

## Congenital Ectropion

A six month old girl presented with eversion of the upper eyelids since the neonatal period. The eversion was more prominent on crying. She also had clinical features of Down syndrome like generalized hypotonia, upward slant of eyes and a protruding tongue (*Fig.1*). Skin examination was normal. A clinical diagnosis of upper eyelid congenital ectropion was made. As the ophthalmological



**FIG. 1** Bilaterally everted upper eyelids in Down syndrome.

examination revealed no exposure keratopathy she was managed conservatively with corneal lubricants. On follow-up at six months, her condition is stable.

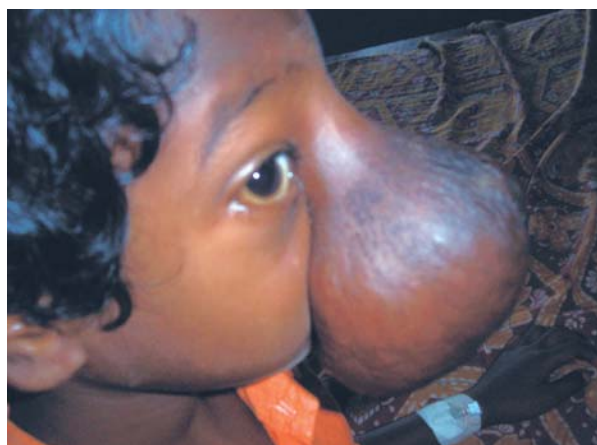
Ectropion is also associated with conditions like collodion baby where the neonate is covered at birth by a thick membrane, flattening of the ears and nose and o-shaped fixation of lips. Some forms of chronic dermatitis like epidermolysis bullosa also cause ectropion in the long run. Bells palsy and facial nerve palsy were ruled out by the absence of facial palsy. Eyelid cysts, tumors and scarring due to trauma were ruled out by their absence. Though the ocular findings in Down syndrome are varied, ectropion is one of the rarest. This condition may resolve spontaneously with conservative treatment or may need surgical intervention to prevent complications if conservative treatment fails.

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## Frontonasal Encephalomyelocele

A seven year tribal male child presented with a painless swelling on the upper part of the nose since birth which had gradually increased to the present size (*Fig.1*). He has average intelligence and was otherwise asymptomatic. He was being worshipped as Lord Ganesha because of the typical appearance.

Examination revealed a large, soft swelling of 10×8×7 cm in size arising from the right side of the root of the nose. Skin over the swelling was



**Fig. 1** A large mass of size 10×8×7 arising from the root of the nose.

hyperpigmented. There was cough impulse but no trans-illumination. A provisional diagnosis of frontal meningocele was made. Differential diagnoses included dermoid cyst, hemangioma, neurofibromatosis, rhabdomyosarcoma and encephalocele. Dermoid cyst, hemangioma, and neurofibromatosis are easily distinguished by their peculiar external appearance. Rhabdomyosarcoma is a rapidly progressing tumor; a relatively benign appearance of this tumor after a long period is against its diagnosis.

CT scan confirmed the diagnosis of right frontonasal encephalomyelocele with partial corpus callosal agenesis. He was referred to a higher center for surgical management.

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## Ichthyosis Linearis Circumflexa

A 7 years old girl, born of a non-consanguineous marriage, presented with multiple asymptomatic dry and scaly patches over her body since infancy. The patches were constantly changing their shape and pattern, resolving in 2-3 weeks with mild transient hypopigmentation, and were recurrent. Lesions tended to get better in summer with aggravation in winter. On examination, she had multiple, erythematous annular and polycyclic scaly patches with double-edged scales at the periphery of the lesions. They were present bilaterally and in asymmetric manner over face, neck, trunk and extremities. Hair, nail and mucosa were normal (**Fig. 1**). Clinically the child was diagnosed as Ichthyosis linearis circumflexa (ILC). Routine investigations, KOH mount and Wood's lamp examination were unremarkable. Histopathological findings were consistent with this diagnosis. The child was treated with oral isotretinoin 10 mg a day for 1 month and lesions resolved within 2 weeks. No new lesions have appeared since then. ILC is a rare autosomal recessive disorder of keratinisation. Association of ILC, hair abnormality and atopic diathesis is called Netherton's syndrome (NS). Common differential diagnoses include erythro-dermic atopic/seborrhoeic eczema, staphylococcal/ Candida infection, tinea corporis and familial peeling skin syndrome. In neonatal period, intensive medical, nursing and nutritional care must be available to



**FIG. 1** Lesions distributed symmetrically on trunk.

manage erythroderma and its complications. In older children, regular emollients (e.g. ammonium lactate 12%) are helpful.

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