Positional Moulding in Premature Hydrocephalics

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Summary

Seven premature hydrocephalics presenting with lambdoid positional moulding (LPM) were reviewed. All were treated for hydrocephalus secondary to aqueductal stenosis, Dandy Walker Syndrome and infection. Parenchymal hemorrhage, intraventricular bleed, cortical atrophy, septal agenesis, cortical anomalies and subdural hygroma were the other common associations. These children did not show expected improvement in their higher mental functions at 6 months to 5.4 years of follow-up, following the management of hydrocephalus. It was not the LPM but associated intracranial anomalies, which were most probably responsible for their poor outcome. The differentiation from posterior plagiocephaly is also highlighted.

Key words: Positional moulding, Craniosynostosis, Lambdoid positional moulding.

Introduction

Lambdoid positional moulding (LPM), expressed by asymmetrical head shape, is on the rise among infants and pediatric population. Posterior head deformations are rarely associated with lambdoid suture fusion and must be differentiated from true lambdoid synostosis (posterior plagiocephaly). Many cases of LPM have been labelled as posterior plagiocephaly in literature, hence, the differential diagnosis between the two is very important while deciding the approach and management. This condition is associated with many other congenital anomalies i.e. occult spinal defects, common birth related trauma, torticollis, hydrocephalus and brain tumors. The current literature describes the LPM mainly in infants and small children up to some extent, but its occurrence and associations in premature babies have not been described.

The present paper deals with seven premature hydrocephalics presenting with lambdoid positional moulding, the associations of LPM and outcome of these children, following treatment.

Material and Methods

Seven premature babies, who had undergone shunt CSF diversion (between years 1993 and 1999) for hydrocephalus, were evaluated for their asymmetrical head deformation and followed in OPD. Details of
### Table I
The Clinical Profile of Seven Children of LPM

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age at presentation</th>
<th>Status of Prematurity</th>
<th>Diagnosis</th>
<th>Sutures and side of LPM</th>
<th>Follow-up following shunt</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>3.5 years</td>
<td>7 months gestation, twins</td>
<td>Gross ventriculomegaly, agenesis of septum, right parieto-occipital periventricular porencephaly, thin cortical mantle, aqueductal stenosis</td>
<td>Lambdoid sutures patent, right LPM.</td>
<td>At 1.5 year: mentally retarded, unable to stand, monosyllable speech.</td>
</tr>
<tr>
<td>2.</td>
<td>2.5 years</td>
<td>7.5 months gestation, jaundice neonatorum, off and on fever, required incubator for one month</td>
<td>Dandy Walker Syndrome, cortical atrophy, Intraventricular bleed, subdural hygroma, / ? pachygyria, bifrontal sequestrated ventricles, porencephaly</td>
<td>Lambdoid sutures open, right LPM.</td>
<td>Six months following shunt: milestones delayed ventricular dilatation +, seizures, functioning shunt.</td>
</tr>
<tr>
<td>3.</td>
<td>1 year</td>
<td>8.2 months gestation, 1.6 kg delayed cry, delayed milestones</td>
<td>Hydrocephalus frontal porencephaly</td>
<td>Lambdoid sutures open, right LPM.</td>
<td>At 5.4 years mentally retarded restless, seizures.</td>
</tr>
<tr>
<td>4.</td>
<td>6 years</td>
<td>7.5 months gestation, delayed milestones</td>
<td>Hydrocephalus, aqueductal stenosis, subdural hygroma</td>
<td>Right LPM, opened sutures</td>
<td>At 2 years: seizures. At 2 years 3 months: subdural hygroma +, blocked shunt, seizures.</td>
</tr>
<tr>
<td>5.</td>
<td>1 month</td>
<td>6.5 month gestation, 1.2 kg, delayed cry, fever in 1st trimester</td>
<td>Hydrocephalus, aqueductal stenosis, biparietal calvarial defects</td>
<td>Sutures open right LPM</td>
<td>At further 2 month: shunt blocked, at 1.2 years delayed milestones</td>
</tr>
<tr>
<td>6.</td>
<td>10 months</td>
<td>8 months gestation, 2 kg, incubated for one month, seizures since birth, fever 2 months</td>
<td>Venticulomegaly, enhancing ependyma, no active infection</td>
<td>Left sides LPM sutures patent.</td>
<td>At 1.5 years: speaks common words achieved normal milestones walks briskly, seizures controlled</td>
</tr>
<tr>
<td>7.</td>
<td>8 days</td>
<td>8 months gestation, occipital encephalocele</td>
<td>Hydrocephalus, Dandy Walker variant, occipital encephalocele, small bleed in occipital parenchyma, Cortical atrophy</td>
<td>Left LPM, subdural hygroma</td>
<td>At 2 years: achieved normal milestones started walking, speaking 2-3 words.</td>
</tr>
</tbody>
</table>

LPM = Lambdoid positional moulding, Perivent = Periventricular, ‘+’ = Positive
individual cases were recorded from case sheets, discharge summaries and follow-up files. Parents of patients were interrogated during follow-up, regarding the achievement of developmental and motor milestones. A clinicoradiological work-up related with positional moulding was carried out in each case during the first admission in the hospital. Lambdoid sutures were either assessed on bone windows of CT scan or on plain skiagrams of head or by both, in each child. Follow-up of these children varied from 6 months to 5.4 years. Brief summary of each case is mentioned in Table I. Three cases are described in detail.

**Case Report**

**Case 1**: Three and a half year female child (born premature at 7 month, twin delivery) presented with history of gradual enlargement and deformation of head since birth. She had episodic vomiting for the last one month. On examination, the enlargement of head was asymmetrical posteriorly, flattened on right side, particularly at occipito-parietal region. Head circumference was 49 cm and biparietal diameter was 28 cm. Anterior fontanelle was wide open and tense. Plain CT head revealed aqueductal stenosis with gross ventriculomegaly, agenesis of septum pellucidum and right parieto-occipital porencephalic cyst communicating with right ventricle. Lambdoid sutures were either assessed on bone windows of CT scan or on plain skiagrams of head or by both, in each child. Follow-up of these children varied from 6 months to 5.4 years. Brief summary of each case is mentioned in Table I. Three cases are described in detail.

**Case 2**: Five month male child (born premature at 7.5 month with history of jaundice neonatorum; required incubator’s support for 1 month after birth) was admitted with 20 days history of progressive head enlargement, excessive cry, poor oral intake and downward deviation of eye balls noticed since birth. On examination, his head circumference was 46 cm, anterior fontanelle was tense and bulging and posterior fontanelle was open. Occipital flattening was marked on the right side. CT scan revealed cortical atrophy, grossly dilated lateral and anterior third ventricles and normal 4th ventricle. A large cisterna magna extending more on the left was noted. Occiput and posterior parietal region was flattened on right side. MRI revealed bilateral hemispherical thin subdural hygroma, bifrontal symmetrical porencephalic cysts and pachygyria (Figs. 2 and 3). In view of intermittent fever, a ventricular tap was done, which yielded 40 ml of homogeneously blood stained CSF. Biochemical examination of CSF revealed no suggestion of infection. The child was put on a week under high pressure. The child was discharged on 7th post operative day. At 1.5 year follow-up she was alert but mentally retarded, able to sit independently but could not stand unsupported. Her sagittal suture was prominent, head circumference was 60 cm and there was no improvement in lambdoid positional moulding.

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long trial of acetazolamide and Ommaya reservoir was placed for repeated tapping of CSF. Ventriculoperitoneal shunt was installed once Ommaya tap showed clear CSF on macroscopic and biochemical examination. At 6 month follow-up, the child developed neck holding, though other milestones remained delayed. Occipital flattening remained unchanged on follow-up. A repeat MRI after 8 months did not reveal a significant change in the ventricular size despite having digitally and clinically functioning shunt. This could have been due to pre-existing cortical atrophy.

**Case 3:** A 10 month old male (born premature at 8 months, having birth weight of 2.0 kg and required incubator for 1 month following delivery) was admitted with history of progressive head enlargement and head deformity since birth. He developed generalized seizures following delivery. History of intermittent fever was present for the last two months. He was receiving phenobarbital in a dose of 30 mg/day. Milestones were delayed historically. Anterior fontanelle was full, head circumference was 43 cm. The head was flattened posteriorly on left side, left ear was relatively placed anteriorly and the left frontal region was prominent (Fig. 4). No apparent cranial nerve deficit could be demonstrated. CECT scan head revealed gross ventriculomegaly with slight enhancement of ventricular ependyma on contrast injection, but ventricular CSF was not suggestive of infection. Ventriculoperitoneal shunt was performed; ventricular CSF was documented to be under high pressure. The child was alert, accepting orally and seizures free but fullness of fontanelles was noted on the 7th post operative day. CT scan showed a decrease in the size of the ventricles with bilateral subdural effusion. CT bone windows revealed open lambdoid sutures. At one and half year follow up, his head circumference was 43 cm with pronounced occipital flattening on left side. His milestones revealed gradual improvement: uttering sentences of 2 - 3 words, he was able to walk briskly.

**Discussion**

Lambdoid positional moulding (LPM) is a condition characterized by occipital flattening, alopecia, anterior displacement of ipsilateral ear and the petrous and frontal bones. LPM is rarely associated with lambdoid suture fusion (posterior plagiocephaly), hence the differentiation between the two is essential from the management point of view. Unlike in lambdoid synostosis, in LPM there is no lambdoid suture fusion. The overall incidence of LPM is unknown. Dunn reported an incidence of atleast 1 in 300 of plagiocephaly in neonates. Muakkassa studied 74 patients treated for apparent lambdoid synostosis, comprising 18.5% of their 404 craniosynostosis patients. However, 52 (13%) of these patients had occipital moulding deformities with sclerosis along the lambdoid suture margin and not true suture fusion. The etiology of LPM is less understood. Clarren suggested that when a rapidly growing fetal
head is maintained in an abnormal intrauterine position, calvarial moulding occurs.  

It is well known that positioning in neonatal period can affect head shape, an example is Scaphocephalic appearance of premature infants maintained with head in lateral, constrained position is one such example. Incidentally, the classical positional head moulding has been recognized amongst the flat headed Indians of North America. It seems that prematurity is one of the important predisposing factors in the development of LPM. It is apparent that if a supine position or another particular position is maintained constantly in prematures for long periods, it may result into LPM, as it had probably occurred in seven premature babies in our study. In one of these (the case of twin births), intrauterine constraints might have also contributed to the moulding. Right sided LPM is reportedly more common and this has been noted in five of our seven cases also.

Thirty seven percent cases of LPM were found to have associated thinner cranial bones, wider sutures and relatively greater cranial weight. Most probably all these factors remained contributory in these premature babies because all had hydrocephalus. Neurological developmental delay was observed in 19% cases and systemic abnormalities in 42% cases of LPM. Twenty eight percent cases, however, may have no association. A constellation of other neurological conditions was noted in 20% cases. These included hydrocephalus, intraventricular hemorrhage, CNS perinatal trauma or infection, Dandy Walker malformation, spina bifida and cerebellar hemorrhage. The other neurological manifestations included subdural hygromas, mild communicating hydrocephalus and atrophy. Hydrocephalus remained the presenting feature amongst all the premature babies in the present report. Aqueductal stenosis was responsible for hydrocephalus in three, while Dandy Walker syndrome in two, a possible infective origin in one. Etiology of hydrocephalus could not be ascertained in one child. Intraventricular and small parenchymal hemorrhage was seen in one case each, porencephalic cyst in three, subdural hygroma (with hydrocephalus) and cortical atrophy in three, agenesis of septum pellucidum in one, biparietal calvarial defect in one and meningoencephalocele in one patient were the other associations. Developmental delay remained a constant feature amongst six of these children. Case no 4, where there was no other cranial anomaly, developed an average intelligence. The developmental delay was most probably on account of associated cranial cortical/ventricular abnormalities. A high frequency of associated CNS anomalies emphasize the need for thorough neurological evaluation and neuroimaging studies in these children. Development of seizure was observed in five of our children during follow-up.

The vast majority of children labelled as posterior plagiocephaly did not have true synostosis, but had positional moulding. The differentiation between the two is very important in view of their divergent management. The routine use of CT, supplemented if necessary by three dimensional reconstruction for cases where the patency of lambdoid suture is in question on plain radiographs, facilitates a more realistic assessment of true lambdoid synostosis. In view of patent sutures in positional moulding, these children have relatively normal growth potential of cranium, unlike the posterior plagiocephaly. There is a strong argument to manage these children conservatively, at least initially, as the compensatory contralateral occipital or ipsilateral frontal growth provides enough room for the growing brain. The treatable associated cranial anomalies may require surgical intervention, as was carried out in our children, mainly for hydrocephalus.

In summary, premature children with hydrocephalus have relatively more frequency of LPM in comparison to posterior plagiocephaly. This apparently may be because of the maintenance of supine position with a thin calvarium and heavy weight of head due to hydrocephalus. The developmental outcome in these cases is poor, not because of LPM but because of associated anomalies.

References