

The electromyographic findings though non specific are highly suggestive in an appropriate setting. Motor unit potentials are of normal size and duration and occur as grouped discharges, the repetitive potentials. During spasms a continuous activity is recorded extending into the period of relaxation. Prolonged activity is also evident on eliciting a deep tendon jerk, i.e., shortened silent period(2,3,6,7).

The manifestations of tetanus are now known to result from spinal and brainstem disinhibition of motor neurons. Tetanospasmin reaches the spinal cord via retrograde axonal transport from nerve terminals and blocks presynaptic neurotransmitter release around motor neurons(4,8,9). Generalized tetanus may result from blood borne dissemination of tetanospasmin to other muscles and subsequent retrograde axonal transportation(3,8).

The diagnosis of local tetanus is easy, provided the possibility is considered. Variable rigidity and involuntary spasms are sufficiently characteristic. Paucity of other neurological findings and when present abdominal rigidity and opisthotonus clinch the diagnosis(2,3,10).

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Congenital Skull and Scalp Defect

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Congenital defects of bone and scalp are rare abnormalities. Scalp defects are more common than bony defects. Recently we came across a neonate with bone defect

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in the parietal region and because of rarity, it is being reported.

Case Report

A 22-day-old infant was seen in Neurosurgical Outpatient with complaint of boggingness in the right side of head since birth. Infant was full term, weighed 3200 g at birth, delivered vaginally and was first born. The antenatal, natal and immediate postnatal events were uneventful. There was no such history in the family. The examination inclusive of detailed neurological assessment was noncontributory except the presence of an irregular scar tissue over the right parietal region with a palpable defect in the parietal bone underneath. X-ray skull revealed a well defined oval-circular defect in the parietal region (Fig. 1). After one year of follow up, there was no neurological deficit although the skull defect had slightly increased in size. The milestones were achieved normally.

The hemogram, urinalysis and chest X-rays were normal. Ultrasound revealed normal sized ventricles and no evidence of encephalocele. CT scan confirmed findings of ultrasound. Cranioplasty of the bone defect was carried out using rib. During the one year of postoperative follow up, the child has grown normally.

Discussion

The defects in the skull bones are rare and probably occur more frequently than one would gather from the literature. It is likely that small lesions are noted, but not reported.

The condition is slightly more common in females and first born infants(1-5). Skull defect is associated with scalp lesion in

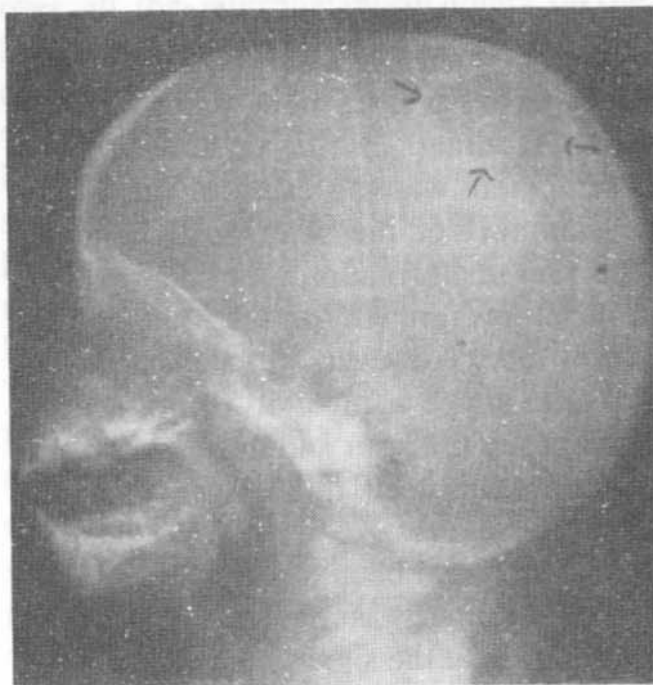


Fig. 1. Lateral view of plain X-ray skull showing defect in the parietal bone (arrow marks).

about 20%(2,6,7). Vertex, occipital and post-auricular areas are the commonest sites in that order for this type of defect. Defect may vary from 1-3 in number and from 0.5-6.5 cm in diameter.

Congenital abnormalities like hydrocephalus, meningocele, micro-ophthalmos, syndactyly, limb deformity, pulmonary atelectasis and polycystic kidneys may co-exist.

An inherited tendency for these defects to occur has been reported. Inheritance may be recessive or dominant. New bone formation from the margins over a period of time reduces the skull defect which may not require cranioplasty. In our patient, the defect did not decrease over one year observation period, hence cranioplasty was done. Cranioplasty has been recommended by some authors at the age of 2-3 years(1,5). Because of the scar in the scalp, the area has remained hair free.

These bony defects occur over the fontanelles, sagittal sutures or inferior angles of parietal bones; this can be explained on the basis of delayed bony union at these sites. At times in the underlying brain development was the cause of the lack of development of the skull bones and that the brain malformation may take the form of local overgrowth of the walls of the neural tube. However, we could not demonstrate any evidence of overgrowth of brain in the CT scan and defect was in the parietal bone itself rather than at angles where the sutures are present. We feel that there must have been a scalp defect which healed *in utero* as evidenced by thick scarred skin at the site of bony defect.

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Farber's Disease

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Farber's disease is a rare genetically determined lysosomal storage disorder of lipid metabolism. Classically it manifests in infancy and consists of a triad of subcutaneous nodules, arthritis and laryngeal involvement. There may be moderate nervous system dysfunction and the lungs, heart and lymph nodes may also be involved. Only 40 cases have been reported so far(1) and to the best of our knowledge, this is the first case report from India.

Case Report

A 3-year-old boy, born of a nonconsan-

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