Congenital melanocytic nevus is deposition of pigment producing cells of melanocytic lineage in the dermis. We present an extremely rare case of congenital melanocytic nevus with a scrotal mass associated with deposition of melanin in the brain. The mass may mimic like a testicular tumour on clinical presentation.

Keywords: Congenital melanocytic nevus, neurocutaneous melanoses, pediatric scrotal mass

INTRODUCTION

Congenital melanocytic nevus (CMN) is a rare disorder with an incidence of one in 20,000. The incidence of giant type (>20 cm diameter) is not known.[1] These nevi often have a garment-like distribution and commonly referred to as bathing trunk nevus. Melanocytic lesions on the genital area are rare and poorly documented, occurring more frequently on the vulva and less often on the perineum and male genitalia. Most commonly reported findings of associated central nervous system (CNS) involvement includes melanocytic accumulation in the temporal lobe, amygdala, thalami, and cerebellum and enhancement of leptomeninges in the brain and spine.[2] Here, we present a rare and unusual case of congenital large melanocytic nevus, with inguinoscrotal mass and CNS involvement. To the best of our knowledge, this has not been reported in literature yet.

CASE REPORT

A 1-month male baby was brought to our hospital with complaints of dark pigmentation of the skin below the neck and inguinoscrotal swelling since birth. The swelling was firm and irreducible, and the right testicle could not be palpated separately from the swelling. The left testis was normal [Figure 1]. The nevus was found to be involving both the dorsal and ventral aspects of the body. On the ventral aspect, it extended from below the nipples up to the bilateral knee joints, and on the dorsal aspect, it was extending from below the axillary region up to the bilateral knee joints. There were multiple satellite lesions in the bilateral upper and lower limbs. The CNS examination and serum alpha fetoprotein levels were normal. An ultrasound study was done, in which the testis could not be separately visualized from the swelling.

Further, magnetic resonance imaging (MRI) showed a large, heterogeneous mass measuring 8 cm × 5.2 cm completely engulfing the scrotal sac, shaft, and root of the penis. The right testis was visualized and was normal. The left testis was not visualized separately [Figure 2]. MRI brain showed neuroparenchymal melanin deposition in the amygdala.

With a strong suspicion of paratesticular tumor in mind, we took the patient for inguinal exploration. Intraoperatively, the mass was seen closely adherent to the penile shaft and along the penile urethra. There

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was no clear surgical plane which made the dissection difficult. Both the testes were normal and separate from the swelling. It was difficult to dissect the mass from the urethra and penis \textit{in toto}. Postoperatively, the child did well and the catheter was removed on the postoperative day 3. To our surprise, the histopathological findings were suggestive of neurofibroma.

The child is under follow-up for 3 years and there is no recurrence. The skin lesions are planned for biopsy to rule out neurocutaneous melanosis (NCM). The child is currently under follow-up with the dermatology department for the management of nevus.

**DISCUSSION**

Congenital nevi are pigment-producing cells of the melanocytic lineage that are clustered in the dermis and produce pigment at birth.\cite{Amir}

\cite{Amir} et al described a baby born of twin pregnancy, out of which one baby had a giant pigmented nevus associated with a scrotal swelling and the other baby was normal.\cite{Amir} The study focussed mainly on the genetic aspect of the disease and concluded that it does not transmit genetically. Melanocytic nevus is more common in the trunk and seen less frequently on the genital or perineal region. Bathing trunk nevus may be associated with vitiligo, neurofibroma, diffuse lipomatosis, NCM, structural brain malformations, hypertrophy of skull bones, and skeletal asymmetry. The genital pigmented lesions arise mainly on the vulva. Although relatively rare, it may be seen on perineum, pubic region, and male genitalia (penis and scrotum). Friedman and Ackerman first described an atypical melanocytic nevus of genital type in a series of seven unusual vulvar nevi.\cite{Friedman}

NCM is a rare neuroectodermal dysplasia defined by large or multiple congenital cutaneous nevus which is seen associated with meningeal melanosis or melanoma. Revised criteria for diagnosis was proposed by Kadonaga and Frieden in 1991:  

1. Large nevus (that means $>20$ cm in adults, $>9$ cm on the infant scalp, or $>6$ cm on the infant body)
2. Multiple nevus (greater than or equal to three)
3. No evidence of cutaneous melanoma, except in cases where the meningeal lesions are histologically benign
4. No evidence of meningeal melanoma except in cases where the cutaneous lesions are determined benign.

The clinical features of NCM tend to appear in early infancy with a median age of 2 years. However, it may present as late as 20 years. In the present case, MRI findings were similar to the literature already published, showing high signal intensities on T1 in the right amygdala (parenchyma deposition) which is a frequent location for melanin deposition.\cite{Kadonaga} Other neurological associations comprise communicating hydrocephalus, syringomyelia, arachnoid cysts, tumors (including astrocytoma, choroid plexus papilloma, ependymoma, and pineal germinoma), and malformations such as Dandy–Walker or Arnold–Chiari.

Giant CMN with scrotal mass can be a diagnostic dilemma, and approach to it can be tricky in centers with limited resources. The challenges that we faced included lack of literature regarding its management, need for a multidisciplinary approach, namely, pediatric surgeon, dermatologist, pediatrician, pathologist, radiologist, and psychologist, and social challenge for the parents to overcome the social stigma attached to it. The natural course of the disease is not known due to a lack of literature. Hence, a regular follow-up is recommended. The management of bathing trunk nevus itself is often challenging. The literature review does
not give satisfactory treatment for nevus of such big dimensions, and the results are not encouraging. Various treatment options include YAG laser, curettage, and staged excision and grafting along with the use of tissue expanders. In our case, we could not have a histological diagnosis of the brain lesions, so NCM was kept as a provisional diagnosis. The regular counseling of the parents with psychologist and their understanding about the disease and the line of management are important in its management.

Since it is not possible to differentiate a paratesticular mass as benign or malignant, early surgical intervention should be done. As there is limited literature available, we recommend regular follow-up to look for recurrence.

**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 other clinical information to be reported in the journal. The patients understand that their names and initial s 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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