Rotatory atlanto-axial dislocation in an infant with osteogenesis imperfecta

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Abstract This is a case report of a 15-month-old patient with osteogenesis imperfecta (OI) who sustained atlanto-axial dislocation. Our objective is to report a unique case of traumatic atlanto-axial subluxation in a child with osteogenesis imperfecta associated with bilateral femoral fractures. The management is discussed. Atlanto-axial dislocation occurring with associated osteogenesis imperfecta is very rare. There have been no previous reported cases. A 15-month-old girl with osteogenesis imperfecta sustained a traumatic atlanto-axial dislocation. The child was followed-up through presentation, diagnosis, management and post-discharge. The initial diagnosis was confirmed with a CT scan. The patient was treated conservatively with a halo-traction for 4 weeks followed by a halo jacket for a further 4 weeks. Both appliances were fitted under general anaesthetic. An anatomical reduction was achieved. There was no neurological deficit at any stage. The child has had a successful outcome. She is asymptomatic with a full range of movement at the atlanto-axial joints at 9 months. In conclusion, this paper records our management of this rare problem.

Keywords Atlanto-axial dislocation · Osteogenesis imperfecta · Bilateral femoral fractures · Halo-traction · Conservative treatment

Case report

A 15-month-old girl with known OI (Type III) presented to the John Radcliffe Accident and Emergency Department in May 2007. She had fallen accidentally down stairs while crawling at home sustaining leg injuries and had knocked her head. The exact mechanism of injury is not known. The child was initially diagnosed with bilateral femoral fractures and was treated in Gallow's traction. Once admitted onto a paediatric ward it became apparent that there was 60° of fixed rotation to the left. The patient was neurologically intact. CT scan demonstrated fixed atlanto-axial rotation.

A 1 kg halter traction was applied increasing to 2 kg. Over 72 h, this failed to reduce the dislocation even with the application of rotation. A halo was applied with eight pins. The skull was very soft as expected and the pins were applied using finger pressure as recommended in the manufacturer's instructions. A 2 kg weight was applied longitudinally and 0.1 kg weight was applied rotationally. This resulted in reduction of the dislocation. After 48 h the rotational traction was removed and the child remained in longitudinal traction for 4 weeks. A halo jacket was applied 4 weeks after the original injury once the femoral fractures had healed and the Gallow's traction was removed, and post-operative films again confirmed the adequate alignment and reduction. The child remained in the halo jacket for a further 4 weeks when it was removed under general anaesthetic (see Fig. 1).

She is now pain free and has regained full movements of the cervical spine at 9 months from the injury.

Discussion

Osteogenesis imperfecta is caused by dominant genetic mutations in the genes COL1A1 AND COL1A2. These

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Fig. 1 A 15-month-old girl with a halo jacket applied under general anaesthetic

code for collagen 1 found in bone, ligaments, tendons and skin. Type III OI is the severe form which is associated with short stature and major deformity as well as multiple fractures.

Traumatic atlanto-axial instability is an uncommon injury, and most commonly occurs in children. Stability of the atlanto-axial joint is primarily dependent on various ligamentous structures including the transverse ligament, the facet joint capsules and the tectorial membrane. The ligamentous laxity in children renders the joint susceptible to rotatory instability. Also children have shallower and more horizontal facet joints allowing greater freedom of motion increasing the force on the atlanto-axial complex [13]. A relatively large head and small neck increases load on the cervical spine during a traumatic event [8]. The mechanism of injury is often secondary to minor trauma but it can occur spontaneously, most often in Down's syndrome, Grisel syndrome, os odontoideum, juvenile rheumatoid arthritis and following neck surgery [1, 4, 12]. Although the majority of patients remain neurologically intact it is important to perform a thorough examination [6]. A previous study reviewing management of atlanto-axial subluxations highlighted a mean age of 6.4 years, with a range of 3–11 years [9]. A total of 15 months falls well below the normal range of presentation.

We have not found a similar case reported in the literature. The patient has OI (III) which made the management

of the problem more difficult. First there were bilateral femoral fractures which required management and second the presence of OI makes the placement and management of the halo more difficult because of the marked osteopenia. OI not only causes osteopaenia but also results in weakening of the ligaments, and this probably predisposed the patient to dislocation at the atlanto-axial joint.

Many patients with osteogenesis imperfecta have associated occipito-cervical pathology [7]. Although in this case, there was no evidence of other significant malformations.

Failure to reduce the fracture required halo traction allowing reduction of the dislocation. The options for management after reduction were to leave the neck free, to immobilise the neck in a collar (not easy in a 15-month-old child) or to treat in a halo jacket once the femoral fractures were healed and the traction removed. We elected the third option because we believed that it would give the best chance of stabilisation of the upper neck with a reduced chance of recurrence. To date (40 weeks after the dislocation) this has not occurred.

Most cases of rotatory subluxation will reduce with traction, dislocation may be more difficult to reduce. The earlier treatment is commenced, the more likely closed reduction is achieved. Where closed reduction is impossible or recurrence occurs, open reduction may be indicated usually with internal fixation between C1 and C2. A posterior approach is used with either screw or wire fixation [10].

It is often difficult to confirm the diagnosis of atlanto-axial subluxation with plain cervical spine radiographs [5], and even in this case with dislocation diagnosis was difficult on the plain X-rays (Fig. 2). Computerised tomography with

Fig. 2 Lateral cervical spine and oblique skull radiograph. Atlanto-axial subluxation is not evident highlighting the limitations of a plain film

Fig. 3 Posterior and anterior CT reconstruction of the cervical spine showing rotatory subluxation of the atlanto-axial joint

Fig. 4 Coronal CT of the cervical spine showing rotatory subluxation of the atlanto-axial joint

coronal and sagittal reconstructions provide more information. However the abnormal head and neck positions make the images difficult to interpret. Dynamic computerised tomography is used to confirm the diagnosis [2]. Atlanto-axial subluxation/dislocation can be classified into five categories: bilateral anterior or posterior translatory, unilateral anterior or posterior rotatory and bilateral rotatory injuries [14]. The child in the case report suffered bilateral dislocation of the facet joints (Figs. 3, 4, 5). This equates to a Fielding and Hawkins type III injury where there is anterior subluxation or dislocation of both facets of the atlas on the axis [3].

First presentations of acute atlanto-axial subluxations in children can be treated conservatively. The injury needs to be reduced initially through skeletal halter traction or sometimes this can occur spontaneously, followed by immobilisation with a collar for 6 weeks [11]. Failure of the deformity to improve within 2 weeks indicates further

Fig. 5 Para-sagittal CT of the cervical spine showing rotatory subluxation of the atlanto-axial joint

intervention with halo traction. The main factor correlating with recurrence and failure of conservative measures is length of delay prior to reduction. If the dislocation is not reduced and immobilised within 3 weeks there is a higher chance of recurrence and permanent deformity [9]. This child was reduced within 5 days with halo traction, and the delay may have made reduction of the dislocation with halter traction more difficult.

To date the dislocation has not recurred. We have no idea what the likelihood of recurrence is in this unique case, but we think that recurrence is probably unlikely unless there is recurrence of a similar injury to the neck and head.

Key points

- A unique case of a 15-month-old female child who suffered atlanto-axial dislocation with associated osteogenesis imperfecta.
- It important to perform a thorough musculoskeletal examination with osteogenesis imperfecta patients following trauma.
- The child benefited from a good anatomical and functional outcome with early reduction and halo traction/immobilisation for 7 weeks.

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

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