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Nature or nurture: a latent ossification of the posterior longitudinal ligament after atlantoaxial fusion. Illustrative case

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BACKGROUND The natural history of ossification of the posterior longitudinal ligament (OPLL) remains poorly understood and multiple etiologies have been reported. However, most have focused on the characteristics of the patient rather than alternation of mechanical stress after spinal fusion.

OBSERVATIONS This report describes, for the first time, a de novo OPLL found at the subaxial cervical spine 7 years after an atlantoaxial fusion surgery. A 57-year-old female initially required atlantoaxial arthrodesis for os odontoideum and stenosis that caused myelopathy. The posterior fusion surgery went smoothly without complications and the patient had good recovery of neurological functions. There was no associated instability, trauma, or reoperations during the follow-up. Seven years later, the patient presented with slight neck pain and a newly developed OPLL at C3–4 caudal to the C1–2 fusion construct.

LESSONS Conflicting with the conventional concept that OPLL is common in elderly men with genetic or hormonal factors, or associated spondyloarthropathies, OPLL could develop in women even after solid C1–2 fusion. The adjacent subaxial cervical spine is not free of risks for subsequent development of OPLL and cervical spondylotic myelopathy. This case illustration extends the scope of etiologies of OPLL within the present literature.

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KEYWORDS ossification of the posterior longitudinal ligament; OPLL; atlantoaxial fusion; C1–2 subluxation; de novo OPLL

Ossification of the posterior longitudinal ligament (OPLL) is a relatively rare pathological process that can lead to a wide range of symptoms, from completely asymptomatic to severe nerve compression and spinal cord injury.[1] Higher incidences are observed in elderly and male patients of Asian populations.[2,3] The pathophysiology, etiology, and natural history of OPLL remain poorly understood. Multiple risk factors have been identified and divided into primary (idiopathic) and secondary etiologies, including hormonal disorders and degeneration.[4,5] However, despite all the current knowledge, there is a paucity of data to clarify the natural history of OPLL. It is unclear how the OPLL first seeds and grows at which vertebral levels. Commonly proposed risk factors of OPLL focus on personal physical characteristics, while few involve the mechanical stress alteration corresponding to dynamic change after any intervention, such as surgical procedures of fusion. Segmental fusion of the cervical spine reportedly might result in shifting the load to the adjacent segments, and thus enhanced mechanical stress, which might lead to adjacent segment degeneration.[6] Therefore, it might be reasonable to infer that previous surgical interventions could play some role in OPLL development.

ABBREVIATIONS CT = computed tomography; CVJ = craniovertebral junction; OPLL = ossification of the posterior longitudinal ligament.

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In common scenarios, these patients usually present with preexisting OPLL that caused symptoms of various degrees. Therefore, the vast majority of research has debated the various approaches and optimal timing of surgical intervention. Very few studies address the subsequent changes of the bulk after surgery or the very initial growth of OPLL. In fact, most spine surgeons rarely catch the opportunity to observe the development of OPLL from scratch.

This case illustration describes, for the first time, a de novo OPLL at an adjacent mobile spinal segment after spinal fusion surgery of the most cephalad cervical spine. The clinical and radiological findings are detailed. According to this rare but valuable experience, the authors sought to address the acquired development of OPLL in a middle-aged female. Different from the commonly referred etiologies of personal characteristics, a biomechanical concept is proposed, such that the mechanical stress alternation after atlantoaxial fusion surgery can nurture OPLL at the subaxial cervical spine.

Illustrative Case

A 57-year-old female had no other medical history of underlying diseases such as autoimmune disease, endocrine disease, or paravertebral ligamentous ossification and neither did her family. She also had no previous history of spine surgery. She sustained persisting neck stiffness and numbness over four limbs for months. Progressive weakness of all extremities was noted. Upon presentation to our clinic, the patient manifested significant ataxia. On physical examination, the remarkably increased deep tendon reflexes indicated upper motor neuron disease, which prompted a thorough investigation. A series of clinical and radiological studies finally made the diagnosis of cervical myelopathy due to os odontoideum associated with atlantoaxial subluxation and stenosis at the craniovertebral junction (CVJ) (Fig. 1).

After admission, the patient first received a halo-ring traction for alignment correction and then halo-vest external fixation. Subsequently, standard posterior fusion of C1–2 with screw instrumentation and wiring of autologous bone grafts was performed. The operation included insertion of bilateral C1 mass screws and C2 pars screws, and posterior wiring by Sonntag method with autologous bone graft from the iliac crest. A decompressive procedure was not necessary as the atlantoaxial alignment had been corrected adequately after reduction of the C1–2 subluxation. After surgery, the patient significantly improved from myelopathy and was discharged home on postoperative day 6. Upon discharge, the patient could walk independently with full muscle strength and steady gait. A series of imaging studies taken 1 year later verified completely clinical improvement by radiological clearance of the preoperative pathologies (Fig. 2). The lordotic angle at C1–2 level was successfully restored and there was no more stenosis at the CVJ. Solid bone fusion was noted at the fractured dens and the posterior elements of C1 and C2 by computed tomography (CT) scans. Furthermore, adequate expansion of the dura sac associated with release of the spinal cord from compression over the CVJ was demonstrated. These radiological findings were compatible with the gradual and stable recovery of the clinical manifestations.

The postoperative clinical course was uneventful until 7 years postoperatively. The patient complained of insidious neck pain for a while, although she stayed neurologically intact. There were no symptoms or signs of myelopathy. The plain radiographs confirmed the proper alignment of the cervical spine, and good bony fusion between C1 and C2. However, a new-onset OPLL was found incidentally by CT scans. The OPLL was mainly located at the C3–4 levels (Fig. 3). Since there was no corresponding new-onset neurological deficit, symptomatic treatment and clinical observation were recommended. To date, the patient has remained free of long-tract signs.

Discussion

The present case illustration describes a subaxial OPLL that was newly developed after the atlantoaxial fusion surgery. The de novo OPLL was found near the initial surgical field, which was properly realigned and surgically fused with satisfactory results. According to the clinical history and radiological findings, for uncertain reasons the development of this OPLL might be strongly associated with prior atlantoaxial fusion surgery. To the best of current knowledge, previously identified factors of OPLL formation and progression have included genetic, hormonal, environmental, and lifestyle factors.[4,5] However, the present case is the first to suggest that

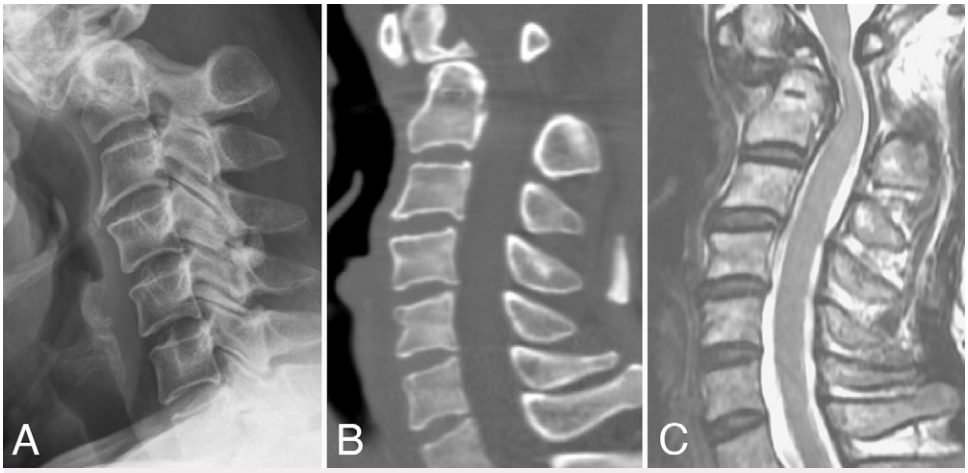

FIG. 1. Preoperative imaging studies confirming the diagnosis of os odontoideum: chronic fracture of the dens with C1–2 subluxation (**A and B**), severe stenosis at the craniocervical, and compression to the spinal cord (**C**).

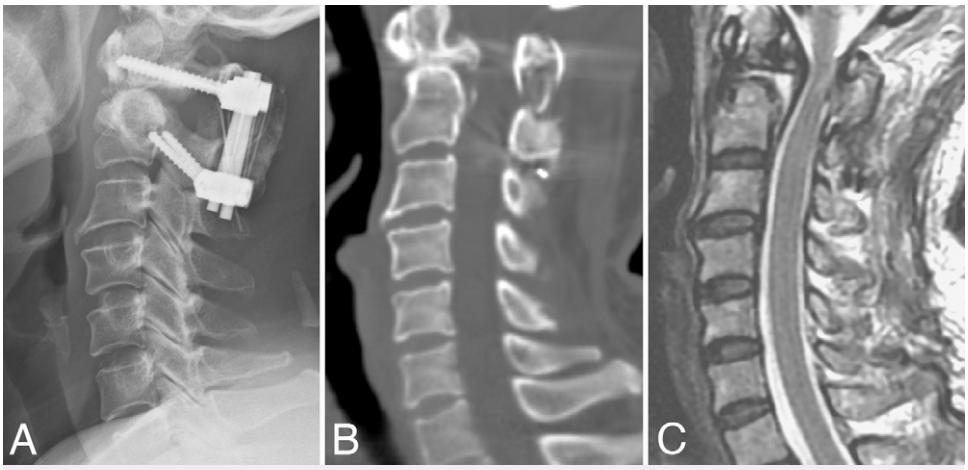

FIG. 2. A series of imaging studies obtained 1 year later verified radiological improvement from the operation: a restored lordotic angle at C1–2 level (**A**), solid bone fusion at the fractured dens and between the posterior parts of C1 and C2 (**B**), and adequate expansion of the dura sac associated with release of the spinal cord from compression over the craniocervical junction (**C**).

prior fusion surgery may be another contributing factor and therefore extends the scope of etiologies within the present literature.

It was reported that the C1–2 angle was significantly enlarged while the C2–7 angle was reduced significantly after the posterior atlantoaxial fusion surgery for patients of os odontoideum.[7–10] This evidence supports the prediction that the subaxial alignment parameters will also change in response to the correction of the C1–2 angle. In the present case, we noted the range of motion at the C3–4 level between neck flexion and extension increased from 4 degrees preoperatively to 9 degrees postoperatively. In addition, the rotation activity of C1–2 was inevitably greatly reduced after atlantoaxial fusion. Consequently, subaxial segments are bound to endure more mechanical stress. From the viewpoint of molecular pathophysiology, mechanical stress has been reported to promote OPLL formation via both increasing the expression levels of various genes inducing OPLL development and its progression, and decreasing the expression level of vimentin suppressing the mineralization in osteoblasts.[11] In addition, it was also suggested that OPLL cells would be transformed into cells that are highly sensitive to mechanical stress, which may induce the progression of OPLL.[12] Therefore, it is reasonable to assume that the mechanical stress aggravation as a consequence of atlantoaxial fusion nurtures the acquired OPLL at the subaxial cervical spine.

There might be debates on why the de novo OPLL just occurred at C3–4 level while spared at other subaxial segments. In the preoperative radiographs, there was remarkable osteophyte at the C3–4 level, suggesting preexisting instability and stress prior to the atlantoaxial fusion surgery. The authors propose that postoperative aggravated mechanical stress, together with the preexisting stress, led to the development of OPLL at the index level. Whether the OPLL should be managed by which surgical approaches, at what timing, and with what risks of complications remain controversial.[13–17]

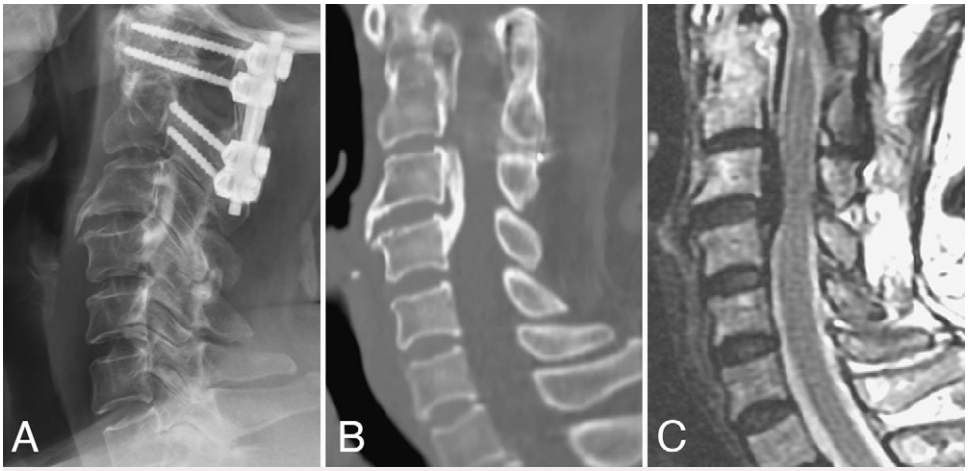

FIG. 3. A series of imaging studies obtained 7 years postoperatively revealing proper alignment of the cervical spine (**A**), good fusion between C1 and C2, and incidental findings of OPLL, mainly at the C3–4 level and partly at the C1–2 level (**B**), and corresponding spinal stenosis at the C3–4 level (**C**).

To sum up, OPLL is a multifactor disorder. Genetic factors are believed to contribute to OPLL development by evidence that OPLL is most commonly found in Asian populations. Some collagen genes, single nucleotide polymorphisms, and mutations have been proposed to be associated with OPLL.[5] Regarding endocrines and hormones, diabetes mellitus, hypoparathyroidism, and acromegaly have been reported to be related to OPLL.[4] Also, old age and mechanical stress have been reported as predisposing factors, implying that the OPLL formation can be a process of degeneration.[1–3,12] With the present case report, the authors point out that atlantoaxial fusion surgery may aggravate subaxial mechanical stress, thereby accelerating cervical spine degeneration. It is noteworthy that cervical spine surgery may predispose patients to adjacent OPLL formation.

Observations

This report describes, for the first time, a de novo OPLL found at the subaxial cervical spine 7 years after atlantoaxial fusion surgery. The 57-year-old female initially required atlantoaxial arthrodesis for os odontoideum and stenosis that caused myelopathy. The surgery went smoothly without complications and the patient had good recovery of neurological functions. There was no associated instability, trauma, or reoperations during the follow-up. Seven years later, the patient presented with slight neck pain and the follow-up radiological examinations demonstrated a newly developed OPLL at C3–4 with substantial narrowing of the spinal canal caudal to the C1–2 fusion construct. Since the postoperative course was free of other events, the de novo OPLL that developed at adjacent segments years postoperation might be attributed to the additional mechanical stress on the subaxial cervical spine following the atlantoaxial fusion surgery.

Lessons

Conflicting with the conventional concepts that OPLL is commonly found in elderly males with genetic or hormonal factors, or associated diseases (e.g., diffuse idiopathic skeletal hyperostosis, ankylosing spondylitis, and other spondyloarthropathies), OPLL could develop in females even after solid spinal fusion of C1–2. The adjacent subaxial cervical spine is not free of risks of subsequent development of OPLL, and follow-up for cervical spondylotic myelopathy is thus warranted. This case illustration extends the scope of etiologies of OPLL within the present literature.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Ko, Tzeng, Wu. Acquisition of data: Tzeng, CH Kuo, Tu, CC Chang, Wu. Analysis and interpretation of data: Tzeng, YH Kuo, CH Kuo, Tu, CC Chang, Wu. Drafting the article: Tzeng, HK Chang. Critically revising the article: Ko, CH Kuo, HK Chang, Tu, CC Chang. Reviewed submitted version of manuscript: Ko, HK Chang. Approved the final version of the manuscript on behalf of all authors: Ko. Administrative/technical/material support: YH Kuo, Cheng, Huang. Study supervision: Ko, Cheng, Huang.

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