Spontaneous Atlanto-Axial Dislocation in a Mongoloid Child with Spinal Cord Compression

Case Report

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Stability of the atlanto-axial junction depends on the elasticity and length of the craniovertebral ligaments that govern mobility about the upper cervical spine,4,17,27 the attitude of the articulating facets9 and the integrity of the odontoid process.16 Since the original description of spontaneous atlanto-axial dislocation by Bell (1830),5 this entity has been reported following infections about the nasopharynx or neck in children,1,4,17,19,22,25,27 with rheumatoid arthritis, ankylosing spondylitis,4,11,12,16,18,19,20 poliomyelitis4 and implicated in tuberculosis, syphilis,27 and steroid therapy.12

According to the recent observations of Tishler and Martel,24 and Spitzer et al.,22 nontraumatic or spontaneous atlanto-axial dislocation is a significant common occurrence in mongolism, and may be a reflection of the congenital laxity of ligaments and joints present in these patients. However, no neurological sequelae have been observed in these individuals to date.15,16,22

This is a report of a mongoloid child with spontaneous atlanto-axial subluxation causing spinal cord compression.

Case Report

A 13-year-old girl was admitted to the Riley Hospital for Children on July 23, 1965, with clinical features of mongolism, neck pain and weak extremities. She had been a happy, active child until an ear infection 5 months ago for which she was treated with antibiotics. Although the patient had not been acutely ill, she had become less active, lost weight, and kept her head immobile and tilted to the left. She was constipated, voided infrequently, and was occasionally incontinent.

The patient, born at full term, was the 3rd pregnancy of a 35-year-old mother. The diagnosis of mongolism was ascertained when the patient was a few months old. Four older siblings were all in good health. The patient had had tonsillectomy and adenoectomy when she was 10 years old. Prior to her present illness she had been attending a special school and could read and write.

Examination. At time of admission the patient was a thin, poorly-developed child with the facial and skeletal features, but without the active joviality, of a mongoloid. Her pulse, blood pressure and respirations were normal. She kept her neck flexed and head tilted to the left, resisted active or passive movements and complained of pain. She required support to stand and walked with a broad base. Her grip was weak, she was unable to undress herself and could not raise either arm above the head. Deep tendon reflexes were increased in all extremities, and bilateral toe extensor signs were present. Sensation to pinprick and light touch was present, but vibration seemed impaired in all extremities. There was no papilledema, or limitation of external eye movements, and the visual fields were full by confrontation.

Coarse horizontal nystagmus was present on lateral gaze to either side. The bladder was visibly distended, but caused no apparent discomfort to the patient. She voided 250 cc. of urine with straining, and an additional 135 cc. were obtained by catheterization. Hemogram, urinalysis and blood urea nitrogen were normal.

Roentgenograms revealed an atlanto-axial dislocation (Fig. 1) and scoliosis of the cervical spine. Brain scan (Hg 208) was negative and electroencephalogram minimal, but diffusely abnormal. Cystometrogram con-

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firmed the diagnosis of a neurogenic bladder and an indwelling catheter drainage was maintained. Attempted pneumoencephalography failed to fill the ventricular system.

Skeletal traction was applied to the skull by passing wire loops between 2 Burr holes on either side of the midline in a sagittal direction. With 12 pounds traction, improvement in muscle strength in the extremities was noted, and reduction of the atlanto-axial dislocation was radiographically evident within 24 hours.

**Operation.** After several days of treatment with 5 lbs. of traction, a posterior fusion of the upper 3 cervical vertebrae was performed. At surgery, with continued traction, four 20-gauge wire loops were threaded extradurally beneath the laminae of the upper 3 cervical vertebrae and secured over the bone struts placed on the laminae. An additional loop was passed under the posterior arch of the atlas and around the 2nd and 3rd spinous processes (Fig. 2).

**Postoperative Course.** The patient was kept in traction until the sutures were removed and then fitted with a Minerva cast. With physical therapy, she regained the ability to feed herself and to walk with assistance. When she was discharged on September 12, the indwelling catheter was removed, but because of residual urine volume of 35 to 75 cc. the patient was given decreasing doses of oral cholinergic drugs for 3 weeks.

When she returned for removal of the cast 12 weeks later, the patient walked normally and was able to completely empty her bladder. There was no deviation of the head, and although hyperreflexia persisted, the Babinski sign was no longer present.

Six months after the operation, stress films of the cervical spine disclosed a solid fusion of the upper 3 cervical vertebrae, and the atlanto-odontoid interval measured 2 to 3 mm.

**Discussion**

Neck pain and rigid, tilted head position are common clinical signs of symptomatic atlantoaxial dislocation.2,4,7,11,25-27 Signs of spinal cord compression are unusual,4 and particularly rare in children.12 Martel15 found no neurologic manifestations in 70 mongoloids studied, 14 of whom had atlas dislocations.15 In the present case, the dislocation may have occurred as a sequel to the ear infection, or may have been previously asymptomatic, and become clinically manifest concomitantly with, but not due to, the illness. As no previous cervical spine films were available, either explanation appears equally plausible.

The ultimate diagnosis of atlanto-axial dislocation is made on radiographic findings of the cervical spine.4 The degree of dislocation is reflected by the atlanto-odontoid interval, which in adults normally should not exceed 2 mm.,4,5 but may be 3 to 4.5 mm. in children.18,20,24 Since intrinsic lesions of the brain stem and parasagittal areas can mimic symptomatic atlanto-axial dislocations, further studies (pneumoencephalogram, electrencephalogram and brain scan) were performed to exclude this possibility in our case. The air study was presumably unsuccessful because of a narrowed entrance to the 4th ventricle, and extensive manipulation and neck flexion for optimum position was thought hazardous.

Although symptomatic relief, and even objective improvement, can be obtained with a plaster jacket or neckbrace immobilization,4,7,10,23,27 in progressive or recurrent neurological manifestations, reduction and fixation of the unstable segment of the cervical spine is the treatment of choice.2,14,20,26 In our case, the method of anchoring traction by passing wire loops between Burr holes was preferred because the skull seemed too thin to withstand long traction. Although traction promptly reduced the dislocation, tightening the wire loops about the upper cervical laminae at surgery further decreased the atlanto-odontoid interval by forcing the odontoid anteriorly toward the floor of the spinal canal.

**Summary**

Spontaneous atlanto-axial dislocation has been recognized as a common occurrence in mongolism, but associated neurological manifestations have not been reported. We have reported a case in which a 13-year-old mongoloid girl with progressive quadripareis and a neurogenic bladder, caused by spontaneous atlanto-axial dislocation,