A 360-Degree Surgical Approach for Correction of Cervical Kyphosis and Atlantoaxial Dislocation in the Case of Larsen Syndrome

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Abstract

Larsen syndrome is chronic debilitating disease that presents with multiple joint dislocations and severely affects the cervical spine in the form of cervical kyphosis and atlantoaxial dislocation. Children usually present in early with a myriad of deficits, compressive myelopathy being the most common. In addition to a bony compression, there is sometimes a soft tissue component, which is seldom addressed. We present here a case of atlantoaxial dislocation with cervical kyphosis due to Larsen syndrome, and along with our previous experience on syndromic atlantoaxial dislocations, we try to define an algorithm for the treatment approach of these onerous challenges. The importance of early intervention is also emphasized with a literature review of similar cases. In addition to the obvious physical damage, early intervention can also avoid the more sinister socioeconomic face of this debilitating disease.

Keywords
► cervical kyphosis
► atlantoaxial dislocation
► Larsen syndrome

Introduction

In 1950, Larsen et al described the first series of cases with distinctive facial features, multiple joint dislocations, and spinal anomalies.[1-12] There is also a risk of dramatic cervical instability and sudden neurological deficit and death, as reported by Larsen himself. Since 1950, several authors have reported similar findings and yet a consensus regarding timing of correction, surveillance, pre- and postoperative bracing, and even the preferred surgical approach is missing. This is due to the wide spectrum of presentation and difference in severity at initial clinical evaluation.

Larsen syndrome occurs in 1 in 100,000 newborns, which is caused by mutations in the gene encoding filamin B (FLNB; 603381) on chromosome 3p14 that is important in regulating the structure and activity of the cytoskeleton.[13-15] We present a case of Larsen syndrome, which was managed, at our institute along with a possible protocol for the management of such cases in the future.

Case Report

History and Examination

"Dish face" they used to call him. A 15-year-old boy, studying in eighth grade, right-handed, presented with a prominent forehead and flattened nose (►**Fig. 1A**). He never used to play with the other boys, lest he risk getting injured, a lesson he had learned early in his life. They used to ridicule him for his long thin "spider"-like limbs (►**Figs. 1B** and **C**). Social stigma apart, he started noticing that the school bag was getting heavier. It became nearly impossible to walk to school with it. Gradually, feeding oneself became a task. The food would often slip through the fingers like grains of sand. Assistance was needed to drink water or even go to the toilet. Eventually, he was bed ridden for the better part of last month. However, he had retained function of his bladder and bowel, with inability to walk to the toilet. On inquiring from his teachers, they would describe him as a student with an average scholastic performance.

Fig. 1 Clinical photograph of Larsen syndrome with prominent forehead and flattened nose (**A**) and spider-like limbs (**B** and **C**).

On examination, his height was a 150 cm and weight 40 kg (BMI = 17.8 kg/m²). There was a definite kyphotic deformity of the spine without any local tenderness. He was able to lift all four limbs against gravity and had definite signs of myelopathy in the four of exaggerated deep tendon reflexes with upgoing plantars. Handgrip was worse on the right (30–40%) side than left (50%). Superficial reflexes were absent with graded sensory loss to all four modalities below C4 dermatome. Single breath count was 7 with a breath holding time of 20 seconds.

Past history was suggestive of multiple fracture dislocations of shoulders and knee often while playing in school after which the child had started to refrain from contact sports. There was no history of consanguinity in the family with birth history being unremarkable apart from the fact the child was home delivered. No history of recent trauma

or tuberculosis could be elicited. The history, characteristic facial features, and physical findings were suggestive of a compressive myelopathy due to congenital atlantoaxial dislocation.

X-ray of the cervical spine revealed a gross kyphotic deformity at C6 to C7 and atlantoaxial dislocation. Careful evaluation of computed tomography (CT) of the cervical spine showed kyphotic deformity (►**Fig. 2A**, **B**) due to anterior wedging of C5 vertebrae. The atlantoaxial joints showed degenerative changes. Magnetic resonance imaging demonstrated significant compression of the cord with signal changes at the craniovertebral junction and minimal retroflexion of the odontoid, although the soft tissue compression anterior to the cervical cord was significant.

Surgical Planning

There were two major considerations while doing the surgical planning: Soft tissue compressing the cervical cord at the cervicomedullary junction and the kyphosis at C5 to 7 levels. Both of the pathologies were significant and hence a decision was made for a transoral decompression of the soft tissue component followed by posterior fixation, including the two transition zones at craniovertebral junction and C7 to T1 levels. However, since more than three levels were involved a completely anterior approach with C5 corpectomy was abandoned as a single posterior fusion and decompression would deal with all the levels adequately. Since the compression was due to kyphosis and a soft tissue component, without any dislocation, preoperative traction was not considered.

Surgery

The patient was placed supine with neuromonitoring in the form of motor-evoked potentials and somatosensory potentials. He underwent a *transoral–transpharyngeal approach* that allowed lateral exposure of roughly 15 to 20 mm bilaterally off the midline from the inferior clivus to the C3 body. The anterior arch of C1 was drilled, laterally up to the lateral margins of the odontoid (~15 mm from midline). Safety margins lie within 11 mm from the midline at the foramen

Fig. 2 Plain computed tomographic scan of sagittal section of patient demonstrating improvement in preoperative kyphotic deformity (Cobb angle 27 degrees to postoperative 14 degrees).
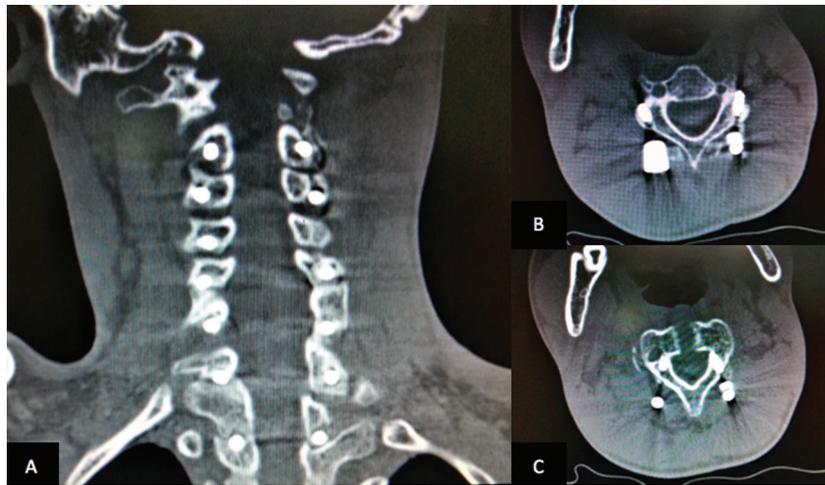
Fig. 3 Postoperative coronal (**A**) and axial (**B**, **C**) computed tomographic scan with posterior stabilization.

magnum, 24 mm at the atlas, and 14 mm at the lower border of the axis. Following this the *odontoid drilling* was done in a top down fashion. This avoided formation of a free-floating fragment of the dens as it was always attached at its base. A thick pannus was seen below the odontoid composed of elastic collagenous tissue and was gradually teased out using nibblers till pulsating dura was visible. After hemostasis, the closure proceeded sequentially using vicryl 2–0 and then monofilament 2–0 suture in an intermittent pattern. *Immediate posterior stabilization* was done by occipitocervical fixation including T1. Intraoperative manipulation was done to correct the kyphotic deformity (►**Figs. 2** and **3**) with the neuromonitoring parameters remaining unchanged throughout.

Postoperative Course

The patient was transferred to the postoperative intensive care unit and was extubated the next day followed by Ryles tube feeding started on day 2. He was transferred to the rehabilitation unit, from which he was later discharged neurologically intact with reduced spasticity on day 7 after surgery. Postoperative CT demonstrated complete correction of the deformity (►**Fig. 2**). Cervical collar was maintained for 3 weeks. Follow-up at 18 months showed excellent recovery of power to 4+/5 with independent ambulation and ability to take care of daily needs with return to school.

Discussion

Atlantoaxial dislocation is different in this case when compared with other syndromic and nonsyndromic varieties. We have found, in our case, that not only does the bony kyphosis cause compression and myelopathy, but also there was a soft tissue component to the compression.[16-18] During anterior decompression after removal of the odontoid, there was no dura visible and neither any pulsations were seen. On removal of soft cartilaginous tissue using rongeurs and nibblers, the dura was finally seen and compression relieved. Consequently, to achieve complete decompression of the cord, a 360-degree approach is necessary. Madera et al[14] were the first to perform a synchronous anterior decompression and fixation, posterior fusion, and fixation for a case of Larsen syndrome.[14] On extensive review, there have been 22 cases of surgically treated cervical deformity in patients with Larsen syndrome (►**Table 1**). The authors have significant previous experience in managing syndromic atlantoaxial dislocation and proposing an algorithmic approach (►**Fig. 4**) for the management of such cases.[2,3,8,9,19]

The treatment is dictated by the natural history of the disease. Although advocates of nonsurgical management have proposed continuous cervical traction and total spinal column bracing in patients with severe deficits since birth and have noted improvement in ventilator and motor functions, they too hypothesized that surgical intervention might be needed later.[20,21] Regarding the approach to subaxial cervical spine fusion, there is a simple approach that can be followed. In cases of minimal kyphosis with no myelopathy, a short segment fusion posteriorly may suffice. However, if severe kyphosis or myelopathy is present, anterior decompression followed by a 360-degree fixation should be aimed for.[22,23]

One thing that is noncontroversial in this syndrome is the need for intervention. With the review of literature and our own personal experience, it is clear that the stage of intervention matters.[5] Patients who were operated earlier or at a stage where they had minimal to no deficits fared much better than those allowed to deteriorate. Further the impact of multiple falls or chronic cord compression cannot be overstated. In severe cases with apnea and respiratory distress, an even earlier decompression with Halo stabilization followed by the earliest allowable opportunity for fixation may be a safe alternative.

Conclusion

Larsen syndrome cases have several defects, the most severe of them being cervical kyphosis and atlantoaxial dislocation. A 360-degree decompression and fixation of the

Table 1 Review of all cases of surgically corrected cervical kyphosis in patients of Larsen syndrome

Author	Age at first surgery	Trauma	Traction/ Collar	Preoperative condition	Surgery	Collar/ brace	Follow-up
Micheli et al 1976[15]	10mo	None	Semirigid cervical	UE and mild LE weakness	C3-T2 posterior fusion	Minerva jacket	Unknown
Muzumdar et al 1977[21]	13.5 y	Fall	NA	Bilateral numbness, weakness in all four	Cervical decompression	NA	Minimal improvement, later deteriorated
Bowen et al 1985[5]	19 y	None	NA	No deficit, progression of kyphosis	Occiput–C4 posterior fusion, later scoliosis correction	Minerva jacket	No deficit
Miz and Engler 1987[20]	14 mo	Motor vehicle accident	NA	Hyperreflexia, decreased perineal sensation	Occiput–C2 posterior fusion	Minerva jacket	No recurrence
Francis and Noble 1988[7]	5 y	None	NA	Weakness, inability to right self	Anterior cervical decompression, fibular strut placement	Halo vest	No deficit
Forsee et al 1995[6]	5 y	None	NA	Arm weakness	Anterior, later posterior fusion	NA	No improvement
Johnston et al 1996[10]	10 mo	Fall after first operation	NA	No deficit initially, fall after first operation induced quadriparesis	Posterior cervical fusion f/b 2 anterior decompression and fusion	Minerva jacket and Halo vest	Improvement in walking
	14 mo	None	NA	No deficit	Posterior fusion	Halo vest	No deficit
	14 mo	None	NA	No deficit	Posterior fusion	Minerva jacket	No deficit
	16 mo	None	NA	No deficit	Posterior fusion	Minerva jacket	No deficit
	12 y	None	NA	Myelopathy, weakness	Anterior and posterior fusion	NA	No improvement
Luk and Yip 2002[13]	8 y	None	NA	No deficit at first surgery, sensory in UE later	Anterior T12-L3 fusion, anterior decompression and fusion, later posterior fusion	Halo vest	Transient weakness, later no deficit
	6 y	None	Halo	Myelopathy	Posterior C1-T1 fusion, anterior cervical fusion and repeated anterior fusion	Halo vest	Myelopathy resolved after first anterior cervical fusion
Banks et al 2003[1]	13 y	Fall	Halo	Myelopathy, weakness in all extremities	Posterior C1-T1 fusion, anterior cervical decompression and fusion 4 d later	Halo vest and then hard cervical collar	Transient increased weakness postop w/ later improvement in better than preop status
Katz et al 2005[11]	3 y	Falls	NA	Weakness before first operation, inability to walk before third operation	Two failed posterior cervical fusions, posterior decompression and fusion, later anterior fusion w/ post sublaminar wires	Halo vest and then collar after second operation, Halo vest through fourth operation, later collar	After fourth operation, transient weakness w/ improvement but persistent C5 and C6 weakness

(continued)

Table 1 (continued)

Author	Age at first surgery	Trauma	Traction/ Collar	Preoperative condition	Surgery	Collar/ brace	Follow-up
Sakaura et al 2007[18]	34 mo	None	Minerva Brace	Spastic quadriparesis with sleep apnea	Anterior decompression corpectomies C4–C5 arthrodesis C3–C6 using tibial strut bone grafts via a lateral approach and later revision of anterior fixation with C2–C7 fixation	NA	Quadriparesis and respiratory dysfunction improved
	58 mo	None	None	Hyperreflexia	Posterior fusion C3–C5	Halo vest	No deficit
	10 mo	None	Halo traction	No deficit, kyphosis worsening	Postcervical arthrodesis. At 29 mo: Anterior decompression C4–5 corpectomy, C3–C6 arthrodesis and later occiput-T4 arthrodesis	Halo vest	No deficit
Madera et al 2008[14]	2.5y	None	Hard collar	No deficit	Synchronous ant decompression and fusion/fixation, posterior fusion/fixation	Halo vest	Transient postoperative weakness and Horner syndrome resolved.
Kumar et al 2013[16]	36 y	None	NA	Mild spasticity of all four limbs	Anterior C2–C5 decompression and fixation and later posterior C1–C6 fusion	Philadelphia collar	No deficit
Yonekura et al 2015[22]	18 y	None	NA	Airway obstruction and repeated aspiration pneumonia	3 y age: post cervical arthrodesis. 18 y: Anterior mediastinal tracheostomy	NA	No deficit
Sahoo et al 2016[17]	56 y	None	NA	Neck pain with spastic quadriparesis	Posterior C1-C2 fusion	NA	Improvement in spasticity
Present case	15 y	None	Hard cervical collar	Neck pain with spastic quadriparesis	Transoral decompression f/b occipito-T1 fusion	Hard cervical collar	Improvement in power and spasticity
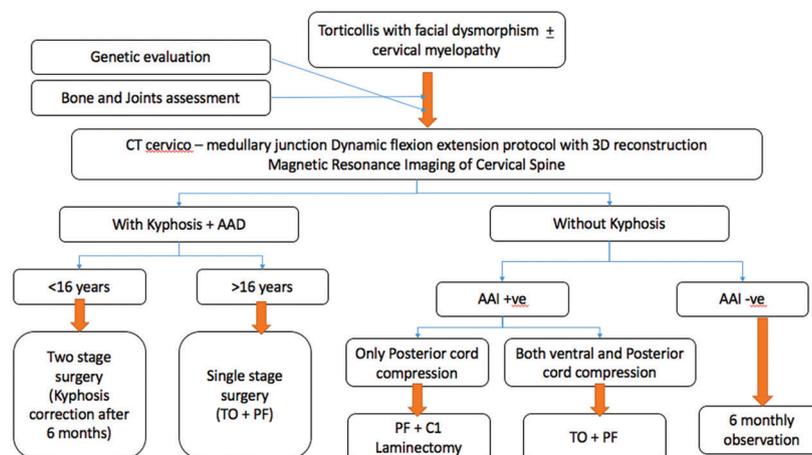
Fig. 4 Algorithmic approach to a case of Larsen syndrome. AAD, atlantoaxial dislocation; CT, computed tomography; PF, posterior fixation

atlantoaxial dislocation followed by deformity correction can prevent further deterioration due to chronic cord compression or trivial falls. We attempt to delineate here that anterior decompression at every level may not be necessary and a single posterior fusion is sufficient. Decompression anteriorly is only needed in case of significant soft tissue compression

that otherwise cannot be addressed posteriorly. Neurological condition at presentation dictates outcome.

Funding
None.

Conflict of Interest
None declared.

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