Spontaneous rotatory atlantoaxial dislocation without neurological compromise in a child with Down syndrome: a case report

Iain Au-Yong · Bronek Boszczyk · Hossein Mehdian · Robert Kerslake

Received: 27 March 2007 / Revised: 5 December 2007 / Accepted: 24 December 2007 / Published online: 31 January 2008
© Springer-Verlag 2008

Abstract Spontaneous atlantoaxial dislocation is a rare recognised complication of Down syndrome. In the majority of cases, dislocation takes place in an anteroposterior direction and is often associated with abnormalities of odontoid development or ossification. Rotatory atlantoaxial dislocation is extremely rare in Down syndrome and this is to our knowledge the first reported case in which modern imaging methods have been described; surface shaded reformats derived from a multislice CT scan were of fundamental importance in making the diagnosis.

Keywords Spine trauma · Atlantoaxial dislocation · Rotatory · CT · Down syndrome

Background

Spontaneous atlantoaxial dislocation is an uncommon complication of Down syndrome [2] and is much rarer than atlantoaxial subluxation which is a common feature of

Down syndrome not thought to predispose to dislocation [4]. Dislocation is almost invariably in an anteroposterior direction and neurological deficit is usual [2]. The current case describes an unusual case of rotatory atlantoaxial dislocation, without neurological deficit, in which the use of surface shaded CT images was immensely helpful in establishing the diagnosis.

Case report

A 5-year-old boy, with Down syndrome presented having awoken with a stiff neck on the day of presentation. There was no apparent history of trauma or recent otolaryngeal inflammation. He had a background of attention deficit hyperactivity disorder and glue ear. Examination revealed that he was holding his neck laterally flexed to the right and rotated to the left. Neurological examination was unremarkable. Any attempt of gentle manual manipulation or traction was resisted by the child.

It was not initially thought that radiographs would be helpful owing to the child's position. CT of the cervical spine was therefore performed (see “[Materials and methods](#)” and figures for detailed discussion of the findings) and a diagnosis of atlantoaxial rotatory dislocation was made.

The patient was initially treated by Halo traction, which was not successful due to lack of patient understanding and cooperation. He was subsequently placed in halo traction under general anaesthesia and fitted with a halo-vest for 3 weeks. Subsequent radiographs confirmed that normal alignment had been restored at C1/C2. Clinical review 2 months after initial presentation revealed a full range of craniocervical motion. A lateral plain film of the cervical spine and further clinical review were normal at 10 months (Fig. 1).

Electronic supplementary material The online version of this article (doi:10.1007/s00586-008-0596-y) contains supplementary material, which is available to authorized users.

I. Au-Yong · R. Kerslake (✉)
Department of Radiology, Queen's Medical Centre Campus,
Nottingham University Hospitals, Nottingham,
Nottinghamshire NG7 2UH, UK
e-mail: robert.kerslake@nott.ac.uk

B. Boszczyk · H. Mehdian
Department of Spinal Surgery, Queen's Medical Centre Campus,
Nottingham University Hospitals, Nottingham,
Nottinghamshire NG7 2UH, UK

Fig. 1 Lateral radiograph of the cervical spine obtained 10 months after initial presentation demonstrating normal craniocervical alignment

Materials and methods

Computed tomography CT was performed under oral sedation using a 16 slice multidetector CT scanner (Philips MX8000 IDT).

Overlapping 1 mm sections of the craniocervical junction were obtained from which multiplanar (MPR) and surface shaded (SSR) reformats were derived using third party proprietary software (Voxar, Barco NV, Kortrijk, Belgium) running on a standard personal computer-based Picture Archiving and Communication System (PACS) workstation. Interpretation of the conventional axial and multiplanar reformatted images was challenging (Fig. 2a, b). However, surface shaded reformats rapidly revealed that there had been complete forward dislocation of the right C1 articular mass in relation to C2 with consequent rotation of about 45° and lateral inclination of the head and C1 in respect to C2 (Fig. 3; Supplementary movie). The anterior

Fig. 2 **a** Axial CT shows rotation between C2 (*a*) and the mandible (*b*). The dislocation at C1/C2 could not be demonstrated on single axial images. **b** Sagittal reformatted CT images shows a hypoplastic odontoid process (*a*). Normal atlanto-dental alignment is maintained

and posterior arches of C1 were noted to be bifid and the odontoid process was hypoplastic but remained central in relation to the C1 anterior arch (Fig. 4a, b); the transverse ligament was therefore presumed to have remained intact. The foramen magnum and upper cervical canal were not compromised, accounting for the lack of neurological deficit.

Discussion

Acute torticollis is an important presenting symptom which can be associated with serious pathologies such as atlantoaxial dislocation [7] and atlantoaxial rotatory subluxation.

Atlantoaxial dislocation occurs rarely in Down syndrome and is rarer than rotatory subluxation and atlantoaxial subluxation, neither of which are thought to predispose to dislocation. A summary of 38 published case reports [2] showed that trauma was the cause in only six, other precipitating causes including upper respiratory tract infections. In many of these cases, as in our patient, there was no discernible precipitating cause.

Fig. 3 A surface shaded image of the occipito-cervical junction viewed from below shows complete rotatory dislocation of the lateral atlantoaxial joints (*a*). The lack of vertebral canal compromise and bifid anterior arch of C1 can also be appreciated

The vast majority of Downs' patients presenting with anteroposterior atlantoaxial dislocation have an associated neurological deficit due to cord compression [5]. Underlying congenital anomalies of C1 and C2 are common; in one series 12 out of 21 had congenital abnormalities such as ossiculum terminale, os odontoideum or hypoplasia of the odontoid process [6], predisposing the odontoid process to slip beneath the transverse ligament.

The current case is unusual in that the dislocation was rotatory rather than anteroposterior, as described above. Because the odontoid remained unchanged in position with regard to the anterior arch of the atlas and there was no canal compromise, neurological sequelae were avoided. Rotatory atlantoaxial dislocation is recognised but extremely rare in patients with Down syndrome [10].

The recommendations for management of atlantoaxial instability in Down syndrome focus on the incompetence of the transverse atlantal ligament. As there are no conservative means of improving or restoring the biomechanical function of the ligament, significant translatory C1/C2 instability is addressed surgically by posterior fusion [8]. The condition in the presented case with isolated rotatory dislocation however, did not indicate biomechanical compromise of the transverse atlantal ligament. It was therefore assumed that repositioning through axial traction might be possible if performed without significant delay. This was achieved under general anaesthesia and maintained after removal of halo fixation. Subsequent radiographic examination ruled out translatory instability indicating maintained transverse atlantal competence.

The diagnosis, which would have been difficult to make by other means, was reached readily using surface shaded CT reformats and is the first reported case of such an

Fig. 4 **a** Surface shaded image of the occipito-cervical junction viewed from anteriorly shows the bifid C1 anterior arch (*a*) and rotatory dislocation at C1/C2. **b** Posterior surface shaded image of the occipito-cervical junction shows the bifid C1 posterior arch (*a*)

unusual presentation described in the literature. The ability in this case to obtain an overview of the relevant anatomical area from multiple viewing angles was of fundamental importance in understanding the nature of the deformity. In the majority of previous case reports, conventional radiographs were relied on for the diagnosis of atlantoaxial dislocation [9]; this approach would probably have failed in this instance where the atlantoaxial distance remained normal. Conventional sagittal and coronal CT reformats,

which have been utilised in previous reports [1], were difficult to interpret as the relevant anatomical areas were displayed on adjacent sections or different viewing planes because of the rotatory nature of the deformity. MRI was also rejected as an initial imaging modality, as the risk of a general anaesthetic in an uncooperative child was considered to be higher than that of the radiation dose. MRI would also have been inferior in demonstrating bony relationships.

A study analysing the application of reformatted CT images in a series of patients presenting with atlantoaxial subluxation showed that surface shaded reformatted 3D images were superior to other reconstructions in demonstration of this injury [3]. Their findings, although in a slightly different group of patients, illustrate the poor sensitivity and specificity of plain film diagnosis and would support the use of CT scanning with surface shaded reformats in patients presenting with acute torticollis.

Rotatory C1/C2 dislocation in Down syndrome should be therefore be distinguished from the more common translatory C1/C2 instability and may be treated conservatively if the transverse atlantal ligament is competent, as was inferred in this case from the position of the dens relative to the anterior arch of the atlas. This CT technique should be considered as a first line of investigation in other children in whom disorders of the craniocervical region are suspected.

Conflict of interest statement None of the authors has any potential conflict of interest.

References

1. Aoki N (1988) Atlantoaxial dislocation presenting as sudden onset of quadriplegia in Down's syndrome. *Surg Neurol* 30:153–155
2. Davidson RG (1988) Atlantoaxial instability in individuals with Down syndrome: a fresh look at the evidence. *Pediatrics* 81:857–865
3. Duan SY, Lin QC, Pang RL (2004) Application of CT 3D reconstruction in diagnosing atlantoaxial subluxation. *Chin J Traumatol* 7:118–121
4. Elliott S, Morton RE, Whitelaw RA (1988) Atlantoaxial instability and abnormalities of the odontoid in Down's syndrome. *Arch Dis Child* 63:1484–1489
5. Hungerford GD, Akkaraju V, Rawe SE, Young GF (1981) Atlanto-occipital and atlanto-axial dislocations with spinal cord compression in Down's syndrome: a case report and review of the literature. *Br J Radiol* 54:758–761
6. Kobori M, Takahashi H, Mikawa Y (1986) Atlanto-axial dislocation in Down's syndrome. Report of two cases requiring surgical correction. *Spine* 11:195–200
7. McGuire KJ, Silber J, Flynn JM, Levine M, Dormans JP (2002) Torticollis in children: can dynamic computed tomography help determine severity and treatment. *J Pediatr Orthop* 22:766–770
8. Poeschel SM, Herndon JH, Gelch MM, Senft KE, Scola FH, Goldberg MJ (1984) Symptomatic atlantoaxial subluxation in persons with Down syndrome. *J Pediatr Orthop* 4:682–688
9. Saad KF (1995) A lethal case of atlantoaxial dislocation in a 56-year-old woman with Down's syndrome. *J Intellect Disabil Res* 39(Pt 5):447–449
10. Sherk HH, Nicholson JT (1969) Rotatory atlanto-axial dislocation associated with ossiculum terminale and mongolism. A case report. *J Bone Joint Surg Am* 51:957–964