Multiple Spinal Stabilization As A Treatment For Hirayama Disease: Experience With Seven Cases

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Disclosure

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INTRODUCTION

- Relatively rare.
- **Clinical features**: Insidious onset asymmetrical weakness and disabling wasting of muscles of upper limbs which progress for few years and followed by arrest.
- **Radiological features**: Hallmark focal myelomalacia of the lower cervical cord with appearance of extra-dural mass in flexion.
- **Treatment**: Ranges from fixation and decompressive spinal procedures.

Here, we discuss the role of *multilevel spinal stabilization* as a treatment of Hirayama disease and role of cervical instability in its pathogenesis.
METHODS

• **Study Period**: June 2014 to January 2019
• **Total Cases**: 7
• **Basis of diagnosis**: Classical described clinical and radiological features.
• **Procedure**: Multilevel cervical fixation in all cases that included fixation of atlantoaxial joint in 6 patients by the adoption of facetal fixation methods. No dural or bone decompression was performed.
• **Follow-up**: ranged from 10 to 50 months (average 23 months).
RESULTS

• All patients had improvement in hand strength in the immediate post-operative period and progressive improvement in the symptoms of weakness, and deformity of hands in all patients.

• The other remarkable feature was postoperative reduction in extradural mass in all patients and its complete disappearance in 4 patients.

• No sagittal imbalance and no implant- or fixation-related complications.
Asymmetrical hand muscles wasting
Posterior extradural enhancing mass from C3 to C6
C1-C6 posterior fixation

Asymmetrical hand muscles wasting
Posterior extradural enhancing mass from C4 to C7
C1-C6 posterior fixation
<table>
<thead>
<tr>
<th>Case</th>
<th>Age, years/ Sex</th>
<th>Duration of Symptoms</th>
<th>Preoperative JOA Score</th>
<th>Radiologic Findings with Neck in Flexion</th>
<th>Postoperative JOA Score</th>
<th>Levels of Fixation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>21/M</td>
<td>1.5 years</td>
<td>15</td>
<td>Posterior extradural enhancing mass from C3 to C7</td>
<td>17</td>
<td>C3-C7 trans-articular fixation</td>
</tr>
<tr>
<td>2</td>
<td>17/M</td>
<td>2 years</td>
<td>14</td>
<td>Posterior extradural enhancing mass from C3 to C6</td>
<td>15</td>
<td>C1-C6 posterior fixation</td>
</tr>
<tr>
<td>3</td>
<td>20/M</td>
<td>2 years</td>
<td>15</td>
<td>Posterior extradural enhancing mass from C4 to C7</td>
<td>16</td>
<td>C1-C7 posterior fixation</td>
</tr>
<tr>
<td>4</td>
<td>28/M</td>
<td>2 years</td>
<td>13</td>
<td>Posterior extradural enhancing mass from C4 to C7</td>
<td>15</td>
<td>C1-C7 posterior fixation</td>
</tr>
<tr>
<td>5</td>
<td>16/M</td>
<td>1 year</td>
<td>13</td>
<td>Posterior extradural enhancing mass from C4 to C7</td>
<td>16</td>
<td>C1-C7 posterior fixation</td>
</tr>
<tr>
<td>6</td>
<td>18/M</td>
<td>1 year</td>
<td>12</td>
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<td>17</td>
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</tbody>
</table>
DISCUSSION

• First defined by Hirayama in 1959
• Pathophysiology: differential growth of bone and dural structures during the time of growth spurt leading to compression of the cervical cord by an extradural mass of unknown etiology that emerges on flexion of the neck and reduces on neck extension.
• Affects predominantly C7, C8, and T1 myotomes with male preponderance between 15 and 25 years of age.
• EMG study: chronic denervation, with or without acute denervation changes (fasciculations, positive sharp waves, and fibrillations potentials).
SUMMARY

From the observations this can be summarized as:

- Atlantoaxial and subaxial spinal instability plays a major role in the pathogenesis of Hirayama disease.
- Multi-level spinal stabilization done alone on the basis of understanding of the fact that instability forms the nodal point of pathogenesis of Hirayama disease, has not been discussed previously in the literature.
REFERENCES