Combined ossification of the posterior longitudinal ligament at C2–3 and invagination of the posterior axis resulting in Myelopathy

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Received: 12 March 2012 / Revised: 7 November 2012 / Accepted: 28 December 2012 / Published online: 20 January 2013
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Abstract

Purpose Spinal stenosis at the C2–3 segment is a rare occurrence, and when it occurs myelopathy infrequently results. Furthermore, only a handful of cases involving congenital abnormalities of the posterior arch of the axis have been described resulting in cervical myelopathy many of which described simultaneous congenital abnormalities at adjacent levels and none of which identified ossification of the posterior longitudinal ligament (OPLL) at the same level. We report a case of a previously undescribed combination of abnormalities at the C2–3 segment resulting in clinical myelopathy.

Methods A 49-year-old Chinese male presented with a progressive cervical myelopathy (C-JOA score 11 immediately pre-op). Segmental OPLL at the C2–3 disk space was visible, together with invagination of the bilaterally hypoplastic C2 lamina into the spinal canal. Signal abnormalities of the spinal cord were evident on both T1 and T2 sequences.

Results The patient underwent a posterior decompression and instrumented fusion at C2–3 using pars screws at C2 and lateral mass screws at C3. Following surgery there was a rapid and significant improvement in the neurological symptoms, with the C-JOA score improving to 14 at final follow-up. A successful fusion was evident.

Conclusions Deficiencies in the posterior arch of the axis are rare and have not previously been reported in conjunction

with OPLL. Advanced imaging is helpful to define the abnormality and site of compression. In the setting of a progressive neurological dysfunction, surgical decompression and stabilization is a reasonable intervention and can be associated with neurological and symptomatic improvement.

Keywords Ossification of the posterior longitudinal ligament · Cervical myelopathy · Axis · Congenital abnormality · Decompression

Introduction

Spinal stenosis at the C2–3 level is uncommon, given the relatively wider spinal canal diameter than more inferior cervical levels [1], and when it is present it usually does not result in myelopathy. Congenital abnormalities of the posterior arch of the axis are one of the rarest causes of cervical myelopathy, with only a small number of cases reported in the literature [2]. In contrast, ossification of the posterior longitudinal ligament (OPLL) is a well-established cause of cervical myelopathy, but the isolated occurrence of OPLL at the C2–3 segment is unusual. We report a case of combined anterior and posterior spinal cord compression at the C2–3 level, with both OPLL and invagination of the hypoplastic C2 lamina contributing to the development of a progressive cervical myelopathy and review the existing literature regarding isolated congenital abnormalities of the posterior axis.

Case report

A 49-year-old male presented with progressive weakness in the lower extremities for approximately 3 years. Subjective

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loss of balance was also present together with a change in his gait. In his upper extremities he was experiencing numbness and weakness in his hands, particularly on the right, for 2 months. There was no bowel and bladder dysfunction present.

His physical examination was significant for positive Hoffman's reflexes bilaterally, hypertonicity, hyperreflexia with a positive Babinski reflex and sustained ankle clonus present. Motor weakness was seen in the intrinsic (right greater than left) and generally in the right upper and lower extremities. His pre-operative C-JOA score was 11 points.

Magnetic resonance imaging showed severe spinal cord compression circumferentially at the C2–3 level. Anteriorly there was a central protrusion of the C2–3 disk. CT scan was also performed to further define the osseous anatomy. Segmental OPLL at the C2–3 disk space was visible. Posteriorly invagination of the hypoplastic C2 lamina was evident bilaterally into the spinal canal. Hypotrophic and shortened pedicles were also present at C2. Signal hyperintensity of the spinal cord was evident on the T2 sequence; extending from the superior third of the odontoid process to the mid-vertebral body of C3. A corresponding hypointensity on MRI was also visible on the T1 sequence. No other abnormalities were seen in the thoracic or lumbar spine (Figs. 1, 2, 3, 4).

The operative time was 2 h and 30 min. The C2 lamina was noted to have fibrous soft tissue attachments in communication with the C2 lateral masses bilaterally. The lamina was visibly unstable and compressed the cervical cord. A complete laminectomy was performed at C2 and the entire floating lamina was removed. We used Leksell and Kerrison rongeurs to remove the lamina. When the C2 laminectomy was performed there was a venous plexus surrounding the C2 isthmus, and some blood loss occurred prior to obtaining hemostasis. Considerable mechanical

instability was present at C2–3 with gross motion evident. A fusion was performed utilizing pars screws at C2 and lateral mass screws at C3 connected with rods. A limited fusion at the C2–3 articulation was performed to preserve the cervical motion segments. Given pre-operative imaging as well as operative findings, there was evidence of retro-vertebral spinal stenosis behind the C3 body that extended to the mid-vertebral body level of C3 and to a lesser extent to the C3–4 disk space. Given this we felt that the best way to adequately address this stenosis was to decompress the entire lamina of C3 and the superior lamina of C4 (Figs. 5a, b, 6, and 7a, b). The total blood loss was 300 ml. The patient was instructed to wear a hard collar for 3 months and had an unremarkable postoperative recovery without complications. Post-operatively, neurological recovery occurred rapidly with the C-JOA score improved to 14 points at initial follow-up and maintained at the time of final follow-up.

Discussion

Unlike defects of the posterior atlas, abnormalities in the posterior arch of the axis are rare and typically do not occur in isolation or result in cervical myelopathy. Isolated OPLL at the level of the C2–3 disk is also quite uncommon and suggests a possible correlation. The occurrence of these two defects simultaneously at the C2–3 segment has not been reported previously.

To our knowledge this case represents the 16th case reported in which abnormalities of the C2 lamina resulted in cervical myelopathy [1, 3–11] (Table 1). Several types of defects in the posterior elements of the axis have been described in such cases. Congenital hypertrophy of the posterior arch of C2 has the potential to cause myelopathy

Fig. 1 Pre-operative lateral radiographs in flexion and extension, respectively

Fig. 2 Pre-operative T-2 sagittal MRI right para-sagittal and mid-sagittal, respectively

Fig. 3 Pre-operative sagittal CT scan right para-sagittal and mid-sagittal, respectively

Fig. 4 Axial CT scan at the level of the C2–3 revealing invagination of the C2 Lamina

[6, 10]. Hypoplasia of the posterior elements of the axis can also paradoxically have the same clinical effect, presumably secondary to instability. Direct posterior compression

is also one potential cause of compression at the level of the axis. In the presented case, a portion of the stenosis resulted from invagination of the C2 lamina. Defects

Fig. 5 Post-operative lateral and AP radiographs at final follow-up

Fig. 6 Post-operative sagittal MRI

involving the axis have been divided by Van Gilder et al. [12] into the following categories: irregular atlantoaxial segmentation, dens dysplasias: os odontoideum, hypoplasia and aplasia, and segmentation failure of C2/3. Defects involving the posterior portion of the axis are extremely rare relative to those that occur anteriorly. The axis originates from four primary ossification centers in the pro-atlas, first and second cervical sclerotomes [12], with posterior arch chondrification beginning at 6 weeks embryogenesis and ossification completing by 3 to 4 years of age [3]. The paired posterior ossification centers typically fuse posteriorly by 2–3 years. Failure of them to fuse results in a bifid arch. Ossification centers fuse to the body at about 7 years of age. Failure of fusion is considered spondylolysis, which had been reported recently imitating Hangman fractures in several patients [13–16]. The occurrence of these two deformities in conjunction with each other is thought to be the cause of a floating lamina. This has been described in five previous cases with invagination of separated and dysplastic C2 lamina and spinous processes [1, 3, 8, 9, 17]. Schematics of the development of the axis are provided in Fig. 8a–c. In our case the two laminae were fused but were not attached to

Fig. 7 Post-operative axial MRI demonstrating adequate decompression

Table 1 Documented cases of upper cervical stenosis and congenital abnormalities of the posterior axis resulting in clinical myelopathy

General category	Authors	Cases	Age/ gender	Clinical symptoms	C2 abnormality	Associated abnormality	Location of spinal stenosis	Surgical treatment	Clinical result
Hypoplasia of posterior C2 arch with Invagination	Koyama et al. [9]	1	54 M	Pain and tingling in lower back, left thigh and left shoulder girdle	Spina Bifida Occulta with dysplastic bilateral laminae	C2–3 abnormal articulations	C2	Decompression with removal of dysplastic structures	Improved
	Kawano et al. [8]	1	42 M	Motor and sensory disturbance in all extremities with gait disturbance	Separation of laminae from base of spinous process with invaginated spinous process	None	C2	Decompression with C2 posterior arch removal	Improved
	Asakawa et al. [3]	1	46 M	Numbness of hands and stiffness in all extremities with gait disturbance	Spina bifida occulta with invaginated bilateral bifid lamina	Enlarged C3 spinous process	C2–3	Decompression with removal of bifid C2 laminae and C3 laminectomy	Improved
	Sakai et al. [1]	1	53 M	Bilateral lower extremity stiffness and left arm paresthesias with gait disturbance	Bilateral invaginated hypoplastic C2 laminae with disruption in posterior arch	C2 spinous process fused to superior articulating C3 process	C2–3	Decompression with removal of invaginated laminae and C3 laminoplasty	Improved
	Jiang et al. [17]	1	16 M	Paroxysmal dizziness, chest pain and bowel/bladder incontinence	Invaginated hypoplastic right C2 laminae with disruption in posterior arch and absent left lamina	C2 spinous process fused to C3 spinous process	C2–3	Occipitocervical fusion with removal of right C2 and C3 hemilaminae	Improved
Aplasia of C2 lamina	Chau et al. [2]	1	14 M	Weakness of right upper and lower extremities with gait disturbance	Bilateral invaginated laminae and floating spinous process	Aplasia of posterior arch of atlas	C2–3	Decompression with C2 and C3 laminectomy with C2–3 lateral mass fusion	Improved
	Current Case	1	49 M	Weakness in all extremities with numbness in the hands and gait disturbance	Invaginated hypoplastic C2 laminae with hypotrophic shortened pedicles	C2–3 OPLL	C2–3	C2 laminectomy with C2–3 fusion	Improved
	Goel et al. [4]	2	42 M	Neck pain and weakness of all extremities	Complete aplasia of posterior arch of C2	C2–3 anterolisthesis	C2–3	1. Posterior C2–3 fusion with plate and wiring 2. Transoral decompression of C2–3	Worsened → Improved

Table 1 continued

General category	Authors	Cases	Age/ gender	Clinical symptoms	C2 abnormality	Associated abnormality	Location of spinal stenosis	Surgical treatment	Clinical result
			16 M	Neck pain with numbness and weakness in all extremities	Complete aplasia of posterior arch of C2	C2–3 anterolisthesis	C2–3	Anterior decompression of C2 with iliac crest structural graft and posterior Occiput–C5 instrumented fusion	Improved
	Muzumdar and Goel [26]	1	31 M	Weakness and stiffness in all extremities	Complete aplasia of posterior arch of C2	C2–3 anterolisthesis	C3 body	Transsoral C2 and C3 decompression with C2–C4 plating	Improved
	Behari et al. [5]	2	12 M	Neck pain and stiffness with quadriparesis	Complete aplasia of posterior arch of C2	Atlantoaxial dislocation; Occipitalization of atlas	Foramen magnum	Transoral odontoid decompression with posterior occipitocervical fusion	Improved
			16 M	L'Hermite's with quadraparesis and change in bowel and bladder function; labored respirations	Absence of C2 laminae bilateral; free	Atlantoaxial dislocation	C2–3	C2 spinous process removed; Occipitocervical fusion to C4	Improved
Hypertrophy of the posterior arch of C2	Prakash et al. [10]	1	40 M	Paresthesias and stiffness in all extremities with urinary urgency	Bilateral laminae and spinous process hypertrophy	Lesser C1 bilateral laminae hypertrophy	C2	Decompression with C1 & C2 posterior arch removal	Improved
	Benitah et al. [6]	1	41 M	Paresthesias in all extremities; leg weakness and L'Hermite's	Bilateral laminae hypertrophy	Lesser C1 bilateral laminae hypertrophy	C2 > C1	Decompression with C1 & C2 posterior arch removal	Improved
Associated with C1–2 stenosis	Goto et al. [7]	1	64 M	Right upper & lower extremity weakness with gait disturbance	Osteophytes projecting into spinal canal	DISH with pseudoarthrosis at C1–C2	C1–C2	Decompression with C1 & C2 posterior arch removal	Improved
	Suzuki et al. [11]	1	61 M	Upper extremity numbness with gait disturbance	C2–3 assimilation	Klippel-Feil; C5–C6 stenosis	C1–C2	C1 & C2 laminotomies with C3–C7 laminoplasty	Improved

Fig. 8 **a** Transverse schematic of the axis approximately 3 months following birth reveals normal development with the two neurocentral synchondroses anteriorly with squaring off of the dens, and ossification extending posteriorly towards the spinous processes. **b** Transverse schematic at approximately 5 years reveals complete ossification of the posterior arch (typically complete by 3–4 years) with incomplete fusion to each other (typically complete by 5 years) resulting in a bifid arch. The anterior neurocentral synchondroses with the body are still visible (typically complete by 7 years). Failure of the neurocentral synchondroses to close (spondylolysis) together with failure of the posterior ossification centers to fuse results in floating laminae [1, 3, 8, 9, 17]. A complete description of the previously described abnormalities of the posterior axis are described in Table 1. **c** Embryology of the craniocervical junction. *Yellow* occipital sclerotomes 1&2, *Magenta* occipital sclerotome 3, *Purple* occipital sclerotome 4, *Light Green* first spinal sclerotome, *Dark Green* first and second spinal sclerotomes, *Brown* second spinal sclerotome

the body by any osseous connections. At the time of surgery there were fibrous connections seen, however. Fibrous attachments to a hypoplastic arch of the axis have been described previously in two cases, with the spinous process and extremely hypoplastic lamina attached to pedicles and are thought to represent a more delayed error in embryogenesis [2, 5]. In these previous reports there was also a significant caudal pull of the residual posterior structures. These authors hypothesized that the part connection with the laminae of C3 might retain this structure in the spinal canal or the paravertebral muscles, such as the semispinalis, attached to the spinous process of the axis and pulled the laminae in the caudal direction [5]. Although such caudal migration was absent in our case, the fibrous attachments visualized potentially contributed to the ventral invagination and posterior compression present by inhibiting normal expansion of the spinal canal during normal ossification.

Interestingly, no associated congenital abnormalities at contiguous levels, particularly in the atlas, were seen in this case, which has been commonly seen with such deformities. Chau et al. [18] reported a complete absence of the posterior arch of the atlas in association with deficiencies of the posterior axis resulting in myelopathy. Behari et al. [5] described two cases of congenital atlantoaxial dislocations both with aplasia of the posterior arch of the axis in addition, while Suzuki et al. [11] reported a case of congenital C1/2 stenosis with C2–3 assimilation posteriorly. The absence of other congenital abnormalities might explain the delayed development of myelopathy in the end of the fifth decade of life seen in our case.

Concordant C2–3 OPLL and an invaginated C2 lamina resulting in myelopathy have yet to be reported. Chau described a small disk herniation contributing to an already compromised spinal canal at C2–3, due to an invaginated and dysplastic posterior arch, but no characteristics of OPLL were seen [18]. The etiology and pathophysiology of OPLL are widely discussed with no clear consensus [19, 20]. Epidemiological studies have identified several risk factors for its development, including, gender, trauma, hormonal imbalance, diet, genetics and diabetes mellitus [21–23]. Instability is also thought to be another potential factor contributing to the development of OPLL. We believe that instability at the C2–3 segment was the major contributing factor in our particular case. Deficiency in the posterior structures of the axis can conceivably result in an increased mechanical loading of the anterior elements. This is evidenced by the development of disk degeneration disproportionate to the remaining subaxial segments and the occurrence of OPLL isolated to the C2–3 level, both of which are rare occurrences at this level. Instability in conjunction with deficiencies of the posterior structures of the axis has been documented before [24, 25]. In

previously described cases, instability was typically manifested as anterolisthesis of C2 on C3, which was not seen in our patient. We attribute this to the attempt at self-stabilization by the development of OPLL at the most mobile portion of the segment, i.e., the disk level, and not the more common retro-vertebral location.

Because of our assumed segmental instability of the C2–3 segment, we chose to perform a fusion in addition to a decompression. In many previously reported cases of posterior abnormalities of the axis resulting in myelopathy, the surgical intervention was limited to removal of the compressive abnormal structures with favorable neurological outcomes resulting. These patients all were assumed to lack segmental instability as a contributing factor to their myelopathy. When posterior axis aplasia results in segmental instability at C2–3, surgical fusion has been shown to be required to guarantee a successful neurological outcome. This presumably relates to the need to improve alignment in cases of anterolisthesis or to eliminate dynamic cord compression. Morizono [24] and Trivedi [25] both reported cases of posterior axis aplasia resulting in anterolisthesis of the C2 body relative to C3 in patients with neck pain but no neurological findings. Treatment varied between the two with Morizono recommending non-operative means and Trivedi an occipitocervical fusion. A handful of other cases have been reported with documented instability at C2–3 with aplasia or hypoplasia of the posterior axis and myelopathy [4, 5, 26]. In such cases cord compression was generally caused by the posterior body of C3 and other anterior structures. All of these cases required both a decompression and stabilization to obtain a successful outcome. As Goel [4] described, performing a fusion without adequate decompression can lead to a poor clinical outcome and revision surgery. No similar cases with associated instability and myelopathy have been treated without a stabilization procedure.

Conclusions

Deficiencies of the posterior arch of the axis have rarely been described with only several case reports having been reported. Isolated OPLL at the C2–3 segment is also a rare occurrence. This article presents a case of circumferential cervical stenosis at C2–3 resulting in clinical myelopathy as a result of the simultaneous occurrence of these two rare abnormalities. This has not previously been reported to our knowledge. Successful management was obtained following surgical decompression and stabilization.

Acknowledgment Approval by the authors' institutional review board (IRB) was obtained. No Funding was received for this project

Conflict of interest None.

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