Giant Congenital Pigmented Nevus of Scalp

4-year-old female admitted for was bronchopneumonia. The child was also noted to have a scalp neoplasm, which was present since birth but had gradually increased to the present size (8X5.5 cm), with thick and wrinkled covering skin (Fig. 1). The lesion was located in the occipital region, was non-pulsatile, with no increase in size on crying and no transillumination, thus ruling out a Neural tube defect. The child had been developing normally with no neurological complaints or similar family history. The patient also had a few pigmented nevi on the back and the extremities. Neuroimaging of the brain was normal and the biopsy showed numerous melanocytes in an intradermal location with occasional melanophages and no cellular atypia. Child was referred to the surgeons for operative management.

Giant congenital pigmented nevus shares the pathological characteristics of common nevi and may invade the skin of any region of the body. Its clinical importance is that its presence on the head and neck region is usually complicated with neurocutaneous melanosis (NCM), a rare phakomatosis consisting of congenital abnormal pigmentation of the skin and meninges, and neurological features like epilepsy, developmental delay, or focal neurological deficits may be present. The meningeal lesions may undergo malignant change, with a very poor prognosis. In



FIG. 1 Giant pigmented nevus of scalp.

NCM, the congenital melanocytic nevi are either giant (>20 cm in greatest dimension) or multiple (>3). Diagnosis is based on clinical features and pathological examination. Treatment is radical or staged excision. It must be differentiated from cutis verticis gyrate, in which the skin lesions are symmetric, and complicated with proliferation of the skin, hyperplasia of the periosteum and bone matrix.

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Large Cavernous Hemangioma

Cavernous hemangioma covering half abdomen and thorax in a neonate is extremely rare. A female neonate born at 32 weeks with a birth weight of 2.3 kg had a large cystic lesion covering left hemithorax,

hemiabdomen with skin over the lesion showing capillary hemangioma (*Fig.*1). Baby had tachycardia which settled in 2 hours and she was stable hemodynamically for initial three days on feeds. Over next three days she developed a necrotic patch which spread centrifugally and developed Kasabach Merritt phenomenon (*Fig.* 2). Despite supportive treatment, propronolol and steroids, the child succumbed to DIC.





Fig. 1 (a, b) Large hemangioma.

This lesion can present with Kasabach Merritt phenomenon which is difficult to manage. Surgical excision for small superficial hemangioma while cryoreduction and graded interferon alpha 2a for large hemangioma remains treatment of choice. Propranolol and steroids might reduce the size.

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Cecoureterocele

A 10 months old female child was brought with the complaints of a mass coming out through her urethra (*Fig. 1*) when the child passed urine since last two months. The child was not able to pass urine whenever the mass prolapsed. There was no history of fever, hematuria, chronic cough or constipation. A diagnosis of cecoureterocele was made based on the presence of cystic mass at the external urethral meatus and urinary retention. Ultrasound showed a cystic mass at the end of ureter with or without hydronephrosis and voiding cystourethrogram demonstrated smooth round filling defect.

An ureterocele is a congenital saccular dilatation of the terminal portion of the ureter. Cecoureterocele is an uncommon type where the ureterocele is elongated beyond its orifice by tunneling under the trigone and the urethra .

Common complications include urinary tract infection, symptoms of obstructive voiding, urinary



Fig. 1 Cecoureterocele.

retention, failure to thrive and abdominal pain. Untreated, these may lead to hydronephrosis or pyonephrosis, A cecoureterocele presenting as a prolapsing mass at urethral meatus should be differentiated from urethral prolapse presenting with bleeding, spotting, dysuria, urinary frequency, introital pain, and urinary incontinence or retention; and sarcoma botryoides which presents as a firm grapelike vaginal mass protruding through the introitus with occasional bleeding.