The various biochemical parameters are given in Table I. Progressive increase in mortality was observed with a progressive reduction in serum albumin level (Table II). In those with serum total lipids and phospholipid levels below the normal range, the mortality was increased. On analysis with the Chi-square test for association, this observation was statistically significant at 1 and 5% level, respectively (Table III). No such association was noted with serum cholesterol levels.

Discussion

Even though serum albumin level is a good predictor of mortality, these days since frank Kwashiorkor cases with low serum albumin levels are becoming rare(1), it is not applicable in the majority of the cases of PEM. While considering serum lipids, an inverse relationship has been documented between the various serum lipid levels and fatty liver(8). The statistically significant increase in mortality noted in those with low serum total lipid and phospholipid levels is of clinical importance. Hence, we suggest that low serum total lipid and phospholipid levels may be regarded as probable predictors of mortality.

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Hydranencephaly/Multicystic Encephalomalacia: Association with Congenital Rubella Infection

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Hydranencephaly, defined as congenital absence of cerebral hemispheres, is a

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rare malformation of the brain(1). There is paucity of literature regarding association of hydranencephaly with congenital rubella infection. We are reporting two cases of hydranencephaly (one with a rare association with congenital rubella) for their rarity and because of the difficulty in differentiating them from multi-cystic encephalomalacia.

Case Reports

Case: A 11½-month-old girl, born of a non-consanguineous marriage was brought with severe developmental retardation. She was a full term normal hospital delivery. At the age of one month she was admitted somewhere else for fever and convulsions, and treated as a case of bacterial sepsis and falciparum malaria without the necessary laboratory evidence for the same. Other than fever and mild tachypnea at that time there were no other abnormal clinical findings. Her hemogram, urine, stool, blood sugar, serum calcium and CSF examination were normal. The cranial CT scan done at that time was of very poor quality and therefore noncontributory. Subsequently, at 11½ months the child was brought to us for further investigations for her severe developmental retardation. There was no other significant past or family history.

On examination the child was averagely nourished. The skull and spine were normal. Head circumference was within normal limits. There were no dysmorphic features. There were bilateral cataracts and the right eye was shrunken and soft. There was no perception to light, generalized spasticity was present without focal neurological signs. Developmental assessment revealed severe retardation. Other systems were normal. The clinical impression was either a congenital rubella infection or congenital cataract with pos-

tencephalitic brain damage.

The investigations were as follows: X-ray skull was normal; rubella IgM was negative but IgG was strongly positive; cranial CT scan (Fig. 1) showed absence of cerebral hemispheres and the entire supratentorial region occupied by CSF and multiple septae; and MRI (Fig. 2) showed absence of cerebral hemispheres, multiple septae in the supratentorial region, small brainstem, small right orbit with vitreous hemorrhage.

Case II: A 1½-year-old female child, born of a non-consanguineous marriage was brought with history of severe developmental retardation. There was history of difficult delivery and the baby cried late after birth. There were no other neonatal problems. There was no history of convulsions.

On examination, the child had a large head (head circumference 52 cm more

![Figure 1. CT scan of Case I showing hydranencephaly.](image-url)
than 95th percentile) with a positive transillumination. There were no dysmorphic features. Severe mental retardation and generalized spasticity were present. Vision was absent but fundus examination was normal. The other systems were normal. CT scan done to differentiate between severe hydrocephalus and congenital brain malformation showed hydranencephaly (Fig. 3).

Discussion

Hydranencephaly is defined as congenital absence of the cerebral hemispheres. The cerebral hemispheres are replaced by a fluid filled cavity. It is most probably due to an occlusive disease of supraclinoid part of internal carotid artery, occurring any time between 3 months gestation to the 2nd postnatal year (2). However, it can be caused by any severe insult to the cerebral hemispheres during the same period. Prenatally it may be due to various causes like congenital infection (mainly viral, toxoplasmosis) developmental malformation, trauma or raised intracranial pressure, giving rise to bilateral symmetrical lesions (hydranencephaly). Postnatally it may be due to severe meningitis (ventriculitis) or encephalitis specially Herpes simplex (3) producing asymmetrical lesions leading to multicystic encephalomalacia (3, 4). The basis of this type of damage relates to the remarkable property of immature brain to undergo rapid dissolution, with removal of degraded cellular components. The child survives as the less damaged brainstem is capable of maintaining vital functions.

Clinically hydranencephaly patients look apparently normal at birth and present with normal or increasing head size, delay in milestones, convulsions, absence of vision (optic nerve maldevelopment) and spasticity. The skull and scalp are
well formed thus distinguishing it from anencephaly or encephalocele. They have no facial or extracephalic malformations as in holoprosencephaly. Transillumination is positive and massive hydrocephalus is a close differential which can easily be diagnosed by CT scan or MRI.

Hydranencephaly and multicystic encephalomalacia may be the same end result of severe brain damage, differing only by the timing of actual insult. Early insult gives rise to hydranencephaly and later insult to multicystic encephalomalacia. Table I shows the differences between the two conditions and compares the above two cases with them. The diagnosis of the two cases is in favour of hydranencephaly for the following reasons: (i) Severe mental retardation; (ii) Absence of vision; (iii) Symmetrical nature of lesions on CT scan; (iv) Absence of ventricles; and (iv) Hypoplastic posterior fossa contents.

The diagnosis of congenital rubella in Case I is on the basis of clinical features like severe mental retardation and bilateral cataracts and a strongly positive titre of IgG antibody against rubella(16).

Hydranencephaly per se is usually an isolated finding with unknown etiology. Hydranencephaly in association with congenital rubella is very rare and the more common cerebral lesions in congenital rubella are microcephaly and subependymal cysts(7). Fetal viral encephalitis as a cause of hydranencephaly has been postulated in the past(3,4) and Case I further strengthens this view.

**TABLE I—Comparison Between Hydranencephaly, Multicystic Encephalomalacia and the 2 Cases**

<table>
<thead>
<tr>
<th>Parameters</th>
<th>HYDRN</th>
<th>Case I</th>
<th>Case II</th>
<th>ME</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mental retardation</td>
<td>++++</td>
<td>++++</td>
<td>+++</td>
<td>+</td>
</tr>
<tr>
<td>Vision</td>
<td>Abs</td>
<td>Abs</td>
<td>Abs</td>
<td>Pr</td>
</tr>
<tr>
<td>Seizures</td>
<td>Pr</td>
<td>Pr</td>
<td>Abs</td>
<td>Abs/Pr</td>
</tr>
<tr>
<td>Neonatal injury</td>
<td>Abs</td>
<td>Pr</td>
<td>Pr</td>
<td>Pr</td>
</tr>
<tr>
<td>Large head</td>
<td>Abs/Pr</td>
<td>Abs</td>
<td>Pr</td>
<td>Pr</td>
</tr>
<tr>
<td>CT scan</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hypodense lesions</td>
<td>Sym</td>
<td>Sym</td>
<td>Sym</td>
<td>Asym</td>
</tr>
<tr>
<td>Cerebral hemispheres</td>
<td>Abs/Pr</td>
<td>Abs</td>
<td>PPr</td>
<td>Pr</td>
</tr>
<tr>
<td>Post fossa contents</td>
<td>N/H</td>
<td>H</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Ventricles</td>
<td>Abs</td>
<td>Abs</td>
<td>Abs</td>
<td>E</td>
</tr>
<tr>
<td>Carotid angiography</td>
<td>Atretic supra clinoid int. carotid</td>
<td>ND</td>
<td>ND</td>
<td>Deviation of vessels</td>
</tr>
</tbody>
</table>

HYDRN = Hydranencephaly; PPr = Small amounts present; ME = Multicystic encephalomalacia; N = Normal; Abs = Absent; H = Hypoplastic; Pr = Present; E = Enlarged; Sym = Symmetrical; ND = Not done; Asym = Asymmetrical.
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REFERENCES


Ataxia Telangiectasia with Acute Lymphoblastic Leukemia

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Ataxia telangiectasia (AT) is an autosomal recessive syndrome characterized by ataxia, oculocutaneous telangiectasia, recurrent sinopulmonary infections and variable immune deficiencies. Individuals with AT are prone to develop lymphoreticular malignancies like leukemias and lymphomas(1). The management of patients of AT with acute lymphoblastic leukemia has not been uniform. We hereby report a case of AT who developed acute lymphoblastic leukemia (ALL) and was treated with reduced doses of chemotherapeutic drugs. He was in complete remission for 5 months but died following relapse of the disease.

Case Report

A 6-year-old boy, born of a nonconsanguinous marriage presented to the Tata Memorial Hospital in January with history of fever for a month and failure to thrive. On examination, we saw a dull appearing boy whose height (117 cm) and weight (16 kg) were between the 5th and 25th percentile for age. Prominent bulbar telan-