



CASE REPORT

Congenital double-level cervical spondylolysis: a case report and review of the literature

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Abstract

Purpose We report a rare Japanese female who was affected with three genetic-linked diseases: double-level cervical bilateral spondylolysis in association with spina bifida occulta, cleft lip and monostotic fibrous dysplasia of the right proximal femur. The case was considered to be congenital in origin. We also review the pertinent literature of cervical spondylolysis, with a focus on the pathogenesis of multiple-level cervical spondylolysis.

Methods A 40-year-old female presented with progressive clumsiness and numbness of the hands. Japanese Orthopedic Association (JOA) score for the cervical spine was 14.5. Plain radiographs of the cervical spine showed bilateral spondylolysis of the articular mass portion, with an adjacent dysplastic change and spina bifida occulta of C4 and C5. Cervical laminoplasty from C4 to C6 was performed.

Results The postoperative course was uneventful, and the patient had some recovery of muscle power and sensation, with JOA score improving to 15.5. At the 8-year follow-up, the patient had no recurrence of symptoms, but did show kyphotic and degenerative changes at the C4/5 and C5/6 level with no apparent instability.

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Conclusions This case is a rare presentation of bilateral cervical spondylolysis involving C4 and C5, presumably congenital, accompanied by combined dysplastic changes of the cervical spine, cleft lip, and fibrous dysplasia, possibly through an error involving an ossification center during the embryonic stage.

Keywords Cervical spondylolysis · Multiple-level cervical spondylolysis · Cleft lip · Fibrous dysplasia · Congenital

Introduction

Cervical spondylolysis is described as a cleft in the articular mass of a cervical vertebra that occurs at the junction of the superior and inferior facet joints and is considered the cervical equivalent of pars interarticularis in the lumbar spine [1]. Hadley first described cases of cervical spondylolysis in 1946 as a congenital separation of the pedicles from a cervical vertebra [2]. Since then, just over 100 cases have been reported in the English literature, mainly in adults [3].

Lumbar spondylolysis is a common condition and is reported to occur in around 6% of the general population [4]. However, multiple-level lumbar spondylolysis occurs in only 0.3% of the general population [4]. Multiple-level cervical spondylolysis is even rarer, occurring far less frequently than multiple-level lumbar spondylolysis. To our knowledge, only four cases of multiple-level cervical spondylolysis have been reported in the literature [5–8].

We herein report a case of myelopathy due to bilateral spondylolysis of C4 and C5, associated with dysplastic changes at the involved level and a history of cleft lip and monostotic fibrous dysplasia, which was presumably

congenital in origin. We also review the pertinent literature of cervical spondylolysis, with a focus on the pathogenesis.

Case report

A 40-year-old female presented with progressive clumsiness and numbness of the hands, especially in the left hand. She had monostotic fibrous dysplasia in the right femur and had undergone an operation to correct a cleft lip soon after birth. There was no history of trauma.

A physical examination revealed no remarkable posterior neck pain. The neck motion was not limited. Muscle strength was reduced to 4/5 in the left myelopathic hand. The patient presented with numbness of the hands and the left leg, mainly in the left hand. The tendon reflexes of both upper extremities were slightly diminished. Both patellar tendon and Achilles tendon reflexes were markedly increased bilaterally. Spurling's foraminal compression test was positive on the left side. She had no bladder or bowel dysfunction. Japanese Orthopedic Association (JOA) score for the cervical spine was 14.5 out of 17 points. There were no pigmented skin lesions or endocrinopathy, and no complaints of pain in other bones.

Plain radiographs of the cervical spine showed bilateral spondylolysis of the articular mass portion, with an adjacent dysplastic change and spina bifida occulta of C4 and C5 (Fig. 1). The posterior fragments were not displaced posteriorly. The sagittal alignment of the cervical spine was regarded as a loss of cervical lordosis and a segmental kyphosis at the C3–C6 level. Lateral dynamic views revealed no apparent intervertebral instability. Plain radiographs of the lumbar spine did not show lumbar spondylolysis or spina bifida occulta of the lumbar spine.

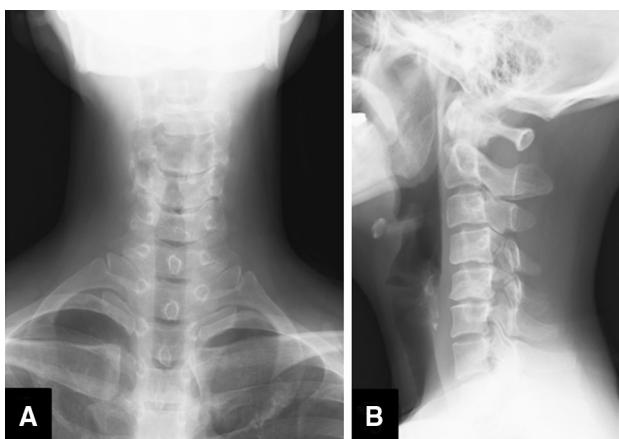


Fig. 1 **a** In an anteroposterior view of the cervical spine, spina bifida occulta of C4 and C5 was found. **b** A lateral radiograph showed a well-corticated defect between the superior and inferior articular pillars of C4 and C5, associated with hypoplasia of the pedicle, and dysplastic lamina without spondylolisthesis

Magnetic resonance imaging (MRI) of the cervical spine revealed cervical spinal canal stenosis. The spinal cord was severely compressed at the C4/5 and C5/6 level, and high signal change was found in the spinal cord on T2-weighted images. The vascular abnormalities of vertebral artery were not found.

We decided to perform only a cervical laminoplasty from C4 to C6 without fusion because the patient did not have severe neck pain or any apparent instability. Among the intraoperative findings, bilateral spondylolysis and spina bifida occulta at C4 and C5 were recognized, and the lamina at these levels was completely unstable. Decompression of the dura mater was sufficiently achieved.

The postoperative course was uneventful, and the patient had some recovery of muscle power and sensation, with JOA score improving to 15.5. At the 8-year follow-up, the patient had no recurrence of symptoms, but did show slightly progressive kyphotic and degenerative changes at the C4/5 and C5/6 level with no apparent instability (Fig. 2).

Computed tomography (CT) taken postoperatively showed a well-corticated, smoothly marginated spondylolytic defect in the mid-articular mass, and dysplastic bilateral lamina (Fig. 3). MRI showed sufficient decompression with residual high signal change in the spinal cord (Fig. 4).

Discussion

Etiology

Cervical spondylolysis is an exceedingly rare condition. It is usually diagnosed after minor trauma or as an incidental finding on a radiographic examination. It most commonly

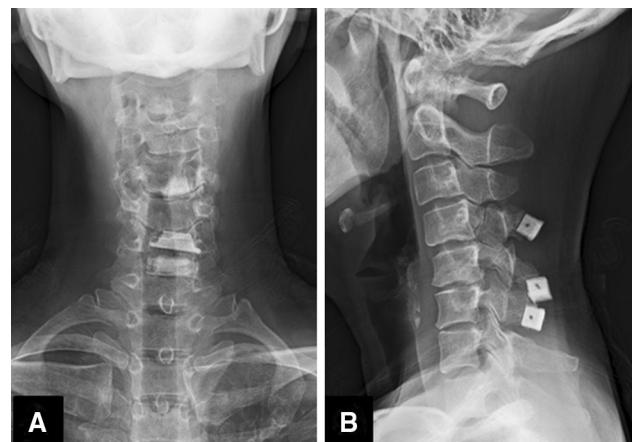


Fig. 2 **a, b** Eight years after cervical laminoplasty of C4–6. A lateral radiograph showed kyphotic and degenerative changes at the C4/5 and C5/6 level

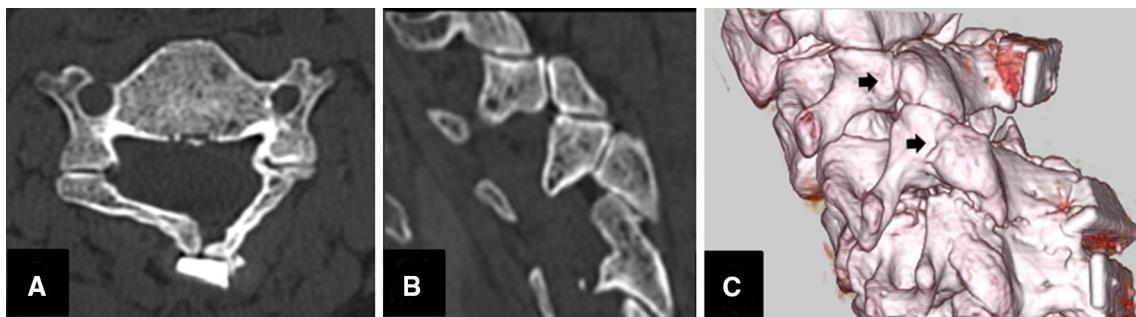


Fig. 3 **a, b** Postoperative axial CT image of C4 and parasagittal CT image revealed a spondylolytic cleft with a sclerotic margin at the pedicle laminar junction. Dysplasia of the pedicle, articular pillar, and

lamina on both sides were recognized. **c** Three-dimensional CT revealed spondylolytic cleft (arrows) in the dysplastic articular masses of C4 and C5

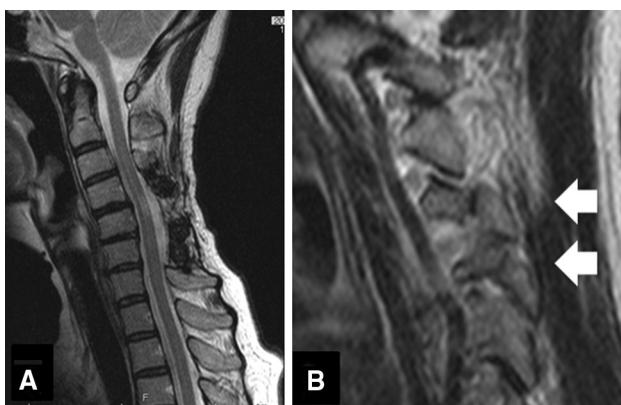


Fig. 4 **a** A postoperative T2-weighted MR image showed sufficient decompression with residual high signal change in the spinal cord. **b** A parasagittal T2-weighted MR image showed the cleft of the articular pillar of C4 and C5 (arrows), which displayed as a thin band of low signal intensity on both T1- and T2-weighted MRI, possibly indicating fibrous connective tissue

occurs at a single level, but has been reported to occur at any level, with the exception of C1 [1, 3, 9, 10]. In 142 case reports, C6 was the most common site of involvement, present in 67 cases (48%), followed by C4 and C5 [3]. Bilateral spondylolysis of the cervical spine has been reported to account for only 25% of 70 cases [1]. Among 69 cases of cervical spondylolysis, 42 were accompanied by spondylolisthesis, but only four were unilateral. Spondylolisthesis appears to mainly be associated with bilateral cases [11]. Spina bifida occulta is another common finding and is reported to be present in around 60% of all cases of cervical spondylolysis [1, 11]. However, it is found in 86% of cases with bilateral defects [11].

Multiple-level cervical spondylolysis

Multiple-level lumbar spondylolysis and spondylolisthesis occur more often in men than in women [4]. Most cases of multiple-level lumbar spondylolysis occurred at two spinal levels from L1 to S1, mainly at the L3–L5 level, and

were associated with sports, trauma, or heavy labor [12]. Similar to the etiology of single-level spondylolysis, the etiology of multiple-level lumbar spondylolysis is thought to be fatigue fractures [12].

In contrast, multiple-level cervical spondylolysis is an extremely rare entity. In the literature where it has been clearly described and defined, to our knowledge, only four cases have been reported, including C4 and C5; C5 and C6; C3, C4, and C5; C2, C4, and C6 [5–8] (Table 1). No cases involving myelopathy and long tract sign secondary to repetitive micro-instability due to multiple-level bilateral spondylolysis have been reported in the literature.

Klimo et al. [13] described the malformation of the spine as “congenital multilevel cervical disconnection syndrome” in children who had multilevel bilateral defects of the osseous structures (namely, the pedicle). This cervical spinal anomaly is distinct from cervical spondylolysis and absent pedicle syndrome which is always absent the pedicle unilaterally [14].

Pathogenesis

The exact cause of cervical spondylolysis remains controversial. Several theories have been proposed, including congenital, embryologic developmental, fatigue fracture, and posttraumatic nonunion with pseudarthrosis [3, 6], although there is no definitive evidence to support these theories.

Morvan et al. pointed out that the most common site affected is C6, because it is a transitional vertebra and is subjected to more stress than other cervical segments [15]. Cervical spondylolysis can thus be regarded as the mechanical response to the microtraumatism on an initially dysplastic vertebral structure as opposed to an acute fracture, as has been proposed in the literature, especially on C6 [1, 3, 9, 15]. In the patients with congenital anomalies [1], C6 spondylolysis was 74%, more common as compared with spondylolysis caused by acute trauma (30%) [3].

Table 1 Literature review findings of reported cases of multiple-level cervical spondylolysis

| Authors, year | Age (years)/sex | Speculated etiology | Symptom | Spondylolysis | Spondylolisthesis | Other findings | Treatment | Follow-up |
|-----------------------------|-----------------|----------------------------|--------------------------------|--|-------------------|-------------------------------------|---|------------------------------|
| Prioleau et al. [5], (1975) | 46/F | Congenital | Neck pain, radiculopathy | Bilateral C3–C5 | C5 on C6 | None | Anterior cervical discectomy and fusion | Without complication |
| Schwartz et al. [6], (1982) | 8/M | Congenital (Down syndrome) | None | C4–C5 (bilateral) | C4 on C5 | Spina bifida occulta C5 | None | None |
| Yochum et al. [7], (1995) | 24/M | Congenital | Neck discomfort, radiculopathy | Bilateral C2 and C4, unilateral C6 (left side) | C4 on C5 | Spina bifida occulta C2, C4, and C6 | Conservative chiropractic management | Complete resolution |
| Garin et al. [8], (1995) | 14/M | Trauma (rugby) | Neck pain | Bilateral C5–C6 | None | Spina bifida occulta C5 | Conservative management | Pain disappeared |
| Present case, (2016) | 48/F | Congenital | Myelopathy, long tract sign | Bilateral C4–C5 | None | Spina bifida occulta C4–5 | Laminoplasty | Residual numbness at 8 years |

F female, M male

The congenital theory suggests that spondylolysis results from a failure of the two centers of chondrification to fuse at the time of ossification after week 8 of gestation [1, 13]. Oh et al. also suggested that the failure of the development of a vertebral chondrification center for the posterior arch of a particular sclerotome, or failure of appropriate ossification, may lead to posterior arch defect, including the absence of a pedicle and spondylolysis [14]. Dysplastic changes at the involved level and spina bifida occulta support congenital and/or developmental theories [1, 16]. Congenital cervical spondylolysis may, therefore, be the counterpart of congenital lumbar spondylolysis [17].

Cleft lip

Cleft lip is a common birth defect linked to genetic influences and is associated with other malformations, including spina bifida occulta [18]. Anomalies of cervical vertebra, such as posterior arch deficiency including cervical spondylolysis, have been reported to occur more frequently in patients with cleft lip and palate [19]. The lip forms between weeks 4 and 8 of gestation. Cleft lip and posterior arch deficiency anomaly of the cervical spine arise from the same paraxial mesoderm. The mechanism involved in palatal shelf fusion during embryonic development also influences the development and fusion of the posterior arch of the cervical spine [18]. Although cleft lip has never been documented together with cervical spondylolysis and fibrous dysplasia, genetic mutations may be the cause of these three conditions.

Familial incidence implicates a congenital disorder, such as a genetic disorder. Saltzman et al. reported on 9 of 12 family members from four generations affected by a form of inherited cervical vertebral dysplasia [20].

Treatment

Most patients with cervical spondylolysis have normal neurologic examination results, although myelopathy caused by instability or spondylolisthesis has been rarely reported [21]. Only two cases (2.8%) among 70 with cervical spondylolysis have shown distinct long tract signs [11]. Surgical treatment for cervical spondylolysis is indicated in cases with symptoms, instability, spinal cord compression, and neurologic deficits [21]. The need for surgery to relieve spinal cord compression is rare. A biomechanical study indicated that increased disc stress induced by hypermobility of cervical kinematics in the setting of cervical spondylolysis is more likely to lead to cervical spinal cord injury than in an intact cervical spine [22]. Combined dysplastic changes adjacent to the spondylolysis may increase the risk of unstable posterior column of the involved cervical vertebra, resulting in instability or cord compression due to spondylolisthesis [16]. When the patient had instability or spondylolisthesis, spinal fusion such as anterior cervical decompression and fusion was indicated.

In the present case, the patient had myelopathy and long tract sign secondary to repetitive micro-instability due to multiple-level bilateral spondylolysis, but no apparent

instability on dynamic radiographic imaging. This condition was caused by repetitive microtrauma, resulting in degenerative changes. Secondary degenerative changes over time due to abnormal spine mechanics may occur, particularly in older patients [1]. Spinal fusion is recommended in the event of an intensification of the radiographic instability or the recurrence of neurological symptoms.

Conclusions

This case is a rare presentation of bilateral cervical spondylolysis involving C4 and C5, presumably congenital, accompanied by combined dysplastic changes of the cervical spine, cleft lip, and fibrous dysplasia, possibly through an error involving an ossification center during the embryonic stage. Spinal fusion may be recommended for multiple-level cervical spondylolysis even without radiographic instability, depending on the symptoms.

Compliance with ethical standards

No funds were received in support of this work. No relevant financial activities outside the submitted work.

Conflict of interest No conflicts of interests exist.

Informed consent Informed consent was obtained from the participant included in the study.

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