Congenital Bathing Trunk Nevus with Meningomyelocele

Sir,

A 3-day-old female neonate, first born to non-consanguineous parents with unsupervised antenatal period was brought with paucity of movements of the lower limbs and a black skin lesion with ulcerated mass protruding through the midline in the lower back noted since birth. She did not have urinary dribbling or retention. At the time of presentation, child was noted to have elevated hyperpigmented lesion with thick hair measuring 15 × 10 cm involving entire posterior torso suggestive of congenital bathing trunk nevus surrounding the meningomyelocele as shown in Figure 1. Initial screening in form of the ultrasound of cranium was normal. Child underwent meningomyelocele repair. Currently child is asymptomatic; and, magnetic resonance imaging (MRI) brain has been planned on follow up.

Congenital bathing trunk nevus is a rare lesion with female preponderance and an incidence of one in 2–5 lakh births. Though named congenital as it is noticed soon after birth, it is not hereditary and occurs due to somatic mutations in NRAS (chromosome 1p13) and BRAF gene.[1] A strong association exists between congenital giant melanocytic nevus, neurofibromas, lipomas, and neural tube defects (NTD).[2]

Congenital nevus is said to be giant when it covers >2% of body surface area or 5 cm in neonates or 20 cm in adults. Lesions may be small in infants and grow proportionately as the body grows. It can occur anywhere on the body with predilection over trunk and is associated with pigmented satellite lesions. It may be flat or raised, hypo- or hyperpigmented, and color may change over time with majority (95%) being associated with thick hair in the lesion. Age of presentation varies; adult presentation is seen when they develop multiple nodules (lipoma, neurofibroma) or when it ulcerates (sclerotic variant of melanoma) while adolescents and girls present for cosmetic reasons.

Neurocutaneous melanosis (NCM) and malignant melanoma are the two most important complications.[3] NCM is characterized by proliferation of melanocytes present in the meninges of brain and spinal cord and present as headache, seizures, movement abnormalities, and brain tumors. Risk of malignant melanoma is 2–5 times higher and occurs in the first 5–10 years of life.[4] High-risk lesions for malignant transformation include those with size >20 cm, satellite lesions and multiple (>3) lesions. Bathing trunk nevus needs continuous follow-up to monitor for signs of malignant transformation. Surgical excision of high-risk lesions is best performed in early childhood.[5] Many non-surgical procedures have been tried and each has its own limitations. Serial excision with tissue expansion is the mainstay of treatment of bathing trunk nevus.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and

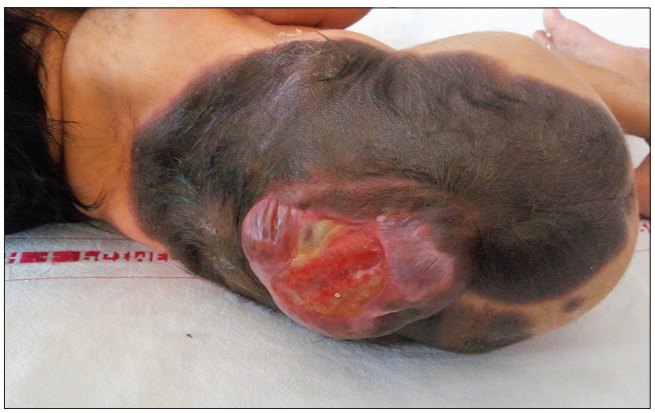

Figure 1: Hyperpigmented giant hairy nevus surrounding the meningomyelocele with satellite lesion noted over buttock area

other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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