KLEEBLATTSCHADEL SYNDROME: A GROTESQUE FORM OF HYDROCEPHALUS LEADING TO OBSTRUCTED LABOR

(A Case Report)

by

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SUMMARY

A full term newborn was delivered by caesarean section after all attempts to deliver the baby vaginally failed. The newborn was found to have gross abnormality of the skull — trilobed or clover leaf skull with proptosis, deformed nose and ears. Radiograph revealed frontal and temporal bossing of the skull with cranio lacunae. This is the third case of this rare syndrome to be reported from India.

Introduction

Kleeblattschadel syndrome is a rare form of hydrocephalus and is also known as cloverleaf skull or trilobed skull. In the literature available for review to us, only 2 cases of this rarity have been reported from India although many reports have appeared in foreign journals (Comings, 1965; Partington 1971; Young, 1973). The first mention of this syndrome as a distinct entity came in 1824 in German literature. In 1965, Comings gave a detailed report on the syndrome.

Sathya Bhama *et al* (1979) reported a case with detailed radiological findings from India.

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CASE REPORT

A primipara was brought to the maternity section of our hospital with obstructed labour. When vigorous attempts to deliver the baby vaginally failed, the woman was taken up for caesarean section with the diagnosis of cephalopelvic disproportion. The mother had never reported for antenatal care. The baby on extraction was seen to have grotesque cranial abnormality and was severely asphyxiated.

The skull had a trilobed appearance with frontal and temporal bossing and proptosis. The ears were low set and malformed. The bridge of nose appeared to be depressed.

The X-Rays of skull (both AP and Lateral projections (Fig. 1 and 2) were taken which revealed bulging of calvanium at the sites of frontal squamosa laterally at the sites of temporal squamosa giving it a trilobed appearance. The squamous portions of the cranial bones were absent being replaced by a peculiar honey combed pattern giving the appearance of lacunar skull. These radiolucent lacunar areas were separated by distinct strips of dense bone.

Orbital cavities, zygomatic arches were present as were maxillary, sphenoid and ethmoid bones. Teeth were present in maxillary and mandibular tissues. X-Rays of chest, abdomen and limbs revealed no abnormality.

The newborn could be revived with great difficulty but succumbed to respiratory failure after half an hour.

Discussion

In the detailed description of the syndrome by Comings (1965), the following features have been listed :

(1) Trilobed skull with depressed ears.
(2) Abnormal development of bone structure of the orbit, nose and jaw.
(3) Characteristic roentgenogram of the skull.
(4) Occasional presence of achondroplastic extremities.
(5) Usually fatal outcome.

The etiology of Kleeblattschadel syndrome has not been firmly established, but is believed to be a form of chondrodystrophy (Caffey, 1970). Support to this school of thought is lent by the concomittant occurrence of thanatophoric dwarfism in some cases (Partington, 1971 and Young, 1973). The possibility of its being severe form of craniosynostosis has also been proposed (Sathyabhama *et al* 1979).

References

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See Figs. on Art Paper II