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Case report

# CASE REPORT OF GIANT CONGENITAL MELANOCYTIC NAEVUS

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## ABSTRACT

This is a case of giant congenital melanocytic nevus, associated with a single lipoma & multiple satellite lesions. Biopsy report showed skin with groups of nevoid cells extending into underlying fat, intimately associated with the lobules & surrounding many blood vessels. The systemic examination was normal. This is a typical case of a giant congenital melanocytic nevus with satellite lesions and a lipoma association. As the surface area is greater than 40cm in diameter; the risk of developing melanoma is higher.

**Keywords:** Bathing trunk nevus, Satellite lesions, Lipoma

## INTRODUCTION

Congenital melanocytic nevi are benign proliferation of melanocytes located in different skin levels that have existed since one's birth. However, the same lesion can also appear within the first 2 years of life, in this case it's called "nevus tardive"[1] Congenital melanocytic nevi are more common in Asian and Black children.[2] The incidence is between 1-2% of newborns[3]. A giant congenital melanocytic nevus has a diameter of 20cm or more & is very rare. One study including over 5000,000 newborns has shown that only one baby in 20,000 has a nevus with a diameter larger than 10cm.[4] It is reported to be associated with leptomeningeal melanocytosis, meningomyelocoele and spina bifida, ocular malformations and glaucoma, auricular malformation,angiomatous lesions,bone atrophy, musculoskeletal deformities, neurofibromatosis, diffuse lipomatosis, vitiligo etc[5] there are other case reports of association of garment nevus with lipoma, neurofibroma & spina bifida occulta.[6,7,8] A case of giant naevi with hydrocephalus was reported in mexico.[9] It is said to occur in the epidermal naevus syndrome.[10]A well substantiated association with neurofibromatosis is documented.[1]

## CASE REPORT

A 5-year-old girl second born by normal vaginal delivery (birth weight was 2.8kgs & cried immediately after birth) to non consanguineous parents hailing from Papum Pare in Arunachal Pradesh, India, presented with a large asymptomatic hyper pigmented lesion encompassing the entire trunk involving the chest, abdomen, back, buttocks and genitalia

covering more than 40% of the body surface area, present since birth [figure 1]. It has been growing in size with age. The lesion was flat initially and gradually started becoming thicker. Distant to the giant nevus, multiple satellite nevi [figure 2] started developing gradually on the upper and lower limbs, that was not present at the time of birth. Hypertrichosis was seen all over the large pigmented lesion converging toward the midline [figure 3]. Scalp, mucosa, palms, soles and nails were normal. There was no significant family history. The patient's cause of worry was the cosmetic disfigurement caused by the large hyper pigmented lesion & the swelling in the left loin which has been gradually increasing in size. [Figure 1] The swelling was first noted by the patient's mother approximately 4 years ago, on examination it was 8×6 cm in size, soft in consistency, non tender, mobile& skin pinches able over the swelling. The diagnosis was made clinically. General, systemic, ophthalmologic & Musculoskeletal examinations were normal. The X-Ray spine & MRI Brain were both normal. Basic blood work & urine routine were normal .The USG of the swelling in the posterior left flank was done using a linear transducer & showed a diffuse well defined thickening of the skin and subcutaneous plane at the site of swelling, no obvious vascularity was noted within, the underlying muscular plane appears normal. Under GA, the swelling in the left loin was sent for excisional biopsy.

Histopathology of the swelling showed, skin with underlying groups of cells with pigmentation [figure 4], which extends into the underlying fat [figure 5]. The nevoid cells are seen surrounding blood vessels in the subcutaneous fat. The cells extend into the fat intimately associated within the lobules & surround many blood vessels. The patient was advised to report immediately if changes in symmetry, color, border, diameter of the lesion or any symptoms of itching, oozing or swellings from the lesion were noted. We recorded all the pigmented moles on her body and are following them up periodically to identify malignant change by the "mole-mapping technique"[11,12]. She was advised to undergo a series of diode laser treatments to treat the hypertrichosis. Patient is under Psychiatric counseling to deal with the cons of this disfiguring disease.

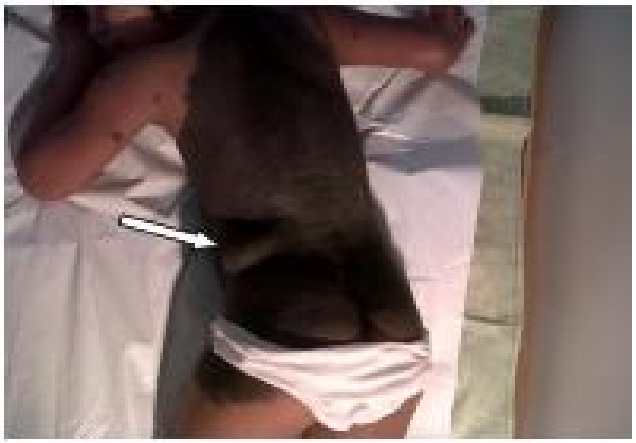

**Fig 1:Giant congenital naevus(>40% BSA involvement) Associated lipoma.**

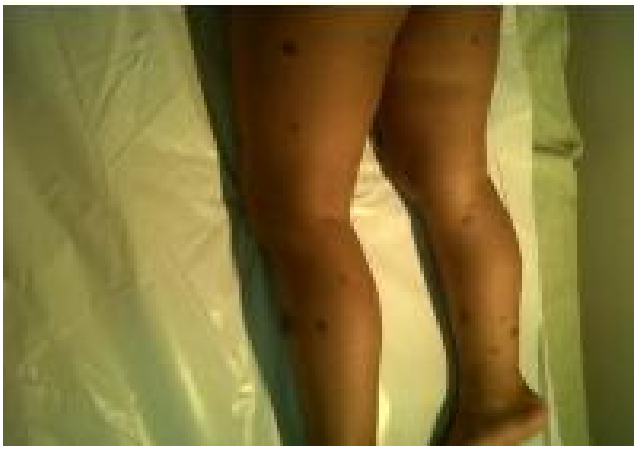

**Fig 2:Satellite lesions distant to giant naevus.**

**Fig 3:Hypertrichosis showing midline convergence.**

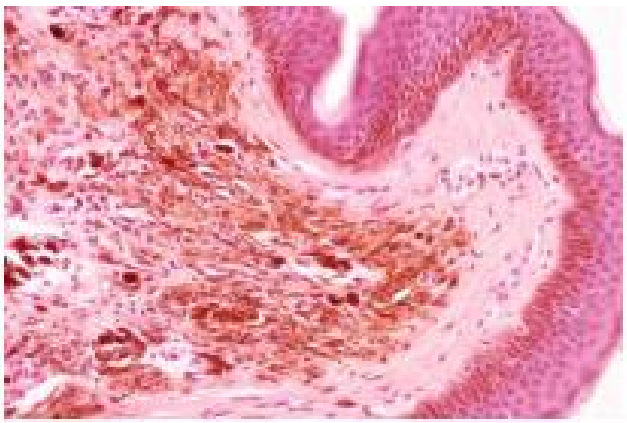

**Fig 4: Histopathology skin with underlying groups of pigmented cells extending into underlying subcutaneous fat. (10x)**

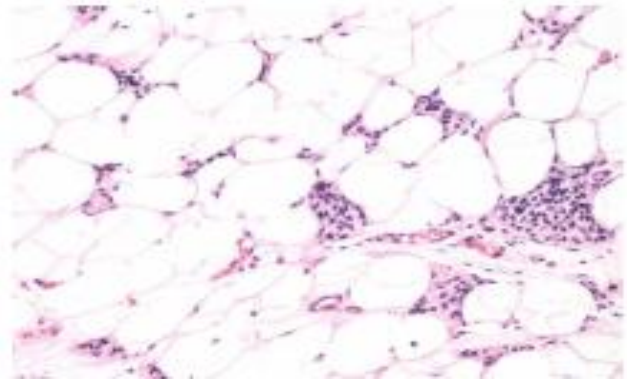

**Fig 5: The nevoid cells are seen surrounding blood vessels in the subcutaneous tissue, extending into the fat intimately associated within the lobules & surround many blood vessels. (40x)**

## DISCUSSION

Synonyms: Giant garment nevi, bathing trunk nevi, Giant hairy nevus

**Incidence/prevalence:** Giant congenital nevi are very rare. One study including over 5000,000 newborns has shown that only one baby in 20,000 has a nevus with a diameter larger than 10cm.[4]

**Classification:** The American national institutes of health (NIH) are used to categorize naevi according to size – small are under 1.5cm in diameter, large between 1.5 and 20 cm and giant naevi having a diameter of 20cm or more.[13]

**Association with lipoma**: The central nervous melanocortin system regulates peripheral lipid metabolism via the beta adrenergic receptors of the sympathetic nervous system. This autonomic outflow shifts substrate metabolism to modulate energy storage and adiposity. This mechanism explains the occurrence of lipoma in giant congenital melanocytic nevus[14].

**Clinical features:** Congenital nevi are usually intensely pigmented macules with hypertrichosis (terminal hair which is darker and more coarse than normal hair with a tendency to be concentrated in the central midline) at birth .As the child grows the nevus grows in proportion and the surface may become warty or get thrown into folds & nodules can develop within a large nevus.

The bathing trunk nevus is very rare but poses a definite cosmetic problem for the patient. Commonest site is the lower back and thigh area & a surface area of a diameter greater than 20cm is involved. This may be accompanied by large numbers of smaller congenital nevi present in areas distant to the giant nevus. An increase in number may develop over time. They are called satellite nevi.

The hairy component, which occurs in 95% of the lesions, tends to become more prominent in late childhood & seems to be more centered and dense toward the midline, but at this stage the nevus ceases to thicken and may become paler. The hair growth pattern usually has a vortex distribution, often centered on the midline in the giant nevus in the back. It is crucial to rule out CNS, ocular and musculoskeletal deformities.

**Histopathology:** In congenital nevi, melanocytes are typically located on the lower two-thirds of the dermis. Occasionally they extend into subcutaneous tissue, with isolated cells or groups of cells within the reticular dermis collagen fibres, with a tendency to be located around cutaneous appendages. [5]

**Prognosis & complications:** Sometimes there is spontaneous regression during childhood or adolescence. This premature regression is commonly due to immunological mechanisms .Rarely, it may result in complications like Superficial spreading melanoma, neurocutaneous melanosis , raised intracranial pressure, hydrocephalus, or space occupying lesions, rapidly growing ulcerative tumor called nodular

proliferative neurocristic hamartoma (neuroectodermal & ectomesenchymal origin[15] .

**Genetic counseling:** It is very important to reassure the parents of a child with a giant melanocytic naevus that, the risk of recurrence is very low in subsequent pregnancies but a few rare cases have occurred which suggest an inherited tendency.

**Diagnosis &Treatment:** MRI scans should be done in babies with nevi over the cranium or spine to exclude significant leptomeningeal melanocytosis.[16] Regular neurological examination is crucial. The treatment aims at improving the cosmetic aspect and balancing the risk of melanoma by excision which may not be possible in giant melanocytic nevi & periodic check up to keep an eye out for changes of malignant transformation. Dermabrasion may help with a mild reduction in pigmentation.

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