Multilevel Spinal Stabilization as a Treatment for Hirayama Disease: Report of an Experience with Five Cases

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■ OBJECTIVE: **To analyze the role of multisegmental spinal instability in the pathogenesis of Hirayama disease.**

■ MATERIAL AND METHODS: **From June 2014 to January 2016, the authors managed 5 patients with Hirayama disease. The patients were diagnosed on the basis of classical described radiologic and clinical guidelines. All 5 patients were treated with multilevel cervical fixation that included fixation of the atlantoaxial joint in 4 patients by the adoption of the facetal fixation methods. No dural or bone decompression was performed. The follow-up ranged from 7 to 26 months (average 17.6 months).**

■ RESULTS: **The most remarkable feature was an immediate postoperative and progressive improvement in the symptoms of weakness, wasting, and deformity of hands in all patients. The other remarkable feature was an immediate postoperative reduction in extradural mass in all patients and its complete disappearance in 2 patients.**

■ CONCLUSIONS: **From the observations, it appears that atlantoaxial and subaxial spinal instability plays a major role in the pathogenesis of Hirayama disease.**

INTRODUCTION

Hirayama disease is a relatively rare but a known and defined clinical entity.[1,2] Disabling wasting, deformity, and weakness of hands in young men are hallmarks of the disease. Despite the fact that clinical and radiologic characteristics have been well defined, an exact understanding of pathophysiology and treatment strategy remains elusive, or at least it is highly debated. All 5 patients in our series were treated by multilevel spinal stabilization that included atlantoaxial joint in 4 patients without any form of bone or soft-tissue decompression. Our remarkably encouraging clinical results have prompted this presentation. Such treatment by multilevel spinal stabilization 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.

MATERIALS AND METHODS

Five patients having the classical clinical and radiologic features described in Hirayama disease were treated during the period from June 2014 to January 2016 (Figures 1 and 2). All 5 patients were relatively young men aged between 16 and 28 years of age (average age 20 years) (Table 1). The main complaints were weakness in both hands, which was more pronounced in the fingers. The symptoms were progressive in all cases. All patients had classical weakness of the intrinsic muscles of the hand with weak handgrip. Four of the 5 patients had flexion deformities of the lateral 2 fingers. The finger and hand deformity was almost similar in all 5 patients (Figure 2I). Three patients also had neck pain. All patients had difficulty in holding objects and writing.

The power of the hands was assessed by the upper limb component of the Japanese Orthopaedic Association score (Table 1). Investigations included dynamic plain radiographs, computed tomography, and magnetic resonance imaging. There was abnormal lower cervical spinal kyphotic curvature on flexion of the neck in all cases. A well-defined extradural mass emerged in the lower cervical spine predominantly posterior to the spinal cord on flexion of the neck and disappeared on neck extension. The subarachnoid spaces were compromised primarily on neck flexion in the lower cervical region. Subarachnoid spaces at other levels including the atlantoaxial region essentially were normal and had no evidence of bone or soft-tissue intrusion. There was no radiologic evidence of facetal instability at the subaxial cervical spine. There was type B atlantoaxial facetal instability (meaning thereby that in neutral head position the facet of atlas was dislocated posterior to facet of axis) in 3 patients and type C facetal

Key words
- Atlantoaxial instability
- Facetal fixation
- Hirayama disease

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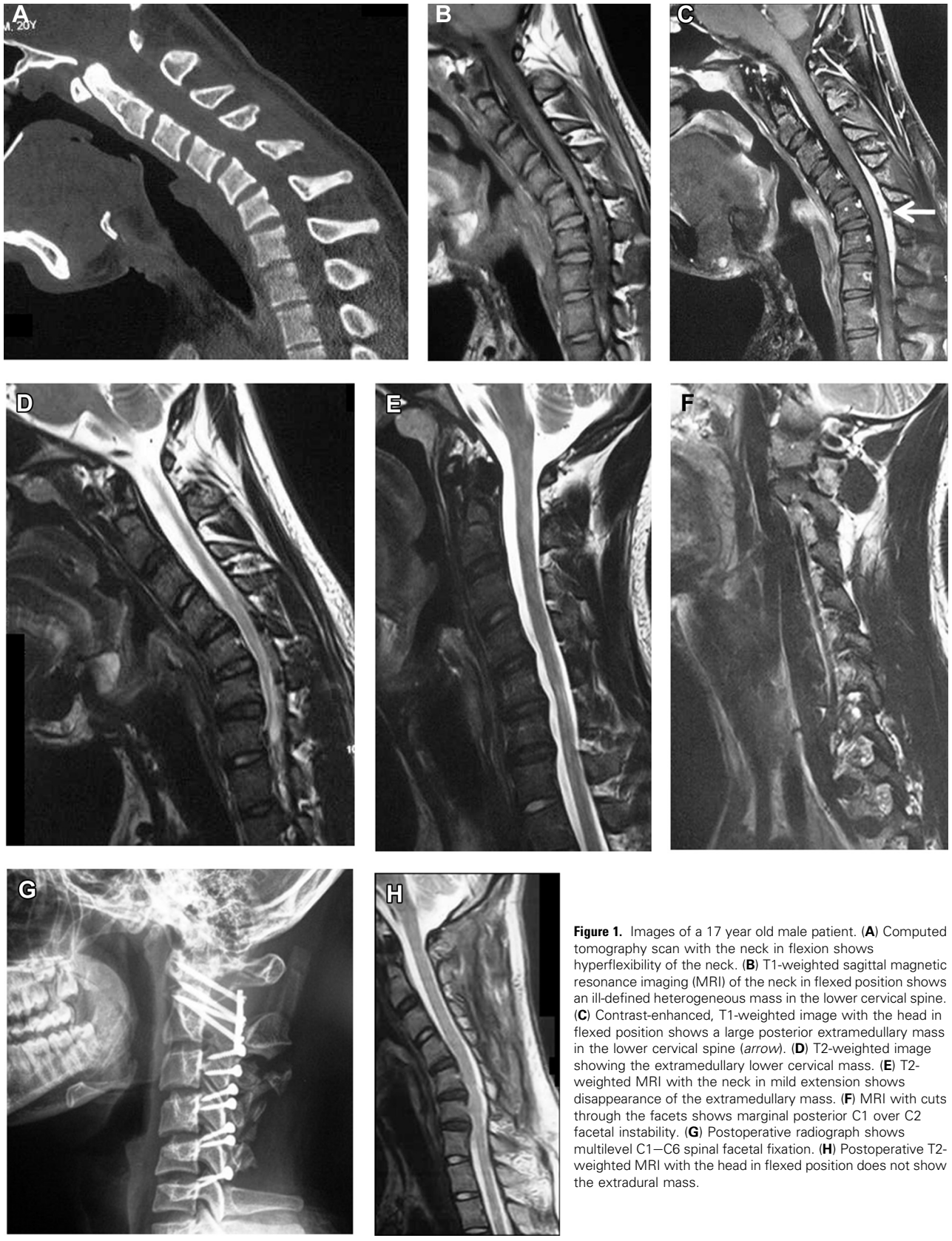

Figure 1. Images of a 17 year old male patient. (**A**) Computed tomography scan with the neck in flexion shows hyperflexibility of the neck. (**B**) T1-weighted sagittal magnetic resonance imaging (MRI) of the neck in flexed position shows an ill-defined heterogeneous mass in the lower cervical spine. (**C**) Contrast-enhanced, T1-weighted image with the head in flexed position shows a large posterior extramedullary mass in the lower cervical spine (*arrow*). (**D**) T2-weighted image showing the extramedullary lower cervical mass. (**E**) T2-weighted MRI with the neck in mild extension shows disappearance of the extramedullary mass. (**F**) MRI with cuts through the facets shows marginal posterior C1 over C2 facetal instability. (**G**) Postoperative radiograph shows multilevel C1—C6 spinal facetal fixation. (**H**) Postoperative T2-weighted MRI with the head in flexed position does not show the extradural mass.

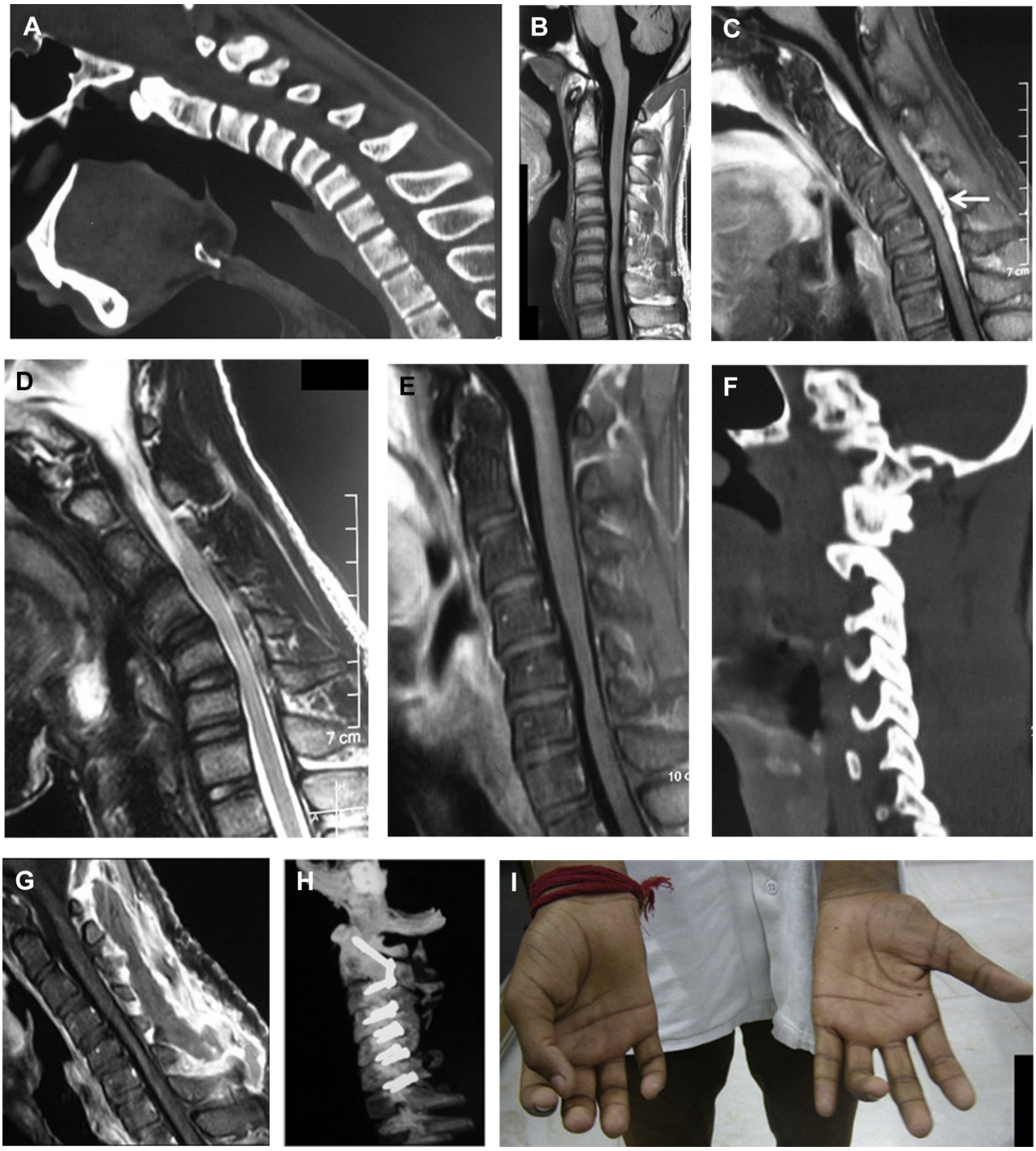

Figure 2. Images of a 16-year-old male patient. (**A**) Sagittal computed tomography (CT) scan shows hyperflexion of the neck. (**B**) T1-weighted magnetic resonance imaging (MRI) with the head in mild flexion shows ill-defined hypointense lesion posterior to the lower cervical cord. (**C**) Contrast-enhanced sagittal MRI with the head in flexion position. Extradural enhancing mass is seen posterior to the lower cervical spinal cord (*arrow*) and anterior to upper cervical spinal cord. (**D**) T2-weighted MRI image with the head in flexion shows the heterogeneous abnormal mass. (**E**) MRI with the head in neutral position showing disappearance of the abnormal extraspinal mass. (**F**) Sagittal cut of CT scan through the facets shows C1 over C2 posterior facetal instability. (**G**) Postoperative T1 contrast-weighted image with the neck in flexed position shows no evidence of extraspinal mass. Restricted neck flexion can be noted. (**H**) Postoperative images shows C1–C7 facetal fixation. (**I**) Image of the hands showing wasting and deformity.

Table 1. Clinical and Radiologic Features

Case	Age, years/ Sex	Duration of Symptoms	Preoperative JOA Score	Radiologic Findings with Neck in Flexion	Postoperative JOA Score	Levels of Fixation
1	21/M	1.5 years	15	Posterior extradural enhancing mass from C3 to C7	17	C3—C7 transarticular fixation
2	17/M	2 years	14	Posterior extradural enhancing mass from C3 to C6	15	C1—C6 posterior fixation
3	20/M	2 years	15	Posterior extradural enhancing mass from C4 to C7	16	C1—C7 posterior fixation
4	28/M	2 years	13	Posterior extradural enhancing mass from C4 to C7	15	C1—C7 posterior fixation
5	16/M	1 year	13	Posterior extradural enhancing mass from C4 to C7	16	C1—C7 posterior fixation

JOA, Japanese Orthopaedic Association.

instability (meaning thereby that the facets were in alignment and facetal instability was diagnosed only on direct bone handling during surgery) in 1 patient.[3] In 1 patient who was the first case managed in the series (case 1), atlantoaxial fixation was not done. The facets were in alignment in this case, and no atlantoaxial bone manipulation was done during operation to assess the nature of instability.

All 5 patients underwent posterior cervical facetal fixation by the transarticular technique. The level of fixation was based on the site of extradural mass, site and extent of kyphotic spinal curvature, and on direct observation of facetal instability during surgery. In 4 patients, atlantoaxial facetal fixation was done by the technique described by us previously (**Figures 1** and **2**). Intraoperative cervical traction assisted in stabilizing the head during surgery and maintained a neutral neck position that was targeted for multilevel fixation.

RESULTS

All 5 patients described improvement in hand strength in the immediate postoperative period (**Table 1**). Although the improvement of power, wasting, and deformity was partial, during the follow-up period that ranged from 7 to 26 months, all 5 patients have continued to progressively and satisfactorily recover. There were no implant- or fixation-related complications. Sagittal balance and kyphotic angle did not worsen despite the multiple levels of spinal fixation. All patients observed restriction of their neck movements, but this symptom was not a major postoperative complaint.

DISCUSSION

The clinical entity was first described in the year 1959 by Hirayama and has been named after him since.[1,2] The disease also sometimes is labeled as juvenile muscular atrophy of the distal upper extremity.[1,2] The disease is fairly rare, and only few isolated case reports and small clinical series are available in literature, more frequently from Asian countries.[1-15] The abnormality has sometimes been related to be a result of differential growth of bone and dural structures during the time of growth spurt.[11] Its absence in older patients and the self-limiting nature of the disease supports this hypothesis.[11]

Although there are variations from the classic characteristics, the primary feature is presentation in young men or older boys. The principal clinical manifestation is wasting and deformity of fingers and weakness of the hands. Leg weakness or stiffness generally is not the primary presentation. Sensory symptoms are remarkably few or absent. The symptoms are progressive and disabling, as they involve the fingers and wrist of both hands in particular. No familial correlation, genetic, or dietary factor has been found to be associated.

The radiologic features are hallmarked by focal myelomalacia of the lower cervical cord.[6] There is evidence of compression of the cervical cord by an extradural mass of unknown or uncertain etiology that emerges on flexion of the neck and disappears or reduces on neck extension.[7,8,12] A number of hypotheses regarding the possible mechanisms involved and the pathology of the dura and extradural tissue have been proposed on this debated subject.[12-14] The most obvious radiologic feature is the apparent cord compression on flexion of the neck by the anteriorly displaced dura at the lower cervical level and cord tissue thinning and altered signals at the level. The theory of cord compression by the abnormally tight dura on neck flexion is the most accepted mechanism.[12] Chronic and repeated trauma by the forward displacement of the posterior dura toward the vertebral body has been identified to be the cause of perfusion impairment and subsequent cord injury.[13] Cord ischemia caused by compression and related to microcirculatory disturbances in the territory of anterior spinal artery has been speculated to lead to the characteristic bilateral symptoms in the hands.[13] Compression by anteriorly located osteophytic bars also has been identified by some authors and implicated to be the cause of symptoms.[12]

Because the etiology is uncertain, the treatment protocol adopted in the reported cases ranges from a variety of fixation and decompressive spinal procedures. Cervical collar immobilization alone had been suggested in the past as a form of treatment, or even conservative monitoring.[15] Multilevel anterior corpectomies,

multilevel discectomies, decompressive laminectomies, dural grafting, "tenting" dural surgery, and a range of laminoplasties have been used in surgical treatment.[16-19] Both anterior and/or posterior fusions and simultaneous decompression or removal of midline bone elements essentially form the current basis of treatment. Although encouraging results have been demonstrated in almost all the articles describing the surgical treatment, it only may be correct to conclude that the philosophically correct form of treatment has not yet been identified.

Although abnormal range of neck flexion has been observed, no definite clinical or radiologic evidence of spinal instability has been demonstrated in any case reported in the literature. Because the use of cervical collar has been identified to be an effective means of treatment, instability might be considered in the pathogenesis of the disease process.[20] The presence of kyphotic cervical spinal deformity, abnormality of spinal alignment and curvatures on dynamic images, and presence of osteophytes do suggest the presence of instability. Identification of multilevel facetal instability was essentially an intraoperative direct observation that was based on our experience in dealing with atlantoaxial and subaxial facetal instability over several years.[21-27] The facetal instability was at multiple levels that included atlantoaxial instability in 4 patients. We observed clear evidence of open and abnormally mobile facet joints that suggested instability at multiple levels. The fact that all 5 patients improved quite remarkably in the immediate postoperative phase suggested the validity of the proposed rationale. The fact that no dural, neural, or bone manipulation or resection was done suggests that instability played a major role in the pathogenesis of the disease.

Although facetal instability in the subaxial cervical spine is difficult or impossible to identify on imaging because of its oblique profile, facetal instability at the atlantoaxial joint can be identified relatively easily because of its rectangular block-like structure and horizontal lie. The facet of atlas was displaced posterior to the facet of axis in three cases (**Figures 1** and **2**). According to our recently described classification, such facetal malalignment is described as type B atlantoaxial instability.[3] In one case, there was no radiologically identifiable atlantoaxial facetal instability but on direct bone handling, atlantoaxial instability was identified and subsequently was treated. We grouped such a form of facetal instability as type C atlantoaxial instability. Both type B and type C atlantoaxial facetal instability have been grouped as central or axial instability because there is no atlantodental interval disturbance and there is no evidence of neural compression by the odontoid process. Some degree of facetal malalignment is within the range of normal and can even be an effect of rotation of the head.

Diagnosis of atlantoaxial instability on the basis of our classification necessarily involves strong clinical corroboration. The validity of the classification of atlantoaxial instability on the basis of facetal malalignment and intraoperative observation will have to be assessed and confirmed by other authors. One patient did not undergo atlantoaxial fixation or intraoperative facetal evaluation. As this case was encountered first in our series, it may be possible that we missed the presence of type C atlantoaxial facetal instability. We identified that type B and type C facetal instability is associated with chronic spinal pathologies like Group B basilar invagination, Chiari 1 malformation, syringomyelia, ossified posterior longitudinal ligament, and multilevel spinal degenerative spondylotic disease.[23,24]

On the basis of our observations and outcome of treatment, it appears that chronic spinal alterations as observed in Hirayama disease could be secondary to multilevel spinal instability. Because atlantoaxial joint fixation was included in 4 cases, the exact role of atlantoaxial instability in the initiation or development of pathologic complex of Hirayama disease only can be speculated.

Our observations of the presence of multilevel spinal instability that includes atlantoaxial instability may point towards the pathogenesis of Hirayama disease. Atlantoaxial instability may be associated with instability of other spinal segments or may even be the primary nodal point of pathogenesis. It may be possible that the spinal encroachment of soft tissue can simulate syringomyelia that also is associated with instability at the atlantoaxial joint. Instability of the spinal segments does seem to be the possible cause because all 5 of our patients had sustained clinical improvement after surgery that involved only spinal fixation. We fixed a number of subaxial spinal levels on the basis of our direct intraoperative observations of instability of the facetal joints around the level of spinal pathology and in the region of kyphosis. It is clear that a longer follow-up is essential to confirm the usefulness of the proposed technique of treatment. It will have to be analyzed whether atlantoaxial instability is uniformly present in all cases or if there is varying and multiple levels of cervical spinal instability.

CONCLUSIONS

Multilevel spinal instability may be the defining cause of Hirayama's disease.

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