

GIANT CONGENITAL NEVI : PECULIAR AND CONTROVERSIAL FEATURES

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Giant congenital melanocytic nevi (GCMN) have several peculiar and controversial aspects in relation to : **Definition, size, location, color, surface, forms of presentation, malignant transformation and management.**

Definition : Conventionally, the term GCMN is reserved for intradermal or compound nevi made of special melanocytes known as nevus cells. Characteristics of GCMN are: Presence at birth, size of more than 20 cm in diameter, dark color, excessive amount of melanocytes, and malignant potential. However several other nevic lesions made of melanocytes share characteristics with the GCMN but are not considered as such. Examples are, among dermal melanocytosis : Giant blue nevus, nevus Ota, nevus Ito, Mongolian spot, and phakomatosis pigmento-vascularis. Among epidermal melanocytosis giant epidermal melanocytic nevus. Among combined epidermal-dermal melanocytosis Becker nevus, and giant nevus spilus.

Conclusion : There are several types of GCMN. Conventionally, only compound or intradermal nevus-cell nevi are considered as such. Other giant melanocytic or combined giant nevi receive special names.

Giant Congenital Melanocytic nevi. Characteristics that matter :

Size – location – color – surface

Size Why is size important ? Because it is directly related to risk of malignancy, usually causes more esthetic problems, the larger the nevus size the greater the therapeutic challenge, and more non-malignant complications

Size measure Current criteria for nevus size classification as giant are : Diameter of more than 20 cm, palm size on the face and twice elsewhere, more than 900 cm², relative area index, etc.

The current classification of nevi (Kopf 1979) in small, medium, and large clumps together nevi ranging in size from 1.4 cm to 19.9 cm and makes no distinction of nevi measuring much more than 20 cm in which the prognosis of malignant transformation, potential complications, and treatment are not the same.

New size classification proposed

The proposed classification for nevus size is as follows :

Small	< 1.5 cm diameter
Medium	1.5 to 10 cm
Large	11 to 20 cm
Giant (G)	
G1	21-30 cm
G2	31-40 cm
G3	> 40 cm

Patients with giant nevi with more than 50 small or medium sized satellite nevi should be classified one group above their corresponding size classification.

Location	Location of GCMN is important in :
Eyelides	Divided nevus of eyelids : As a marker of embryonal development of nevus (< 24 w. gestation).
Limbs	GCMN in the limbs : As a cause of limb hypotrophy (Ruiz-Maldonado R. et al J Pediatr 1992; 120: 906-11)
Scalp	GCMN over the scalp and spinal column : As a marker of neurological alterations and neurocutaneous melanosis (Ruiz-Maldonado R. et al Dermatology 1997; 195: 125-128) GCMN on the scalp often undergo spontaneous depigmentation
Perineum	GCMN of genital and perineal area (Alvarez-Mendoza et al Ped Develop Pathol 2001; 4: 73-81) as a special clinico-pathologic variant GCMN in mucocutaneous, orificial location, palms and soles as therapeutic challenges.
Surface and color	The surface of most GCPN (86%) is pigmented and hairy, some are pigmented only (11%).
Nodules	The presence of benign nodules (19%) is relatively frequent. Rapid growth, pain or ulceration may be signs malignant transformation.
Plexiform growths	A corrugated surface (6%) or plexiform newgrowths (6%) are seldom new observed and do not imply malignancy.
Color	The color of GCPN is usually black (80%), brown in 16%, and mottled in 4%. In newborns GCPN are darker than a few months thereafter. Development of melanoma on a black GCMN is difficult to diagnose. Diffuse loss of pigment in a black GCMN is associated to spontaneous regression (scalp), and to desmoplasia. Focal loss of pigment may denote malignancy (amelanotic melanoma).
Variants	Newly recognized clinico-pathologic variants of GCMN. Bulky nevocitoma of perineum. This GCMN is characterized by its perineal location, its massive dimension and histopathologically structures of neuroid appearance and pseudo-follicular structures lined by nevus cells. (Reyes-Mújica M et al Virchows Arch A Path Anta Histopathol 1992; 420: 87-93). Desmosplastic, hairless, depigmented nevus. This variety of GCMN is clinically characterized by hardening of the nevus that becomes of ligneous consistency , hairless depigmented and in most cases intensely pruritic (four of six patients). Histopathology shows intense dermal fibrosis invading the fat tissue, and absence or diminution of adnexal structures. No clinical or histopathological features of malignancy were found. (Ruiz-Maldonado R et al Br J Dermatol, in press).
Malignant	Congenital melanocytic nevi of all sizes may undergo malignant

Transformation	<p>transformation, however it is accepted that the larger the nevus the greater the risk of malignant transformation. Besides size a factor that theoretically influence malignant transformation is the biologic behavior of melanocytes. If melanocytes have the potential of becoming malignant the larger their number the greater the risk of malignant transformation. It is also known that GCMN located on the vertebral column and scalp have a greater risk of being associated to meningeal melanosis which is a potential source of malignant transformation.</p> <p>GCMN with the exception of size (larger lesions are more prone to develop malignancy) it is not possible to predict, on clinical or histopathological features which will remain benign.</p> <p>A few years ago we tried to find a marker of risk of malignant transformation measuring melanocytes DNA content, anuploidy, and cell cycle by flow cytometry in 28 GCMN who did not develop melanoma. Significant differences were found among the two groups (Pediat Develop Path 2001; 4: 73-81.</p>
Management	<p>Management of GCPN is a controversial issue. The following therapeutic possibilities exist : observation, split skin grafts, serial surgical excision (subcutaneous inflatable expanders), dermoabration, chemical peel, and lasers.</p>
Observation	<p>Arguments supporting observation :</p> <p>Malignant transformation in GCMN may develop from extracutaneous melanocytes (meninges). Most treatments are traumatic and results are often poor.</p>
Surgery	<p>Often require multiple interventions, results even with the use of tissue expanders are often poor. Cost-benefit is often negative.</p>
Dermoabration	<p>Is traumatic, partially removes pigment but not hair, risk of scarring.</p>
Curettage	<p>In large (G1 to G3) lesions curettage results are similar to those obtained with dermoabration.</p>
Peels	<p>Phenol peel is potentially toxic (heart, kidney) partially removes pigment but not hair, risk of scarring. T.C.A. peel is less effective than phenol peel but is not toxic.</p>
Laser	<p>Are partially effective, require several applications, are expensive.</p>
Grafts	<p>Keratinocyte grafting is in most cases esthetically unacceptable and expensive.</p>