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Giant Congenital Melanocytic Nevus In A Neonate

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Abstract

A three hour old baby presented at the special care baby unit of the Ladoke Akintola University of Technology Teaching Hospital, Ogbomoso with giant congenital melanocytic nevus. This rare case is discussed with a view to highlight challenges associated with managing such patients.

Introduction

Nevus is derived from the Latin word knee-vus, meaning a birth mark or mole. Congenital nevus, by definition is a tumour like malformation of the skin or mucous membrane present at birth, while acquired nevus is usually seen after 8 months.¹ The acquired nevus is more commonly encountered in contrast to congenital nevus. Congenital nevi develop between the second and sixth month of gestation and are primarily neuroectodermal in origin. Arrests in the migration of congenital nevus in utero may result in leptomeningeal melanocytosis.² Melanocytic nevus contains melanocytes which are responsible for the pigmentation of the skin. Embryologically melanocytes are found in utero as early as 40days.¹

Congenital melanocytic nevi occur in approximately 1% of newborns.² They present in various sizes viz - small (less than 2cm), intermediate (between 2 and 20cm) and giant (above 20cm).² Giant congenital melanocytic nevi (GCMN) are rare and they may undergo malignant transformation.^{2,3} Benign lesions may be ignored or managed with pulsed laser therapy

for cosmetology, while malignant should be managed surgically.^{2,4} To my knowledge the present case is the only known case of GCMN in a Nigerian. The clinical features of the neonate and the challenges posed in the management are discussed.

Case Report

A three hour old male baby was referred to the special care baby unit of the Ladoke Akintola University of Technology Teaching Hospital, Ogbomoso because of multiple hyper-pigmented patches on the face and limbs with the most extensive single lesion extending from the back of the neck to the buttocks. The lesions were seen on the posterior trunk, lower limbs and face from birth. The lesions on the back were black and confluent and were described to be coarse, while the other lesions on the buttocks and lower limbs were scattered and oval.

The baby was delivered in a mission hospital at term and was referred immediately for expert management because of the skin lesions. Delivery was uncomplicated via the vagina and the birth weight was 3.5kg. The mother is 27 years old and she received ante-natal care at the same hospital where the baby was delivered. Only the routine prescribed drugs such as ferrous sulphate and folic acid were taken during pregnancy. Ante natal period was not eventful and there was no history suggestive of maternal illness, use of steroids or irradiation during pregnancy. There is no similar skin lesion in the

sibling or the parents.

On examination the baby was well and not ill, pale, cyanosed or jaundiced. A full length of 50cm was recorded while the weight was 3.5kg. There was no abnormal facies and the axillary temperature was 36.4°C. Multiple hyperpigmented macules of varying sizes from 2-10cm were observed on the lower limbs, buttocks, lumbar and sacral areas. A few hyper pigmented macules with diameters varying from 2-10cm were observed on the upper limb scalp and face. The back of the child was uniformly hyperpigmented, but the lower quarter of the posterior trunk was partially spared. Pictures of the neonate are shown in figures 1 and 2.

Figure 1



Figure 2



The vital signs were normal and the baby had a heart rate of 158 beats per minute, while the respiratory rate was 46 cycles per minute. Neurological examination was also normal with the baby having a head circumference of 35cm. Muscle tone, sucking and other neonatal reflexes were normal. Examination of other systems revealed no abnormality.

An assessment of Giant congenital melanocytic nevus was made and the patient was admitted to the special care baby unit for further evaluation. Full blood count revealed a packed cell volume of 51%, total white blood cell count of 12,600/mm³ and a differential neutrophils and lymphocyte count of 72% and 28 percent respectively. Blood sugar at presentation was 87mg/dl. Hepatitis B virus screening for the baby and retroviral(HIV) screening for the mother were both negative.

Other tests requested were a trans-fontanelle ultrasound, computerized tomography of the brain and the paediatric dermatologist was informed about the child. The baby was discharged home in a stable condition on the fourth day of admission due to financial constraints. At discharge the parents were counselled about the condition of the child and the need for a further detailed evaluation in view of the risk of leptomeningeal melanocytes and of malignant transformation. The parents however defaulted from the clinic and could not be reached through the telephone communication. The social services department of the hospital could also not trace the child, due to logistics reasons.

Discussion

Giant congenital melanocytic nevi are rarely seen in neonates.^{2,4} The available scanty literature on them are mainly from developed countries and are case reports.^{1,3-6} Probably, the present case reported may

well be the first in a Nigerian. Previous studies on dermatoses in Nigerian neonates suggest that GCMN does not affect this populace.^{7,8}

A number of challenges were experienced in the management of this case because of the constraints in our setting. The first and major huddle was parental financial inadequacy. This hindered full evaluation. Computerized tomography and magnetic resonance image of the brain and spine could not be carried out to rule out leptomeningeal melanocytosis. This was indicated because of the location of the lesions on the face and spine, which are known to be associated with leptomeningeal melanocytosis.^{1,2,5} Proper management of this child required a careful follow up of the melanocytic nevus in order to detect malignant changes. The financial constraints on the part of the parents might probably have contributed to the refusal of the parents to stay long enough in hospital for proper review and failure to attend follow up later.

Coping with a newborn with multiple and obvious disfiguring lesions on the skin, might be a herculean social challenge, especially when dealing with a rare disease without support groups to help or encourage the parents. A previous study showed that a number of Nigerian parents whose children have gross congenital malformations discharge them against medical advice from health facilities.⁹ Death may ensue from complications of the disease or from infanticide, which may seem to the parents an exit from the problem.

Proper management of this kind of case in the future will need the coming together of the government, the health care service providers, philanthropists, the public media and the public.⁹ The government can improve the socio-economic condition of its citizenry by resuscitating the economy and improving the funding and equipments supply to its

health facilities. More attention should be put on the health insurance scheme such that its operational base would be expanded to allow for treatment of such conditions. Donations from philanthropists and public spirited individuals can help and a realistic improvement of the health care delivery services, especially the funding, can go a long way in solving the problems of financial constraints. The public media will be of immense assistance in informing the public about such special cases that need financial assistance.

The managers of health care facilities should spend funds judiciously and give financial concessions to indigent patients. The health facilities also need to see that all their departments function effectively. The social services definitely have a part to play in the management of children in the community. The social services should be able to provide the essential link that can make follow up and / or interventions possible.

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