

CONGENITAL ATLANTO-AXIAL DISLOCATION

C. T. TAN

T.G. LOH

P. BALASUBRAMANIAM

INTRODUCTION

Atlanto-axial dislocation is an uncommon condition. It is usually secondary to trauma, infection (tuberculous and non-tuberculous atlas and axis) and rheumatoid arthritis. Other uncommon causes reported include retropharyngeal abscess, malignant deposits, ankylosing spondylitis (Sorin, *et al.*, 1979). Congenital atlanto-axial dislocation is rare. Wadia (1967) first drew attention to its relative common occurrence among his patients in India. This finding has since then been substantiated by other authors (Bharucha *et al.*, 1964). Congenital atlanto-axial dislocation is also said to be relatively common in Ceylon and Thailand (Wadia, 1972). Rheumatoid arthritis is not as commonly found in Malaysia as in the western developing countries. Its pattern of illness seen here also tends to be of milder form of illness (Toh, *et al.*, 1973). Ethnically Indians constitute about ten percent of the multiracial population of Malaysia. It is thus interesting to note that since the inception of the University Hospital in 1968, we have seen three cases of congenital atlanto-axial

dislocation, two of them were ethnic Indians. Two other atlanto-axial dislocations were also seen during the same period; both of them were of traumatic origin. We are reporting here the three cases of congenital atlanto-axial dislocation to bring awareness to this potentially curable form of illness.

CASE 1

C.R. was a 36 year old Indian male seen in 1977. His symptom first started when he was 21. He developed sudden onset of vertigo while sitting on a bicycle. This was followed by generalised weakness so that he was subsequently admitted to the local estate hospital and was confined to bed for four days. He was discharged well after a month and was able to go back to work as a tin mine labourer.

A year later, he was admitted to Kuala Lumpur General Hospital for seven months from two attacks of vertigo which left him tetraplegic with difficulty in talking and swallowing. He apparently made very slow recovery from these disabilities, eventually went back to work in packaging in a light bulb factory eleven years later.

The admission in 1977 was due to an acute onset of giddiness associated with bifrontal throbbing headache and acute onset of left limbs weakness. Besides this, he also complained of dizziness on extension of the neck. There was no other history of note.

On physical examination, the patient was noted to be of below average intelligence. He had short neck with Sprengel's shoulder on the right side. The neck movement was limited. The speech was

C.T. Tan, M.B., B.S. (Melb.), M.R.C.P. (U.K.)

T.G. Loh, M.B., B.S. (Sing.), M.R.C.P.

P. Balasubramaniam, M.B., B.S. (Cey.), S.R.C.S. (England).

Departments of Medicine and Orthopaedic Surgery,
Faculty of Medicine, University of Malaya,
Kuala Lumpur, MALAYSIA.

Correspondence to:

Dr. C.T. Tan, Department of Medicine,
Faculty of Medicine, University of Malaya,
Kuala Lumpur, MALAYSIA.



Fig. 1 Cervical spine x-ray (case 1) showing the os odontoidenum with the body of the axis being displaced anteriorly. The distal odontoid fragment is shown with arrow.

slightly dysarthric with nasal escape. The cranial nerves were normal except the gag reflex which was tardive. The power was weak on all four limbs with brisk reflexes and extensor plantar responses both sides. Sensory examination was normal and the gait was spastic. Examination of the other systems were normal. The x-ray of cervical spine with tomography demonstrated partial fusion of the C2-C3 vertebrae and the body of the axis was displaced anteriorly (Fig.1). A-P view showed that the odontoid fragment was well corticated and was displaced high in the foramen magnum. Other investigations which were normal included the full blood count, serum creatinine, electrolyte, calcium and blood glucose. The patient was referred to the Neurosurgical Unit in the General Hospital, Kuala Lumpur for fusion of the atlantoaxial joint. He improved functionally after operation and was able to go back to work.

CASE 2

C.A.M. was a 62 year old Chinese housewife. She was admitted in 1979 with the complaint of neck pain of two years' duration, six months of stiffness and weakness in all four limbs starting first on the left side. Because of these, she was unable to carry out her household chores and had to walk slowly. Besides feeling the sensation of spasm of the arms everytime she bent her neck, there was no other significant symptom. There was no history of trauma, and the past history was not significant.



Fig. 2 Cervical spine x-ray (case 2) showing os odontoidenum with forward dislocation of atlas over the body of the axis.

On examination, she was thin and a little wasted. The cranial nerves examination was normal. Neck movements were limited. There was spastic weakness of all four limbs with generalised hyperreflexia and extensor plantar responses. The proprioceptive sensation was impaired over all four limbs, the left upper limb showed reduced sensation to pin prick. Examinations of the other systems were normal.

Investigations which were normal included full blood count, ESR, serum electrolyte, blood glucose, liver function test, serum creatinine, calcium, phosphate, alkaline phosphatase, ECG and chest x-ray. X-ray of the cervical spine with tomography showed forward dislocation of the atlanto-axial joint with the os odontoidenum (Fig. 2).

The patient then had skull traction to reduce the fracture followed by posterior C1C2 wiring. Unfortunately she developed respiratory complications and died two weeks post-operation.

CASE 3

G.M. was a 28 year old Indian who worked as a blacksmith in the Malayan Railway. His main complaint was that since four years before admission in 1980 he had been having progressive unsteadiness and weakness of both legs with paraesthesia of all four limbs. Over the last one year, he had noticed pain in the neck which radiate to the left arm, mainly occurring in the morning and aggravated by neck movement. There was no sphincteric disturbance and no impotence. Despite these symptoms he was able to cope with his job. When he was 18, the patient had a fall from a ten feet height during which he had loss of consciousness lasting about two hours. He did not develop any neurological symptom after recovering consciousness. There was no other history of note.

On physical examination, the patient had a short neck with limited neck movement. Cranial nerve examination was normal. The right trapezius and small muscles of the right hand was wasted. The muscle power of both upper limbs were normal. Both hip flexors, knee extensors and dorsi flexors of the ankle were mildly weak. All the reflexes were brisk with bilateral extensor plantar responses. Sensory examination showed proprioceptive sensation loss in left big toe. The gait was spastic. Examination of the cardiovascular, respiratory systems and the abdomen were normal. Other investigations which were normal included full blood count, ESR, blood urea, serum electrolyte, calcium, phosphate, VDRL, TPHA, ECG, skull x-ray and chest x-ray. Cervical spine x-ray with flexion, extension, tomography and screening showed considerable subluxation between C1 and C2 joint.

The patient was put on skull traction for five weeks to reduce the fracture. While on traction, he developed urinary retention. Occipito-cervical spine fusion was subsequently done. The bladder symptom recovered post-operatively with the neurological sign remaining the same.

DISCUSSION

The first case of congenital atlanto-axial dislocation was described in 1886 by Giacomini. It was definitely rare as Greenberg (1968) was only able to find 94 cases reported in the literature. Wadia (1960) pointed to its unusual high frequency

in India. In his presentation of 52 cases in 1972, there was a male predominance of 4:1. The average age of the onset of illness was 24 years. The duration of symptom ranged from weeks to 48 years.

The pathogenesis of atlanto-axial dislocation causing spinal cord compression is related to the occipitalization of the atlas and the fusion of the C2-C3 vertebrae (Klippel-Feil syndrome) in some patients. These anomalies impose undue stress on the atlanto-axial joint. Abnormal adontoid process which may be too long, too short or failure of fusion between the apical and basal segment (os odontoidenum) are important in other cases. Particularly in patients with mongolism, there may be excessive laxity of the transverse ligament reducing the stability of the joint.

The symptomatology and signs are best exemplified by Wadia's case review (1972). The neurological symptoms consisted of three groups: cervical pain and stiffness, transitory attacks and progressive neurological disturbance. The neck stiffness with limitation of movement occurred in 50 percent of his cases. Transitory attacks were often precipitated by exaggerated flexion and extension of the neck. The attack consisted of paralysis of limbs, parasthesia below the neck, unconsciousness, blurring of vision and blindness.

The neurological sign may be slowly progressive, or persistent after the transitory attack. The most common finding was a spastic tetraparesis. Localised muscle wasting of the hand, shoulder muscles and occasionally fasciculation may be seen. The mechanism may be due to the venous congestion and stagnant hypoxia as shown experimentally in monkey when plastic tumor was inserted at high cervical spine (Taylor and Brynes, 1974). Posterior column sign was less evident and urinary symptom was uncommon. Horner's syndrome and nystagmus were rarely seen. Other associated congenital anomalies such as short neck (as in cases 1 and 3), dysplastic face, kyphoscoliosis, Sprengel's deformity - congenital elevating scapula (as in case 1) may occur.

The diagnosis rests almost entirely on good radiological examination. Lateral view of the cervical spine in flexion and extension and the open mouth view to visualize the atlanto-axial joints is usually required. Lateral cervical tomography is necessary in some cases. Myelography is indicated

when the diagnosis is uncertain; when there are more than one congenital anomalies exist in the cervical spine so that one has to be certain the site of compression is at the atlanto-axial joint; and when differentiation from syringomyelia or basilar invagination has to be made.

It is well known that traumatic os odontoidenum may also cause delayed myelopathy (Fielding and Griffin, 1974; Hawkins *et al.*, 1976). In the traumatic non-union, history of trauma is usually present, the fragments tend to match perfectly and there is no marginal cortex at the level of the fracture line or the rounded off appearance that is found with congenital os odontoidenum (Hensing and McEwan, 1975). Absence of other associated congenital anomaly is also a guide. We favour a congenital aetiology to our second patient's os odontoidenum as there was no past history of trauma. In addition, the odontoid fragment showed smooth outline and was well corticated (Fig. 2).

The other associated congenital craniovertebral anomalies which may be seen with radiological examination include hemivertebra, small hypoplastic dens, cervical spinal bifida and cervical rib. Computerized tomography has been shown to be useful in the diagnosis of atlanto-axial dislocation (Dan, 1978).

Our third patient had a history of head injury six years prior to his first development of symptom. Unlike os odontoidenum, there has not been any previous report of traumatic transverse ligament incompetence causing a delayed myelopathy from atlanto-axial subluxation although Greenberg (1968) has suggested that it may be an aetiological factor in some patients. However, the associated short neck in our patient suggests that congenital anomaly was the main cause for his atlanto-axial subluxation and the head injury could definitely have precipitated his myelopathy.

Management of the patients involves firstly reduction of the dislocation by neck extension. If this is not possible, skull traction is recommended. Fusion of the atlanto-axial joint is then carried out.

ACKNOWLEDGEMENTS

We want to express our thanks to Dr. Mohan Das who operated on case 1, Dr. K.J. Lim for referring and Dr. R. Rapport for operating on case 2, all the staff of the Department of Medicine, Surgery and

Orthopaedic Surgery who have taken part in the care of the patients. Thanks also to Puan Rohani for typing the manuscript.

REFERENCES

- Bharucha, E.P. and Dastur, H.M. (1964) Craniovertebral anomalies, *Brain*, **87**, 469-481.
- Dan, N.G. (1978) Atlanto-axial subluxation: An unusual case elucidated by computerized tomography, *Aust. N.Z. J. Surg.*, **48**, 565-566.
- Fielding, J.W., Griffin, P.P (1974) Os odontoidenum: an acquired lesion. *The Journal of Bone and Joint Surgery*, **56-A**, 187-190.
- Giacomini, C. (1886) as quoted in Greenberg, A.D. (1968) Atlanto-axial dislocations, *Brain*, **91**, 655-685.
- Greenberg, A.D. (1968) Atlanto-axial dislocation, *Brain*, **91**, 655-685.
- Hawkins, R.J., Fielding, J.W. and Thompson, W.J. (1976) Os odontoidenum - congenital or acquired: a case report. *J. Bone and Joint Surgery*, **58A**, 413-414.
- Hensing, R.N., McEwan (1975) Congenital anomalies of the spine. *The Spine* Vol. 1, Ed. R.H. Rothman and F.A. Simeone, Publishers: W.B. Saunders co.
- Singh, S., Dutta, A.K. and Gupta, S. (1969) Cranio-vertebral anomalies with neurological deficit, *J. Assoc. Physician Indo.*, **17**, 469.
- Sorin, S., Askari, A., Maskowitz, R.W. (1979) Atlanto-axial subluxation as a complication of early Ankylosing Spondylitis - two case reports and review of literature, *Arthritis and Rheumatism*, **22**, 273-276.
- Taylor, A.R. and Brynes, D.P. (1974) Foramen Magnum and High cervical cord compression, *Brain*, **97**, 473-480.
- Toh, B.H., Sengupta, S., Ang, A.H., White, J.C., Lau, K.S. (1973) Rheumatoid Arthritis in West Malaysia, *Ann. Rheum. Disease*, **32**(2), 151-156.
- Wadia, N.H. (1960) Chronic progressive myelopathy complicating atlanto-axial dislocation due to congenital abnormality. *Neurology (Bombay)* Vol. VIII. Pg. 81.
- Wadia, N.H. (1967) Myelopathy complicating congenital atlanto-axial dislocation, *Brain*, **90**, 449-473.
- Wadia, N.H. (1972) Congenital atlanto-axial dislocation and its manifestations due to spinal cord compression in the Tropical Neurology, Ed. J.D. Spillaine, Publishers: Oxford University Press.