Cranial Neuroimaging in Infantile Tremor Syndrome (ITS)

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Clinical, hematological and cranial neuroimaging findings of eight cases of infantile tremor syndrome are reported. All had coarse tremors, anemia, hyperpigmentation and delayed or regression of developmental milestones. Five patients had microcytic, hypochromic anemia, three had dimorphic anemia. CT scans of two cases and MRI scans of three cases showed cerebral atrophy. One of these two CT scans, in addition, showed a small hypodensity in right basal ganglia region. Two CT scans were normal. One MRI showed hyperintense signals in frontal and periventricular white matter on T2 weighted images. The changes described are non-specific and also seen in cases of malnutrition and viral infections of CNS.

Key words: Infantile tremor syndrome (ITS), Neuroimaging

Infantile tremor syndrome (ITS) is a well-recognized clinical condition characterized by presence of tremors, anemia, dermal pigmentation and regression of developmental milestones, beginning during later part of infancy(1). Presence of tremors and neuromotor regression are the most prominent neurological manifestations basis of origin of which is poorly understood(1,2). Classically the presence of tremors has been attributed to structural and functional alterations of extra pyramidal system due to various causes. To find any such structural changes of brain, we studied cranial neuroimaging (CT scan/MRI) findings of eight such cases admitted in Department of Pediatrics, M.Y. Hospital and Chacha Nehru Bal Chikitsalaya; tertiary care hospitals at Indore, between March 2005 and October 2005, along with their clinical and hematological parameters.

Results

Mean age of the cases was 17 months. There were six boys and two girls (Table I). Four patients
had PEM grade II; three of them were of grade III; and one patient had 84% of expected weight for age. All of them had plump appearance with sparse and hypopigmented hair. Six children were exclusively breast fed. All had anemia, cutaneous hyperpigmentation and regression or delayed developmental milestones. Hemoglobin levels and peripheral blood smear examination findings are summarized in Table I. Two children had severe tremors involving limbs, head and tongue, while in others the tremors were more marked in distal parts of the limbs. Seven out of eight children had some infection at the time of presentation; five had bronchopneumonia and two had gastroenteritis. Pedal edema was present in two cases. Our clinical cases were similar to those described by others in their studies(3,4). CT/MRI scan findings are also summarized in Table I.

**TABLE I—Clinical and Neuroimaging Features of Eight Cases of Infantile Tremor Syndrome (ITS)**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (months)</th>
<th>Sex</th>
<th>Grade of malnutrition</th>
<th>Hemoglobin level and (g/dL) peripheral smear</th>
<th>Neuroimaging features CT / MRI scan</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>24</td>
<td>F</td>
<td>III</td>
<td>4.8 Microcytic, hypochromic</td>
<td>Diffuse cortical atrophy. Small hypodensity in right basal ganglia region.</td>
</tr>
<tr>
<td>2.</td>
<td>18</td>
<td>M</td>
<td>II</td>
<td>7.0 Dimorphic anemia.</td>
<td>Normal.</td>
</tr>
<tr>
<td>3.</td>
<td>24</td>
<td>F</td>
<td>III</td>
<td>5.0 Dimorphic anemia, few megaloblasts.</td>
<td>Normal.</td>
</tr>
<tr>
<td>4.</td>
<td>24</td>
<td>M</td>
<td>II</td>
<td>7.5 Dimorphic anemia.</td>
<td>Prominence of ventricular system and subarachnoid space. Diffuse white matter hypolasia.</td>
</tr>
<tr>
<td>5.</td>
<td>9</td>
<td>M</td>
<td>II</td>
<td>6.5 Microcytic, hypochromic</td>
<td>Prominence of ventricular system, subarachnoid spaces and cerebellar folia.</td>
</tr>
<tr>
<td>6.</td>
<td>12</td>
<td>M</td>
<td>II</td>
<td>7.0 Microcytic, hypochromic</td>
<td>Cerebral and cerebellar atrophy.</td>
</tr>
<tr>
<td>7.</td>
<td>13</td>
<td>M</td>
<td>–</td>
<td>6.7 Microcytic, hypochromic</td>
<td>Hyperintense signals in left frontal, periventricular white matter.</td>
</tr>
<tr>
<td>8.</td>
<td>12</td>
<td>M</td>
<td>III</td>
<td>6. Microcytic, hypochromic</td>
<td>Generalized atrophy of grey and white matter, more in frontal and temporal regions.</td>
</tr>
</tbody>
</table>

Discussion

Cortical atrophy and prominence of subarachnoid space and ventricular system were the commonest findings in MRIs (present in 3 scans). Hyperintense signals in frontal and periventricular white matter on T2 weighted images in one of the MRI scans may signify demyelination or edema and necrosis secondary to some ischemic insult, which in turn may occur due to some acute viral infection or parainfectious acute demyelinating encephalitis (ADEM)(5). On reviewing the literature relevant to our study we did not find much information, as far as neuroimaging in ITS is concerned. Presence of gyral atrophy and mild ventricular dilatation on CT scans of these cases has been described in literature. Pneumoencephalograms done in such patients also showed evidence of ventricular dilatation and cortical atrophy(2).

Thus, few non specific structural changes were seen on cranial neuroimaging with CT and MRI of 8 cases of ITS which may also be found in cases of malnutrition, especially in acute and recovering phase(6,7) acute viral meningoencephalitis or in ADEM(5). Studies using functional neuroimaging modalities may reveal areas of altered physiological function of brain and can help in determining exact etiology of hitherto poorly understood condition.
What this Study Adds

- Cranial neuroimaging using CT and MRI in cases of Infantile Tremor Syndrome reveals only non-specific structural changes.

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REFERENCES


