Giant Congenital Nevocellular Nevus and Natal Teeth

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Congenital nevocellular or nevomelanocytic nevi (CNN) are considered to be hemartomatous 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 nevous elements(l). CNN
is classified as small, intermediate or giant
types according to their size. Lesions
measuring less then 1.5 cm ii the largest
diameter are small and those between 1.5
to 20 cm intermediate (l,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
procedure(l). 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(l,2). The
lifetime risk of malignant transformation in
 giant CNN is 6.3% and the relative risk is
increased by 17 fold(l,2). 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(l,2). Hence total
excision of giant CNN is recommended
usually between 10-14 months of life. Such
a management was, however, not be
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(l,2). The
adnexal melanocytes are, however, not
removed and hence palliation against
melanoma is doubtful(l). 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.

REFERENCES