

Atlas Hypoplasia Combined with Atlantoaxial Subluxation is a Cause of Non-traumatic Cervical Myelopathy

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Congenital abnormalities of the posterior arch of the atlas (C-1) are uncommon. They may range from partial clefts to total agenesis of the posterior arch. Atlas hypoplasia may cause segmental compression and cervical myelopathy in elderly patients. In this report, we present a case of atlas hypoplasia combined with atlantoaxial subluxation. The patient developed cervical myelopathy gradually for 6 months without preceding trauma. Neuroimaging studies revealed a unilateral cleft of the posterior C-1 arch and widening of the predental space which caused marked compression of the spinal cord at the atlas level. A remarkable neurological recovery followed the removal of the hypoplastic C-1 arch and the constricted ligament along with posterior occipital-C-3 fusion. Hypoplastic posterior arch of C-1 may create marked focal stenosis of the spinal canal and the cord may be further compromised by atlantoaxial subluxation. Posterior decompression combined with occipital-C-3 fusion is an effective treatment for symptomatic patients with such a condition. (*Mid Taiwan J Med* 2003;8:99-104)

Key words

atlas hypoplasia, atlantoaxial subluxation, cervical canal stenosis, cervical myelopathy, posterior occipital-C-3 fusion

INTRODUCTION

Congenital abnormalities of the posterior arch of the atlas (C-1) are uncommon, and may range from partial clefts to total agenesis of the posterior arch. Neurological symptoms may occur after minor cervical trauma in patients with isolated partial agenesis of the posterior arch of the atlas [1-3]. There is, however, increasing evidence that non-traumatic cervical myelopathy may be caused by atlas hypoplasia [4-6]. We present a case of nontraumatic cervical myelopathy caused by unilateral hypoplastic posterior C-1 arch and atlantoaxial subluxation.

CASE REPORT

This 72-year-old woman presented with

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progressive numbness in both hands and weakness of the right upper and lower limbs. These neurological presentations began insidiously 6 months ago without preceding trauma. On admission, she was unable to stand without support of a cane. Coordination of her hand was also impaired.

Physical Examination

The results of a general examination were normal. Tongue and palate movements were also normal. There was neither atrophy nor fasciculation of the sternocleidomastoid muscles. Neurological examination showed hemiparesis and hemihypesthesia on the right side. Muscle power was low in all four limbs but was worse on the right side. Marked wasting of the right deltoid muscle was found. Muscle power of right shoulder abduction was MRC grade 2/5 (Royal Medical Research Council of Great Britain



Fig. 1. Lateral view of the plain radiographs of the cervical spine demonstrates hypoplasia of the posterior arch of C-1, and subluxation of the atlantoaxial joint.



Fig. 2. Axial section of the CT scan shows a hypoplastic posterior C-1 arch (white arrow), and increased prevertebral space due to atlantoaxial subluxation (black arrow).



Fig. 3. Three-dimensional CT demonstrates markedly increased prevertebral space, measuring 19.78 mm (black arrow), and marked spinal canal stenosis due to cleft of the posterior arch of C-1 and atlantoaxial subluxation (white arrow).



Fig. 4. Midsagittal T1 weighted MR image of the cervical spine shows a hypoplastic posterior C-1 arch resulting in severe constriction of the upper cervical canal and cord compression.

strength grading scale). Muscle tone was markedly high in the lower limbs, and Hoffman sign was noticed. The patient's gait was moderately spastic and the Babinski's reflex produced a dorsal response.

Neuroimaging Studies

Plain radiographs of the cervical spine and helical three-dimensional CT disclosed a markedly narrow canal at the level of the atlas due to a cleft of the right posterior C-1 arch and atlantoaxial subluxation (Figs. 1, 2). The

prevertebral space was 19.78 mm (Fig. 3) in diameter which was significantly wider than the normal limits. Cervical spondylosis was present below the level of C-2. Magnetic resonance imaging and CT scan of the cervical spine showed a hypoplastic posterior C-1 arch resulting in severe focal constriction of the cervical canal with cord compression at the C-1 and C-2 levels (Fig. 4).

Operation

The patient was intubated with a fibroscope

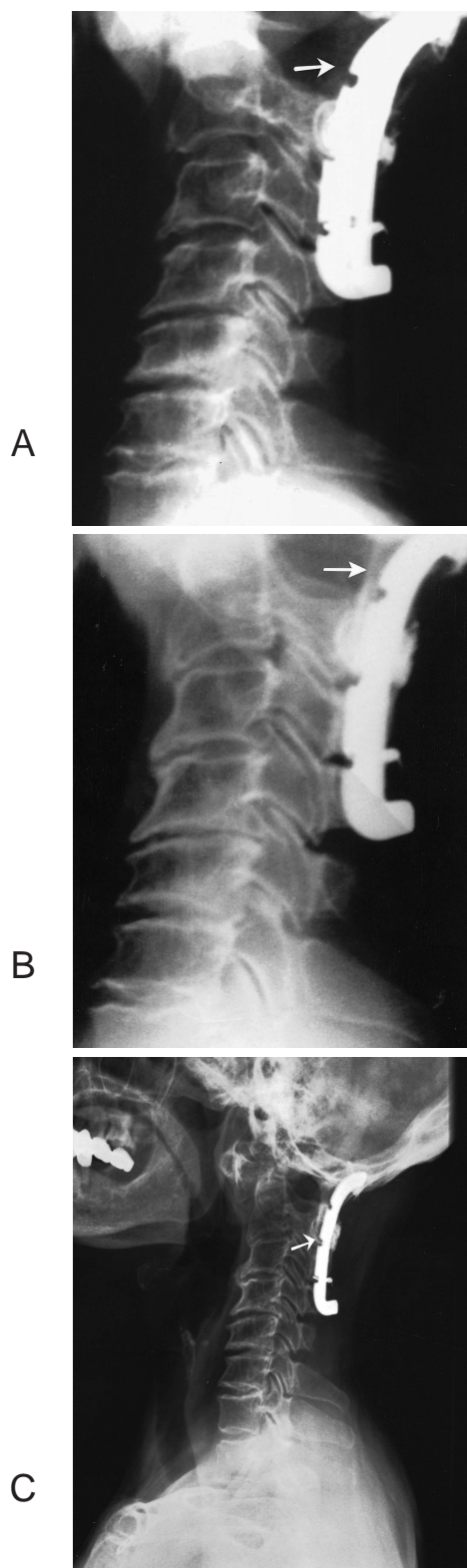


Fig. 5. A: Immediate post-operative plain radiograph of the cervical spine demonstrates widening canal after decompression and fusion; B: The follow-up radiograph of the cervical spine 6 months later, and C: Successful fusion of the occipital bone to C-3 vertebra 12 months after surgery.

when she was awake and then turned to a prone position to monitor the neurological status during positioning. General anesthesia was induced, and the reduction of the dislocated atlantoaxial joint was performed under the guide of C-arm fluoroscopy. The surgical approach was performed from the posterior midline and a partial cleft on the right posterior C-1 arch was identified. The spinal cord was compressed by a hypertrophied ligament on the right side and hypoplastic C-1 arch on the left side. The underlying dura was indented at that level and expanded after removing the hypoplastic C-1 arch and the constricted ligament. Posterior occipital-C-3 fixation was performed by SOF'WIRE cable system and Tiframe plate-cable fixation (Codman & Shurtleff, Johnson & Johnson) (Fig. 5A). Two drill holes were placed into the occipital bone 2 cm superior to the rim of the foramen magnum. Wires were passed from the holes to the foramen magnum. Another three sublaminar wires were also passed safely under the C-2 and C-3 laminae. Tiframe was fixed with the above five wires for stabilization. The occipital bone and lateral masses of C-1, C-2 and C-3 were decorticated. Autologous bone harvested from the right posterior iliac crest was overlaid on the decorticated rough bone surface. Finally, the wound was closed in layers.

Postoperative Course

The patient was able to walk without assistance one week after surgery. The muscle power of the right deltoid muscle began to improve (shoulder abduction MRC grade 4/5) and she was able to lift her right upper limb when she was discharged from the hospital. Muscle tone and deep tendon reflex were still high in the lower limbs.

Post operative plain film of the cervical spine taken 6 months and 12 months later revealed successful bony fusion from the occipital bone to C-3 (Figs. 5B, 5C).

DISCUSSION

The atlas has three primary ossification centers during embryonic period [7]. In approximately 2% of the population, a fourth

ossification center forms on the posterior tubercle of the atlas. During the ossification process of the atlas, two different types of anomalies can develop: 1) the formation of a posterior cleft, and 2) varying degrees of posterior arch dysplasia [8]. Our case belongs to type 2 because of a partial cleft of the posterior arch of the atlas.

The older classification of congenital clefts and defects of the posterior arch of the atlas was divided into five types [1]. Type A: failure of midline fusion of the two hemiarches. It is estimated to occur in 4% of the population and represents 97% of all posterior arch defects [9]. Type B: Unilateral cleft which ranges from a small gap to a complete absence of the hemiarch and posterior tubercle. Our case was classified as type B. Type C: Bilateral clefts of the lateral aspects of the arches with preservation of the most dorsal part of the arch. Type D: Absence of the posterior arch with a persistent posterior tubercle. Type E: Absence of the entire arch including the tubercle. Types B to E occur in 0.69% of all individuals. The classification system was reviewed by Currarinos et al [1], and clinically subdivided into five groups: Group 1, asymptomatic with incidental findings; Group 2, neck pain or stiffness after trauma to the head or neck; Group 3, Chronic symptoms related to the neck or cervical spine; Group 4, various chronic neurological problems; and Group 5, acute neurological symptoms after minor cervical trauma [1-2]. This patient belonged to Group 4 because of her chronic symptoms and neurologic deficits.

The average sagittal canal diameter ranges from 17 to 25 mm at the level of the atlas; the cord diameter ranges from 10 to 12 mm [10]. In cases of cranio-cervical junction lesions, cord compression occurs when the sagittal diameter behind the dens is 14 mm or less [11]. Patients with a sagittal canal diameter of less than 10 mm usually show clinical manifestations [10,12,13]. Payne and Spillane were the first to notice the importance of congenital stenosis of the spinal canal and suggested that the development of myelopathy may also be related to the initial size

of the canal [14]. If the spinal canal is congenitally small, the spinal cord will be encroached upon easily by osteophyte, vertebral subluxation, or hypertrophied ligamentum flavum. Our patient also had subluxation of the dens. Her retrodental space was only 8.8 mm. Posterior displacement of the dens while flexing the neck might further compromise the cervical cord. The combination of atlas hypoplasia, segmental cervical canal stenosis and atlantoaxial subluxation as found in our patient has only been reported by Duong et al [15].

A variety of surgical options are available for the treatment of atlantoaxial subluxation. The patient in this report had a narrow spinal canal caused by partial agenesis of the posterior arch of the atlas and atlantoaxial subluxation. To widen the canal, we decided to remove the remnants of the posterior arch of the atlas. In this situation, the application of C1-C2 arthrodeses is difficult in the absence of a posterior arch of C-1. There are two surgical options: 1) the use of bilateral transarticular screws to stabilize the lateral masses of C-1 and C-2, and 2) fusion of the occiput with C-2 and C-3. The first operation can be achieved by transarticular screw fixation of C-1 and C-2. However, the left lateral mass of C-1 was partially hypoplastic in our case. Therefore, this procedure is difficult to perform because the hypoplastic bone may be fractured while purchasing a screw larger than its diameter. Therefore, we applied posterior decompression, internal fixation with Tifram and bone graft for the treatment of atlas hypoplasia, segmental cervical narrowing, cord compression and cervical myelopathy in this case and the outcome was satisfactory. Solid bony fusion from the occiput to C-3 lamina and good neurological recovery of the patient were obtained.

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寰椎成形不全合併寰椎-樞椎滑脫導致非創傷性頸椎脊髓病變

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寰椎後椎弓的先天異常並不常見，它們可細分為後椎弓局部椎裂到完全缺失。寰椎成形不全可以導致成年人頸椎壓迫及脊髓病變。本篇，我們報告一個因寰椎成形不全合併寰椎-樞椎滑脫的案例，她並無創傷病史卻於近六個月來發展成頸椎脊髓病變，神經影像學檢查發現寰椎後椎弓單側裂及樞椎突前空間過寬導致頸椎脊髓明顯壓迫而造成頸椎脊髓病變，此病例在接受寰椎成形不全的後椎弓切除及後位枕骨-第三頸椎融合手術後有明顯的神經學改善，寰椎後椎弓成形不全可以導致椎管明顯狹窄，若又合併寰椎-樞椎滑脫則會加重頸椎脊柱的先天性狹窄而造成頸椎脊髓病變，後側減壓手術切除寰椎成形不全的後椎弓和周邊退化的韌帶加上枕骨-第三頸椎融合對這類有神經症狀的病人是一項有效的治療。(中台灣醫誌 2003;8:99-104)

關鍵詞

寰椎成形不全，寰椎-樞椎滑脫，頸椎椎管狹窄，頸椎脊髓病變，後位枕骨-第三頸椎融合

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