Now the patient is waiting for anal transposition.

Discussion

Vaginal obstruction due to imperforate hymen with resultant hematocolpos is mentioned in writings of Hippocrates and Celsus but hydrometrocolpos, a comparable condition was not reported prior to 1956(6).

Imperforate hymen presenting as spherical mass protruding out of introitus in neonatal period is quite unusual. Association of hydrometrocolpos with anorectal malformation is also quite unusual(2-5).

Prenatal effects of maternal hormones causing excessive secretion of uterine and cervical glands is an additional necessary factor for development of hydrometrocolpos(3,7). The hydrometrocolpos may be associated sometimes with anorectal malformation(2-5) as in our case.

Clinically the lesion presents with either (*i*) Anterior pressure with urethral obstruction or (*ii*) Posterior pressure with rectal obstruction or (*iii*) upward pressure and intestinal displacement and sometimes respiratory embarrasment(8,9).

Our case was unique as it presented with failure to pass meconium possibly due to pressure effects. However, there were no back pressure effects on urinary tract due to outward and downward ballooning of imperforate hymen, which has not yet been reported.

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Adrenoleukodystrophy

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Adrenoleukodystrophy is a sex linked autosomal recessive disorder in which progressive CNS degeneration as accompa-

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nied with adrenocortical insufficiency. The present case report of this rare disorder is the 3rd case report from India(1,2).

Case Report

A five-year-old boy, 3rd born to consanguineous parents, presented with history of generalized hyperpigmentation of the body for 9 months and progressive deterioration in speech, swallowing and gait for two months. The child had delayed milestones initially and later regression of attained milestones. The child had suffered three distinct episodes of sudden onset of loss of consciousness associated with shock and seizures.

Examination revealed marked hyperpigmentation of the palms, soles, nails, extremities, trunk, palate, gums, tongue and face (*Fig. 1*). The child had blank dull facies, excessive drooling of saliva and was not interested in surroundings. His speech was slurred. Cranial nerves were normal. He had increased tone in all the four limbs. Jaw jerk and deep tendon reflexes were brisk, abdominal reflexes were sluggish and superficial reflexes were normal while extensor plantar response was present. There were no cerebellar signs or signs of meningeal irritation.

A diagnosis of adrenoleukodystrophy was made because the child had clinical signs of corticotrophin deficiency and progressive neurological degeneration mainly involving white matter.

Investigations revealed: Hb 9.6 g/dl peripheral smear–normocytic hypochromic anemia. Serum calcium, serum electrolytes, serum transaminases, abdominal ultrasound and sural nerve biopsy were normal. CT Scan showed bilateral symmetrical hypodensity of parieto-occipital region, with normal ventricles and cisterns (*Fig. 2*).

Fig. 1. Clinical photograph of the patient showing spasticity, contractures at both ankles and hirsutism. (Hyperpigmentation has disappeared following corticosteroid therapy).

Serum C_{26} fatty acids were elevated (very long chain fatty acids).

On follow up, the child was continued on steroids. Though the generalized hyperpigmentation resolved remarkably, the child deteriorated neurologically and died at the age of six years.

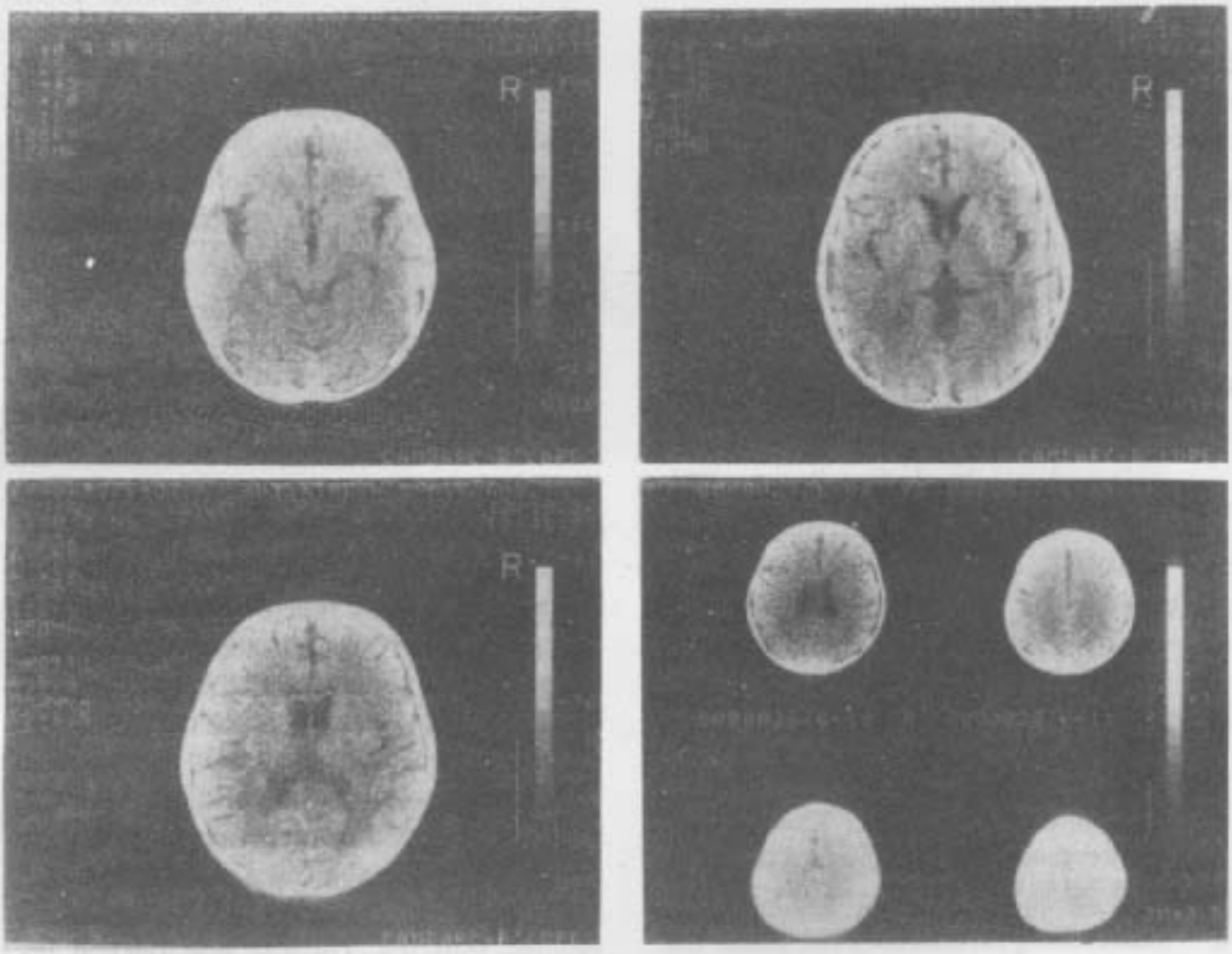

Fig. 2. CT Scan of head showing bilateral symmetrical hypodensity in posterior parietal and occipital areas.

Discussion

A clinical combination of progressive neurological symptoms and signs of adrenal insufficiency occurring in a boy easily arouses the clinical suspicion of adrenoleukodystrophy.

Features of adrenal insufficiency can precede or follow onset of neurological symptoms. In the present case, occurrence of three episodes of shock and seizures coupled with generalized hyperpigmenatation, representing adrenal insufficiency, preceded the neurological symptoms by eight months. In one third of cases, signs of overt adrenal insufficiency will be present at the time of diagnosis of adrenoleukodystrophy.

The CT Scan showing bilateral symmetrical hypodensities on the parieto-occipital region found in the present case is characteristic of this disease. The CT scan findings are due to active demyelination of the CNS and affords an initial clue in the diagnosis of adrenoleukodystrophy(3).

The presence of progressive deterioration of cerebral function associated with generalized hyperpigmentation occurring in a young male child with increased concentration of very long chain fatty acids, and CT Scan findings of bilateral symmetrical hypodensities, more in the posterior cerebrum corroborate with the diagnosis of adrenoleukodystrophy(4,5).

This rare disorder presently has no available treatment. Death usually occurs

between 1 to 1½ years of developing neurological manifestation(3) which also occurred in the present case.

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Spigelian Hernia

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Herniation occurring through a deficiency in the semilunar fascia is called spigelian hernia after the Flemish anatomist Adrian Vander Spiegel. Spigelian hernia is a rare clinical entity and most reports deal with adults(1), with only 18 cases reported in children(2). The rarity of this condition in children prompted the present communication.

Case Report

A 2-year-old boy was admitted on 3.8.1989 with history of a swelling in the left infraumbilical region from the third day of birth. There was no history of birth trauma or intestinal obstruction. Examination revealed a 3 × 2 cm soft, reducible infraumbilical swelling lateral to the left rectus muscle with a positive cry impulse. On complete reduction of the swelling, a 3 cm wide defect was palpable in the underlying fascia. With a diagnosis of spigelian hernia, exploration was done on 5.8.1989. A hernial sac was found emerging through a 3 cm defect in the linea semilunaris and lying between the external and internal oblique muscles. The sac was pushed inside, and the fascial defect along with the internal oblique muscle was repaired with 2/0 ethibond sutures. The external oblique was then double breasted with nonabsorbable sutures. The post-operative period was uneventful and the patient is asymptomatic at follow up 6 months later.

Discussion

Spigelian hernia is rare in children. It occurs through a weakness in the fascia

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