

Giant Congenital Nevocellular Nevus and Natal Teeth

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Congenital nevocellular or nevocellular nevi (CNN) are considered to be hamartomatous lesions of melanocytes apparent at birth(1-3). Though CNN has an incidence of 1% of all newborns, its spectacular variant- the giant or bathing trunk nevus is extremely rare, occurring once in 5,00,000 live births(1). Giant CNN is of devastating consequence due to the cosmetic disfigurement and the great risk for malignant transformation. We report a neonate presenting at birth with giant CNN and natal teeth and the problems in management of such cases.

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Case Report

A 2.44 kg girl was born by spontaneous vaginal delivery at 37 weeks gestation to a 30-yr-old second gravida mother with one live and healthy 4 yr-old-boy. The mother was diagnosed to be a non-insulin dependent diabetic one yr before the present pregnancy and was on diet control. Since 16th week of gestation, she was on regular insulin therapy with good control of the diabetes.

The baby was noted to have deep brown, pigmented hairy verrucous lesions at birth. There was single large lesion in a bathing suit pattern (25 cm across) encircling the whole lower half of the trunk (*Fig. 1*). There were also similar lesions of varying sizes (2 cm x 3 cm to 5 cm x 7 cm) distributed over scalp, forehead and extremities. Nails and mucus membranes were normal. The baby had 2 partially erupted natal teeth (incisors) on the lower jaw. There were no other malformations. Ultrasonograms of the cranium and abdomen were normal. In view of the extensive involvement of the skin, no active management was considered in the neonatal period. Parents were counseled about the nature of the lesions and advised regular follow-up. The baby also developed jaundice on day 3

of life which required phototherapy from day 5 to 7 as serum bilirubin rose to 16 mg/dl. At 3 months of age, the baby's weight was 4.2 kg; there were no seizures or developmental delay. The skin lesions were of the same size and texture with no evidence of progression.

Discussion

CNN is defined as a lesion consisting of pigmented or non-pigmented melanocyte with or without the participation of nevus elements(1). CNN is classified as small, intermediate or giant types according to their size. Lesions measuring less than 1.5 cm in the largest diameter are small and those between 1.5 to 20 cm intermediate (1,2). Giant or bathing trunk or garment CNN are either more than 20 cm in the greatest diameter, more than 120 sq cm in area or which cannot be excised *in toto* with primary suture closure in a single operative



Fig. 1. Giant congenital melanocytic nevus on back with smaller lesions on buttocks and extremities.

procedure(1). Our patient had all the types of CNN.

Apart from the cosmetic disfigurement giant CNN may develop into melanoma and scalp lesions may be associated with leptomeningeal melanocytosis resulting in seizures, features of raised intracranial tension and development delay(1,2). The lifetime risk of malignant transformation in giant CNN is 6.3% and the relative risk is increased by 17 fold(3). Melanoma is diagnosed within the first 3-5 years of life in about 50% of cases and giant CNN are estimated to account for 40-44% of all melanomas in children(3). Hence total excision of giant CNN is recommended usually between 10-14 months of life. Such a management was, however, not possible in our patient because of the extensive spread of the lesion.

Recently, new techniques have been used in the treatment of giant CNN(2). In the dermabrasion procedure, the lesion is given a 'deep shave' preferably in the first week of life with the hope of removing as much nevus cells as possible(1,2). The adnexal melanocytes are, however, not removed and hence palliation against melanoma is doubtful(1). Another procedure is to expand and harvest the epidermis of normal skin of the patient using inflatable balloons and using the same for skin grafting. Despite these procedures, total excision of the giant CNN may be difficult to achieve and close follow up of these lesions is mandatory. Follow-up visits are recommended at 3-6 month intervals for the first 5 yr coupled with nevus photography and biopsy of new or enlarging lesions(4).

Small and intermediate sized CNN also bear a considerable risk towards melanoma(3,5). The relative risk is increased by more than 3 fold and risk upto 60 years of life is 0.8-4.9%(3). Melanoma, however, rarely if ever occurs earlier than 12 yr of age. Hence, excision has been recommended electively under local

anesthesia once the child is 10-12 yr old(2,3)

The management of jaundice in this baby presented a peculiar problem. The effectiveness of phototherapy was in doubt because of very limited area of normal skin. Phototherapy has a mutagenic effect in tissue culture cells, and the potential hazards of using phototherapy in such a situation are unknown. Phototherapy has not been reported to be a risk factor for melanoma(4).

Natal teeth have been described in various syndromes though none of these have giant CNN as feature(6). The association of natal teeth and giant CNN has previously not been reported. Embryologically odontoblasts and melanoblasts have a common origin from neural crest cells(7) and it is likely that the two may be related. However the possibility that these conditions were unrelated and their occurrence incidental cannot be excluded.

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