Cervical Spinal Stenosis and Myelopathy Due to Atlas Hypoplasia

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This paper describes a patient who presented at our hospital with neurologic symptoms due to congenital cervical spinal stenosis at the atlas level. Congenital atlantal stenosis is usually due to hypoplasia of the posterior arch of the atlas. It is a rare cause of spinal stenosis, and only 12 symptomatic patients with isolated atlantal stenosis have been reported. Current treatment is surgical decompression, and all reported patients receiving surgical treatment improved to some degree. [*J Chin Med Assoc* 2007;70(8):339–344]

Key Words: atlas, decompression, myelopathy, spinal stenosis

Introduction

Cervical spinal stenosis is mostly due to degenerative change or trauma. It usually occurs below C3 level. Symptomatic isolated atlantal stenosis is very rare; only 12 patients with symptomatic isolated atlantal stenosis have been reported in the literature. Most of these patients received surgical decompression and gained significant improvement after operation.

Case Report

The patient was a 38-year-old male with no major medical history. The patient suffered from tingling sensations in his abdomen and perineum while flexing his neck. Both hands had sustained numbness for 2 months. No major trauma history was noted. Neurologic examination revealed a deep tendon reflex of 3+ in the upper limbs and 4+ in the lower limbs. Neither spastic gait nor urinary or stool incontinence was noted. There were no motor or sensory deficits except for the sensation of numbness in both hands. Plain cervical X-rays revealed mild degenerative change without significant instability (Figure 1). Cervical computed tomography (CT) revealed an incurving of the posterior arch of the atlas (Figure 2). Magnetic resonance imaging (MRI) revealed marked stenosis at the C1 level with a sagittal canal diameter of 12.46 mm and a sagittal dural sac diameter of 6.23 mm at the atlas level (Figure 3). The patient received surgery for removal of the posterior arch of the atlas with duroplasty. After operation, the numbness in both hands and the abnormal tingling sensations in the abdomen and perineum improved.

Discussion

Cervical spinal stenosis usually occurs below C3, and rarely occurs above the C2 level.[1] It is usually associated with degenerative spondylosis or trauma. Symptomatic isolated atlantal stenosis is very rare, and up to the year 2005, only 12 patients with symptomatic isolated atlantal stenosis had been reported. All of these cases were without history of major spinal trauma or significant cervical spinal instability.

Among the 12 reported cases and the 1 case we present in this article, 10 were Asian, 9 presented with cervical myelopathy, 10 were adult-onset, and 12 received surgical decompression. All the patients who

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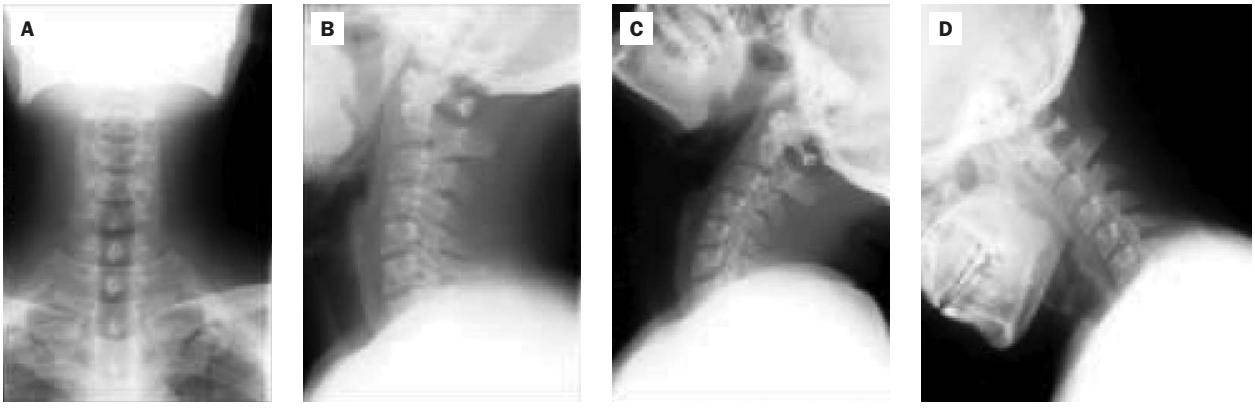

Figure 1. Plain cervical X-ray reveals mild degenerative change without significant instability.

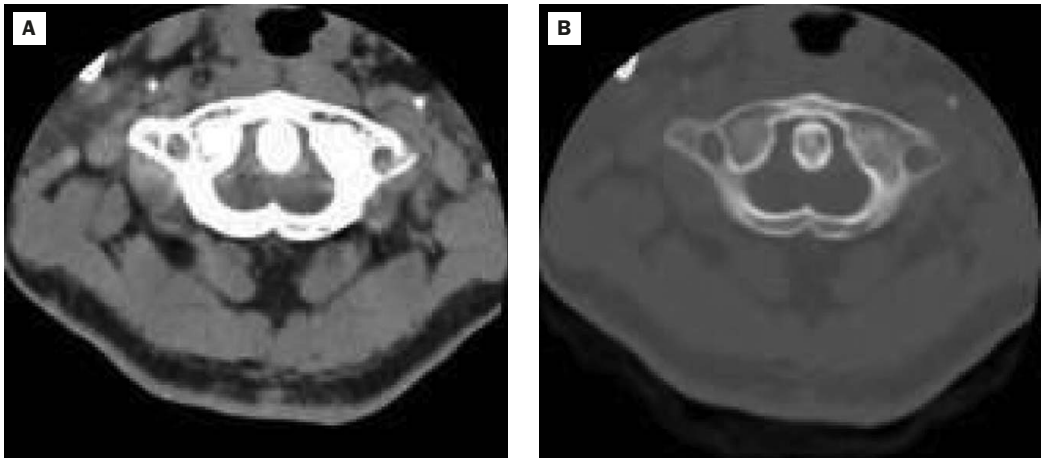

Figure 2. Cervical spinal computed tomography at the atlas level. Note the hypoplasia and incurving of the posterior arch, with ridge formation on the midline within the canal. The narrowest width of the canal is 12.46 mm.

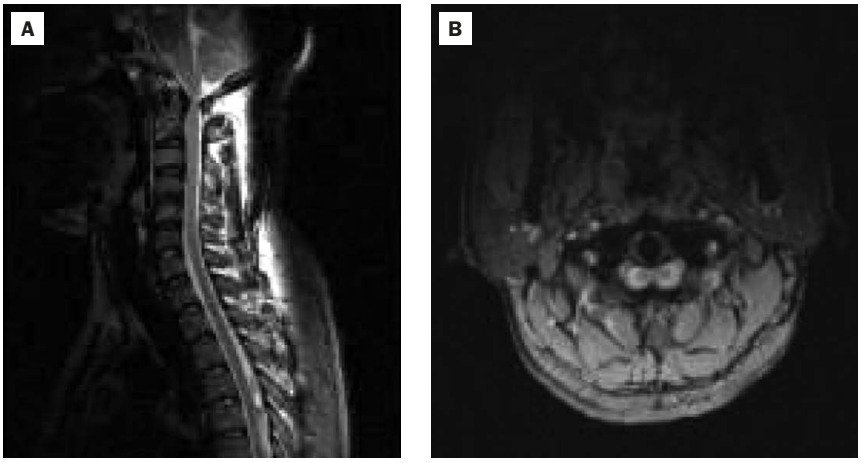

Figure 3. Cervical spinal magnetic resonance imaging reveals stenosis at the atlas level. The axial cut is at the C1 level. The sagittal canal diameter is 12.40 mm and the sagittal dural sac diameter is 6.23 mm at the C1 level. Note the high signal intensity change in the posterior aspect of the cord at the C1 level in the sagittal view, which may indicate compressive myelopathy.

underwent the operation improved to some degree. Interestingly, all of the 10 adult-onset patients were Asian, and both of the child-onset patients were Caucasian (Table 1).[1–9]

Congenital anomaly of the atlas may result from achondroplasia, aplasia or hypoplasia of the anterior or posterior arch, or it may result from cleft formation of the anterior or posterior arch.[10] Congenital anomalies

Table 1. Summary of patients with symptomatic isolated atlantal stenosis reported in the literature

Authors	Sex	Age (yr)	Ethnicity	Clinical presentation	Onset of symptoms to presentation or treatment	Canal diameter at atlas level	Dural sac diameter at atlas level	Treatment	Results
Wackenheim[2] (1974)	NM	NM	NM	Occipital neuralgia, functional obstruction of foramen magendie, no cervical myelopathy	NM	NM	NM	NM	NM
Sawada et al[3] (1989)	Male	38	Japan	Weakness of right upper and both lower limbs; muscular wasting of bilateral arms and forearms; lower limbs spasticity; DTR of all limbs increase; slight sensory disturbance of all limbs	3 yr	7 mm	NM	Removal of posterior arch of atlas	Weakness and spasticity of extremities alleviated
Komatsu et al[4] (1993)	Male	56	Japan	Progressive gait disturbance, spasticity of lower extremities; weakness and muscular atrophy of all limbs; muscle tone increase in all limbs; impairment of sensory functions of all extremities; lower limb hyperreflexia; difficult urination	11 yr	7.7 mm	NM	Removal of posterior arch of atlas, C2 laminoplasty and occipital decompression of foramen magnum	Able to walk with assistance; resolution of urinary incontinence; full recovery of bilateral upper limb muscle power
Tokiyoshi et al[1] (1994)	Male	55	Japan	Numbness in all limbs; left-sided hemihyperesthesia; spastic gait; bilateral DTR increase; bilateral Chaddock signs	5 mo	8 mm	NM	Decompressive suboccipital craniectomy, removal of posterior arch of atlas and duroplasty	Normalization of DTR, gait and sensory function
Okamoto et al[5] (1998)	Male	77	Japan	Paresthesia and paresis of all limbs; bilateral lower limb spasticity; all limb DTR increase; bilateral Hoffmann's sign and Babinski's sign were positive; JOA score was 4	20 yr	11 mm	NM	Removal of posterior arch of atlas and occipitocervical sublaminar wiring	JOA score improved to 5 points; improvement of gait disturbance and fine movements

(*Contd*)

Table 1. Continued

Authors	Sex	Age (yr)	Ethnicity	Clinical presentation	Onset of symptoms to presentation or treatment	Canal diameter at atlas level	Dural sac diameter at atlas level	Treatment	Results
Phan et al[6] (1998)	Male	80	Chinese	Sometimes urinary incontinence; mild atrophy of bilateral hypothenar muscles and interosseous muscles; increased muscle tone, hyperreflexia and impaired sensory function of all limbs; bilateral Hoffmann's sign positive; spastic gait	2.5 yr	8 mm	NM	Suboccipital decompression of foramen magnum; removal of posterior arch of atlas and C2 partial laminectomy	Some improvement in neurologic symptoms and stabilization of gait
	Male	75	Chinese	Bladder dysfunction; unable to walk without assistance; spasticity, hyperreflexia, muscular atrophy and muscle power decrease of all limbs; sensory impairment of bilateral C4 dermatones; bilateral Hoffmann's sign positive	6 mo	7 mm	NM	Removal of posterior arch of atlas	Able to walk without assistance; muscle power increase in all limbs; bladder function returned to normal
Nishigawa et al[7] (2001)	Male	82	Japanese	Spastic gait; hyperreflexia of all limbs; hypesthesia of bilateral C6 to T1 and L4 to S1 dermatones	17 mo	12 mm	7 mm	Removal of posterior arch of atlas and C2 laminoplasty	Near complete resolution
	Male	72	Japanese	Severe gait disturbance; loss of manual dexterity; hypesthesia of bilateral C4 to T1 and L4 to S1 dermatones; unable to walk	3 mo	11 mm in neutral and 8 mm in flexion	6 mm	Removal of posterior arch of atlas	Marked improvement of gait
	Female	22	Japanese	Neck pain; headache; vague right hemicorporal paresthesia; symptoms exacerbated by neck extension, rotation and ipsilateral tilting; no cervical myelopathy	NM	9 mm	NM	Removal of posterior arch of atlas	Most of the symptoms eliminated
Connor et al[8] (2001)	Female	8	Caucasian	Clumsy hands; ataxic gait; lower limb weakness; an episode of	1 yr	10 mm	NM	Removal of posterior arch of atlas	General improvement of walking and

				transient marked weakness of right leg with spontaneous recovery within 1 wk					clumsiness; resolution of gait ataxia
Tubbs et al[9] (2005)	Female	9	Caucasian	Episodes of sudden hypotonia with subsequent falling; this patient has underlying idiopathic growth hormone deficiency, Duane's syndrome, Klippel-Feil syndrome and developmental delay	A few months (exact time is not mentioned)	< 10 mm	NM	Removal of posterior arch of atlas and suboccipital craniectomy	Frequency of hypotonia episodes decreased
Present case (2007)	Male	38	Chinese	Tingling sensation in abdomen and perineum while flexing neck; bilateral hand numbness	2 mo	12.46 mm	6.23 mm	Removal of posterior arch of atlas with duroplasty	Resolution of hand numbness and abnormal tingling sensation in abdomen and perineum

NM = not mentioned; DTR = deep tendon reflex; JOA = Japanese Orthopedic Association (JOA score is the criterion for evaluation of the operative results of patients with cervical myelopathy according to the JOA).

of the atlas usually involve the posterior arch. These posterior arch defects of the atlas are classified into 5 variants: type A is failure of posterior midline fusion of the 2 hemiarches, which forms a midline cleft in the posterior arch; type B is unilateral arch defect; type C is bilateral arch defects; type D is absence of posterior arch with preservation of the posterior tubercle; and type E is absence of the posterior arch including tubercle. The type A defect occurs in 3–4% of the general population and comprises more than 90% of all posterior arch defects of the atlas.[11] The mentioned 12 patients and the presented patient in this article all had complete but hypoplastic posterior arch of the atlas, except the patient reported by Connor et al, whose posterior arch of the atlas had a type A cleft.[8]

During embryogenesis, the atlas originates from 3 ossification centers, 1 anterior and the other 2 on lateral sides. The anterior ossification center gives rise to the anterior tubercle and anterior arch. The 2 lateral ossification centers extend dorsally and fuse in midline to form the posterior arch. The arch is nearly fused by birth, and complete ossification between the 3 ossification centers usually occurs between 6 and 8 years of age.[12] Therefore, hypoplasia of the posterior arch of the atlas may result from premature fusion of the 2 lateral ossification centers or inadequate dorsal extension of the 2 lateral ossification centers at the time of union.[6,7]

The average sagittal dural sac diameter is 10–12 mm, and the normal sagittal canal diameter is 16–25 mm at the atlas level. In the presented case, the patient's sagittal dural sac diameter was 6.23 mm and the sagittal canal diameter was 12.40 mm at the atlas level. There is significant cord compression if the sagittal canal diameter is smaller than 14 mm at the atlas level, and myelopathic symptoms and signs will develop if the sagittal canal diameter is smaller than 10 mm at the atlas level.[1,3–7,9,13]

Most of the atlantal configurations of the reported cases had hypoplastic but complete and smooth-curved posterior arches, or included the presence of a midline cleft of the posterior arch. The atlas shape of the patient we present had the unique feature of a hypoplastic, complete but incurved posterior arch (Figure 4).

Nine of the 12 reported cases presented cervical myelopathy symptoms during adulthood. Although atlantal stenosis is congenital, the hypoplastic atlas itself may not result in neurologic manifestation unless complicated by aging processes such as degenerative spondylosis. Degenerative cervical spondylosis usually occurs below the level of C2, which decreases the range of motion of the lower cervical spine, thus increasing atlantal movement. Atlantal overactivity may contribute

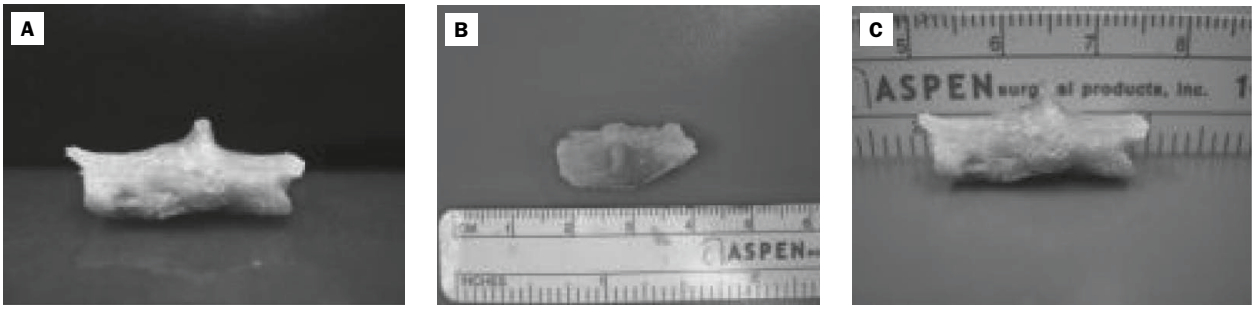

Figure 4. The removed posterior arch of the atlas. Note the ridge formation on the midline, which had caused significant cord compression.

to spinal cord compression and the development of symptoms.[4,6]

In addition to causing neurologic symptoms, severe atlantal stenosis may predispose patients to severe spinal cord injury after a trivial trauma. Liliang et al reported a 3-year-old boy who suffered from quadriplegia and respiratory failure after performing an improper somersault. Neuroimaging of this patient revealed cervical spinal cord contusion with marked canal stenosis at the atlas level.[10]

Treatment is surgical decompression and, in the majority of cases, removal of the posterior arch of the atlas is enough.[7] The case we have presented here and the 11 review cases who received surgical treatment demonstrated significant improvement after operation.

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