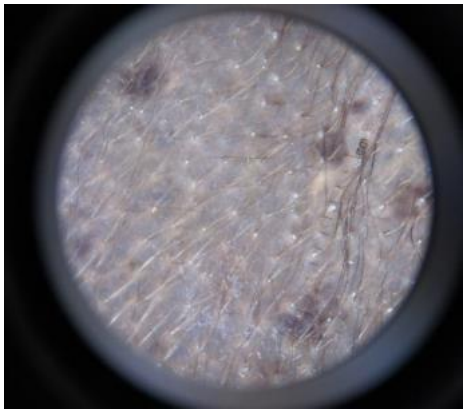


Figure 2. Dermoscopy examination: homogenous brown-black.

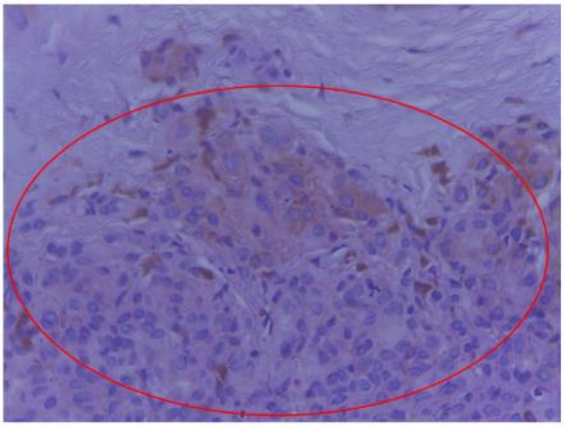


Figure 3. The surface of the epithelium reveals hyperplasia (acanthosis), and the sudoriferous gland is surrounded by nevus cells (red circle) (magnification 10). There are no nevus cells in their follicles (magnification 40).

3. Discussion

Since birth, the woman, in this case, had large blackish rough patches that were neither itchy nor painful over her back, neck, shoulders, left breast, left arm, lower leg, and buttock. GCMN (way rarer) is predicted to impact 1 in 20,000 newborns. The prevalence of CMN is 1-6% among all newborns, but GCMN (much rarer) affects 1 in 20,000 births.[9] Giant congenital melanocytic nevi occur sporadically, although there are relatively few publications about the potential of these conditions having inherited.[13] According to Anamnesa, the patient's father, uncle, and grandmother were identical to the patient.

This patient's back, neck, shoulders, left arm, and lower limbs have exhibited hypertrichosis since infancy, evidenced by extensive, blackish, rough, non-pruritic lesions. GCMN commonly manifests as a brownish lesion or black with well-defined margins and hypertrichosis. As a macule or elevated lesion, it may be lighter in color and have few or no hair follicles in babies.[6] This patient was diagnosed with giant congenital melanocytic nevi based on the history, physical examination, and histopathology.

The GCMN would be further broken into three categories: G1 (21 to 30 cm), G2 (31 to 40 cm), and G3 (greater than or equal to 40 cm) (more than 40 cm). Taking only the size of the nevus into account, the classification of patients with giant nevi and more than 50 satellite lesions would be raised by one level.[8] This patient was classified as having a G3 category giant congenital melanocytic nevus due to the presence of a lesion larger than 40 cm and a satellite lesion larger than 50 cm.

A Becker nevus is considered a differential diagnosis for this patient. Young males are more likely than women to develop the uncommon pigmented smooth muscle hamartoma known as the Becker nevus throughout adolescence. It is characterized by hypertrichosis and hyperpigmentation and often develops unilaterally over the shoulder, upper arm, and scapula. According to histopathology, nevus Becker is distinguished by acanthosis, papillomatosis, keratotic plugging, unequal rete ridge elongation, and epidermal flattening on a regular basis.[14] Dermoscopy findings of a network, localized skin furrow, perifollicular hypopigmentation, hair follicles, and vasculature are characteristics of nevus Becker.[15]

The presence of melanocytes grouped in nests inside the epidermis, elongation of epidermal ridges coupled with an increased number of melanocytes, and hyperkeratosis and hyperplasia are the most typical histological findings of gigantic congenital melanocytic nevi.[11] Dermoscopy is the best method for evaluating mild to moderate CMN. Regarding GCMN, the fact that its nevus cells are found more deeply in the skin is one of the reasons for the limited application of dermoscopy. The majority of giant nevi have a brownish backdrop with darker pigmentation islands. Other dermoscopy observations include hypertrichosis, hypo- or hyperpigmentation of the

perifollicular area, pseudo milia, and vascular structures.[16] In this patient, a tumor measuring 6cmx5cmx2cm was diagnosed as a lipoma, whereas a neurofibroma was the differential diagnosis.

Neurocutaneous symptomatic melanosis is uncommon, and the prognosis is worse for asymptomatic neurocutaneous melanosis; typically, 70% of patients with neurological disease signs die before age 10. In this case, no clinical melanocytic neurocutaneous signs were seen. However, a neurocutaneous melanocytic investigation is required.[7] Melanocytic deposition in the CNS is assessed by radiographic imaging, which includes MRI. The baseline MRI should be performed between 4 and 6 months of age. This patient was referred to the pediatric neurology division for examination. The pediatric neurologist found no neurologic impairments and intended to perform a CT or MRI scan, but the patient refused.

Malignancy risk is further increased when there are more big naevi (>50 cm) on the trunk, head, or neck, when there are several satellite lesions, and when there are nodules, dark patches, junctional activity, deep dermal neurogenic elements, or a blue naevus component.[10] There is substantial evidence in the existing scientific literature that these persons have an elevated chance of developing cancer. It is estimated that the lifetime risk of acquiring melanoma for these persons is between 5 and 10 percent, even though the incidence rate of malignancies in GCMN is still controversial.[17]

Marghoob et al. conducted a literature review and discovered that over 70% of melanomas related to GCMN were detected before puberty, while less than 1% of total melanoma occurrences in the U.S. population occur at this age.[18] In a study by Dedavid et al., which included 289 individuals with GCMN, it was determined that around fifty percent of melanomas were diagnosed before age five.[19] The purpose of pre-melanoma treatment is to lower the risk of cutaneous melanoma and for cosmetic reasons. Excision, curettage, dermabrasion, chemical peeling, cryotherapy, electrosurgery, and ablative laser are possible treatment procedures.[12] The patient did not return for monitoring, so this progression could not be monitored. Due to its unattractive appearance and considerable psychosocial impact on the patient and his family, the patient's plan will include psychological counseling or a consultation with psychiatry.

In order to treat GCMN, surgical procedures such as serial excision and repair with skin grafting, tissue expansion, local rotation flaps, and free tissue transfer may be utilized. The following are some examples of other surgical techniques that might be used: Due to the depth of some lesions. It is probable that excisions will not be able to eliminate the chance of melanoma in some patients. This is especially true if leptomeningeal involvement is present.[12]

Clinical follow-up of GCMN is also difficult because the surface of the lesion often has irregularities and nodules, and it is often dark and has much hair. Patients and their parents should be instructed to undertake regular skin self-examinations and seek medical attention if the nevus or other abnormal symptoms change in color, shape, surface, or appearance. A multidisciplinary team of dermatologists, plastic surgeons, radiologists, and, if necessary, psychologists and neurologists assists with patient follow-up. It is crucial to palpate the lesion and lymph nodes during the periodic examination. Taking sequential images makes it easier to track how GCMN has developed over time.

4. Conclusion

GCMN is a relatively uncommon disorder characterized by a pigmented lesion at birth or within the first year of a person's life, but the underlying cause is unknown. Because of the possibility of developing MM and the problems of neurocutaneous melanoma, it needs to be correctly diagnosed and managed appropriately. The monitoring of the patients is a critical component of the management process. It is essential to remember that most individuals diagnosed with GCMN are capable of living a healthy and productive life, regardless of the type of care chosen, be it surgical or observational.

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