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A Case of Clover-Leaf Skull (Kleeblattschaedel) Syndrome

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Abstract

A case manifesting the typical features of clover-leaf skull syndrome is described. Recognition of this rare condition by clinicians assumes importance since early neurosurgical intervention may lead to cosmetic and neurological improvement.

Key words -
Clover-leaf skull syndrome,
Kleeblattschaedeal syndrome

Holtermueller and Wiedemann [1] described a rare congenital malformation of the head characterised by a huge protrusion of the skull and marked broadening of the temporal region and face. They called it 'Kleeblattschaedal syndrome', which later came to be referred to as clover-leaf skull syndrome because of the characteristic shape of the head.

In this condition, there is a protrusion on top of the skull which is separated from the facial bulges by a furrow representing the rather normal dimensions of the face in the plane of the eyes. The result of these changes is a grotesque, clover-leaf shaped head, with external ears situated very low, in a horizontal position. Corresponding to the externally visible furrow, there is, within the skull, a bony shelf that separates a frontal vesicle of the brain from two temporal vesicles. The other important features include protruding eyeballs, antimongoloid slanting of eyes, premature synostosis of the lambdoid, coronal and sagittal sutures, and marked hydrocephaly. Hypoplasia of maxilla, nasal and zygomatic bones and orbits, high-arched palate, and micromelia with chondrodystrophic changes, may also occur. Most children are still-born or die soon after birth without mental development [2].

The report by Holtermueller and Wiedemann [1], which reviewed 12 cases published till then in German literature, considered the essential elements of the clinical entity to be
(i) trilocular configuration of the cranium,
(ii) associated facial malformations,
(iii) micromelia and anomalies of the skeleton (not obligatory), and,
(iv) typical radiologic configuration and hydrocephalus.

A new classification was proposed by Partington et al [3], in a paper wherein they reviewed 23 previously published cases and described 4 of their own cases. In their classification the cranial malformation was accepted either as a separate entity, or as one associated with other skeletal deformities. Zuleta et al [4]. reported the surgical correction of the cranial

malformation in 4 cases, and reviewed the earlier cases where it was attempted.

In this paper, a case of clover-leaf skull syndrome presented at the Mental Retardation Clinic at NIMHANS is reported.

Case report

L., a 1 year 4 months old female child, was admitted to NIMHANS on 13.12.83. She was born of consanguineous parents; her father was married to his sister's daughter. There was no significant family history. She was the last among 4 siblings, with 1 elder brother and 2 elder sister. The father's age when the child was born was 48 years, and that of the mother was 29 years. There was no significant pre - and peri-natal history. There was a mild delay of motor and mental milestones since birth.

Examination revealed the typical features of clover-leaf skull syndrome. Head was trilocular, with a huge vertical protrusion in the frontal region and marked broadening of the temporal regions (see Figures 1 and 2). The external ears were placed very low, almost in a horizontal position. There was telecanthus, lateral slanting and protrusion of the eyeballs. Hypoplasia of maxillary, nasal, and zygomatic bones were noted. Palate was high-arched. Limbs were short and thin, and the distance between 1st and 2nd toes was increased bilaterally.

- ***Front view of the face, showing trilocular skull***
- ***Profile showing temporal bulge and low set horizontal ear***

X-ray of the skull revealed an increase in the vertical diameter of the skull, much more so in the frontal region, with increased convolutional markings behind the coronal suture. Findings were also suggestive of craniostenosis of the coronal and lambdoid sutures, however, the frontal metopic suture appeared to be non-fused (Figure 3). X-ray of the hands and feet showed the bone age to be less than 1 year. The EEG and haematologic investigations were normal.

- ***X-ray skull A.P. view***

On 19.12.83, the child suddenly developed aspiration pneumonitis and died after a few hours.

Discussion

The clinical features of this case conform typically to the available description of clover-leaf skull syndrome. The absence of significant skeletal abnormalities is in keeping with the separate clinical entity proposed by Partington et al [3]. The fact that the child died at an early age of 1 year and 4 months brings to the fore the low survival of these children. However, survival to age 14 years has been described.

The recent report of successful surgical correction with these cases, resulting in acceptable cosmetic and neurologic improvement should alert the clinicians about this, albeit very rare, disorder.

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