Treatment strategies for severe C1C2 luxation due to congenital os odontoideum causing tetraplegia

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Abstract

Purpose High-grade C1C2 luxation is a rare pathology. There is no clear evidence as to how to treat this deformity. There is only limited evidence about the different surgical techniques and possible approaches including advantages, disadvantages, and complications.

Methods This is an uncommon case of a 13-year-old child with progressive, tetraplegia due to congenital os odontoideum with translational instability between C1 and C2, and progressive luxation of C2. An irreducible dislocation of the C0/C1 complex caused significant compression at the cervicomedullary junction and neurologic deficit. In this

paper we highlight the different types of os odontoideum, a review of existing evidence of surgical correction. We will discuss the different treatment strategies which could be applied and the current solution will be described.

Results Continuous skeletal traction and translational reduction was achieved by a specially designed halo traction system including continuous skeletal traction in a wheelchair for 6 weeks. The surgical treatment consisted of a posterior only release, translational reduction and posterior instrumentation from C0 to C4 with a Y plate and homologous bone graft. Neurological deficits started to improve during halo traction. After surgery the patient was ambulatory without any assistance and reached a Frankel stage E. Postoperative X-rays and CT scan revealed complete reduction at the C1/C2 level and a decompressed cervicomedullary junction.

Conclusion Treatment of severe C1C2 luxation is difficult with limited evidence in the literature. The current case shows a successful treatment strategy to reduce the deformity and lists alternative approaches.

Keywords Congenital os odontoideum · C1C2 luxation · Translational instability · Neurologic deficit

Case presentation

A 13-year-old girl from Angola was admitted to our institution with progressive spastic tetraplegia due to a C1C2 luxation with significant cord compression at the level of C1 causing cervical myelopathy. The translational instability was due to a non-traumatic os odontoideum of the dens axis. At the presentation the patient was wheelchair bound, suffering from progressive tetra spastic weakness accentuated in her left upper extremity. The

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Fig. 1 **a** X-rays of the patient showing a dystopic os odontoideum with anterior luxation of the C0C1 complex. *Red arrow* posterior arch of C1. **b** CT scan showing the translational instability of the C1C2 complex

clinical investigation revealed a tetraplegia with Frankel Grade C, unable to walk or stand by her own.

Diagnostic imaging section

Plain cervical X-rays (anteroposterior and lateral, flexion/extension X-rays) showed a dystopic os odontoideum with anterior luxation of the C0C1 complex (Fig. 1a, b) in an anterior and caudal direction with dislocation of the anterior arch of C1 projecting to the level anterior of C3.

Dynamic X-rays showed a C1C2 translational instability in flexion and extension. A computer tomographic (CT) reconstruction of the cervical spine revealed a significant compression at the cervicomedullary junction at the level of the foramen magnum due to the translatory and vertical instability and dislocation of the C1C2 complex (Fig. 2). The spinal canal narrowed to less than 5 mm in sagittal diameter. The anterior and posterior arches of the atlas showed congenital malformation, whereas a CT-angiography depicted a normal course and diameter of the vertebral artery.

Historical review of the condition, epidemiology, diagnosis, pathology, differential diagnosis

Os odontoideum is defined as an ossicle with smooth circumferential cortical margins representing the odontoid

Fig. 2 Sagittal CT cut showing a dystopic os odontoideum with C1C2 luxation and significant spinal canal narrowing. *Black* posterior arch of C1, *white* os odontoideum, *red* anterior arch of C1

process that has no osseous continuity with the body of C2 [1]. Its pathogenesis as well as its treatment have been controversially debated in literature. It is a rare lesion with

no epidemiological numbers known. Anomalies of the odontoid process are more common in patients with Down's syndrome, Klippel–Feil malformation, multiple epiphyseal dysplasia, or other skeletal dysplasia disorders than they are in the general population [2, 3].

The etiology of os odontoideum remains unknown with evidence for a traumatic genesis to a chronic non-united fracture of the odontoid process [4, 5] or congenital causes as a centrum of the atlas or proatlas [6]. Anatomically it has been classified into orthotopic and dystopic types. Orthotopic os odontoideum moves with the anterior arch of C1, whereas a dystopic type is functionally fused to the basion [4].

The atlantoaxial joint has flat lateral articulations, weak posterior ligaments with the ligamentum flavum replaced with a thin atlantoaxial membrane, and no intervertebral disc and anulus to restrict motion [4].

In an intact spine the stability of the C1C2 complex during translational motion is provided by strong transverse ligaments trapping the intact odontoid process within the anterior arch of C1. A discontinuity of the odontoid process to the body of C2 as seen in traumatic or non-traumatic os odontoideum leads to atlanto-axial weakness and biomechanical insufficiency of the apical odontoid and alar ligaments. This weakness can result in instability as a loss of the ability of the spine to maintain its pattern of displacement under physiological loads [7].

The instability is associated with a translation of the C1 body and the os odontoideum relative to C2. Anterolisthesis describes the anterior subluxation of the C1-os odontoideum complex in relation to C2 during flexion. This mechanism of translational dislocation is comparable to the instability seen in patients with rheumatoid arthritis of the upper cervical spine with destruction of the cruciate ligaments due to the chronic inflammatory process [8]. Translational instability and dislocation result in a posterior spinal cord impairment. In posterior subluxation the os odontoideum and C1 are translating posteriorly into the spinal canal during extension. Anterolisthesis seems to be more often; however, some patients have instability in both directions [4, 7].

Vertical instability with dislocation of the C1C2 complex in a ventro-caudal direction causes invagination of the dens of C2 with brain stem compression and subsequent neurologic injury including respiratory paralysis. This mechanism of vertical instability is also seen in patients with rheumatoid arthritis [8].

Clinical appearance of patients with os odontoideum may vary from complete asymptomatic as in incidental imaging findings as well as neck pain, torticollis or significant neurological impairment due to acute or chronic spinal cord injury [4].

The natural history of untreated asymptomatic os odontoideum appears uncertain. The literature provides cases of asymptomatic courses [4, 7, 9] as well as

symptomatic patients after long-term follow up. Several cases of initially asymptomatic patients who developed symptomatic atlantoaxial instability due to minor trauma have been reported. Clinical symptoms range from pain to paraplegia [4, 7, 9]. Three types of os odontoideum have been described as round, cone, and blunt tooth, whereas the degree of myelopathy seems to be correlated with the round type of os odontoideum [1].

The literature provides no convincing evidence to support a diagnostic standard for os odontoideum. There is no information about the sensitivity and specificity of imaging studies. There is no evidence in literature that other imaging studies than plain X-rays needed to establish the diagnosis of os odontoideum and which patients should undergo supplemental imaging (CT/MRI) after the diagnosis have been made.

Plain X-rays of the cervical spine (anteroposterior, open-mouth odontoid, lateral) show an abnormal odontoid apex with smooth circumferential cortical margins and no osseous continuity with the body of C2 [4].

Plain lateral dynamic X-rays in flexion/extension have been used to depict the degree of abnormal motion between C1 and C2. The maximal distance the os odontoideum moves in the sagittal plane, the inner diameter of the atlas and minimal spinal diameter can be established. There is no correlation between the degree of instability and neurological status but 13 mm or less of anteroposterior spinal diameter is strongly associated with myelopathy [9, 11].

Computed tomography (CT) and magnetic resonance imaging (MRI) provide further information to assist operative planning. CT provides information about osseous abnormalities and MRI shows the degree of spinal cord compression. A CT-based angiogram is performed to show the course of vertebral arteries at C1 and C2 or the position of the transverse foramina at C1 and C2 as well as the diameter of the vertebral artery which is essential to determine the best operative procedure [7].

Even if there is no diagnostic standard it is suggested that in a symptomatic patient with neurologic symptoms X-rays, MRI scans and a CT angiogram are mandatory for diagnostic purposes and for appropriate surgical planning.

Rationale for treatment and evidence-based literature

Treatment of os odontoideum in asymptomatic patients is controversially debated in literature. An evidence-based treatment standard does not exist; however, different expert opinions based on multiple case reports are available. Asymptomatic os odontoideum with or without instability in flexion/extension X-rays can result in a long-term asymptomatic course [9, 20] or late neurological deterioration with or without trauma [4, 7, 9, 12].

Some authors recommend no treatment in asymptomatic or radiographic stable asymptomatic courses [9, 20]. The risk of an operation in those cases may be significantly higher than the risk of sudden dislocation and neurological injury during daily life activities [13].

Others recommend operative treatment in asymptomatic os odontoideum in a radiographic unstable [14] or even stable condition according to own clinical experience. Klimo et al. [7] recommend stabilization in all patients with an os odontoideum regardless of the degree of movement of the ossicle to avoid neurologic injury. The authors argue that, biomechanically, a compromise of the integrity of the odontoid-transverse ligament complex must compromise atlanto-axial stability. This instability can result in spinal cord injury in even minor stressful events [15].

Successful treatment of os odontoideum (regardless if symptomatic or asymptomatic respectively stable or unstable) aims to stabilize the C1C2 joint with or without decompression. A variety of operative techniques have been described. Early techniques use semirigid atlantoaxial fixation with dorsal bone and wire constructs which allow limited stabilization of cervical motion. They are associated with higher rates of pseudarthrosis compared to internal screw fixation techniques and need to be stabilized with a rigid external orthotics.

More modern techniques as transarticular screw fixation or rigid atlantoaxial fixation are associated with high fusion rates and provide immediate spinal stability in all planes [10, 16, 17]. In most cases no external halo orthosis is necessary postoperatively. Other methods of rigid internal fixation are C1 lateral mass screws, C2 pars interarticularis screws combined with a rod, C2 translaminar screws, C2 pedicle screws, and subaxial lateral mass screws. All of them demonstrated excellent stability in adult cadaver studies; however, neither has been studied in pediatric patients [14].

In the current case several different treatment options could be applied.

Strategy 1: intraoperative reduction and posterior instrumentation

Intraoperative reduction and instrumentation from C0 to C2 or C3 is the most straight forward technique, compared to staged procedures or preoperative halo traction techniques avoiding a long hospital stay and complications. A single posterior procedure is less traumatic and reduces risks of staged surgery and appears more comfortable especially for children and their parents. However, this technique can only be applied if the deformity is not fixed and the residual motion is sufficient to perform an intraoperative reduction. Preoperative dynamic X-rays in flexion/extension have to depict motion between C1 and C2.

However, a passive traction test under continuous fluoroscopy should be performed to assess the amount of motion between C1 and C2. This test can be done with the patient awake or in anesthesia. With the patient awake, any neurologic deficit can be detected early and traction can be released; however due to muscle tension only limited reduction will be achieved. In general anesthesia due to muscle relaxation, a maximum of reduction can be achieved, simulating the intraoperative situation; however, neurologic injury may occur. The use of SEP and MEP may help to detect any neurologic compromise early and is therefore recommended.

The literature provides cases where rapidly increasing skeletal traction was a common factor in neurologic injury [19]. Thereby intraoperative reposition can cause severe neurological impairment including sensory deficit, extremity weakness, vegetative syndromes as bradycardia, respiratory paralysis, and remaining spinal cord injury.

Strategy 2: preoperative halo-traction, reduction and secondary posterior only fusion

Preoperative halo traction allows gradual correction of the C1C2 luxation. The effect of slow reduction gives greater increase in lengths and correction with fewer disturbance of function [19]. Gradually increasing weight and close observation of children in halo traction allows contemporary intervention at the first sign of neurologic problems. The degree and progress of correction can individually be modified. In contrast to intraoperative reposition the decrease or release of weights leads to resolution of symptoms immediately in most cases. Neurological symptoms usually resolve within 1–5 months [19]. On the other hand halo traction in children is associated with significant complications in 53 %, including pin-site complications (infection, skull penetration, and brain injury) and neurological complications (extremity weakness, Horner's syndrome, cranial nerve injury, and bradycardia) [19].

Strategy 3: staged surgery including anterior and posterior release, decompression, reduction and posterior instrumentation

Due to the significant cord compression at the cervicomedullary junction at the level of the atlas and the foramen magnum, respectively and in cases of a rigid deformity with no or limited potential of reduction a primary posterior or even combined anterior posterior release with decompression may be performed. The rationale behind this strategy is to remove all posterior elements at the cranio-cervical junction (posterior arc of C1 and enlarge the foramen magnum) to release the cord. An additional

Fig. 3 Halo ring traction in a specially designed wheelchair system allowing a translation of the head

anterior, transoral approach, and anterior release may be needed to increase flexibility. However, an anterior transoral–transpalatopharyngeal approach to the craniocervical junction may be associated with significant morbidity and complications: retropharyngeal wall injury may require surgical repair. There also a risk of infection of the retropharyngeal space. Careful preoperative preparation, such as oral hygiene and prophylactic antibiotic therapy should be done [18]. To reduce the risk of infection preoperative

tracheotomy could be established which itself is associated with several risks.

Procedure (surgery, intervention)

Due to the limited flexibility on functional X-rays, a passive reduction test was performed to assess the residual C1C2 motion, the need for halo-traction or a staged release surgery as described above. Under continuous fluoroscopic imaging we performed continuous manual longitudinal traction. This procedure was not effective due to pain, anxiety and active muscle tension. Therefore, the same procedure was performed in anesthesia, accepting the risk of neurologic injury. During traction we realized that sufficient reduction could be achieved in the craniocaudal direction but not in the posterior direction (translation).

Based on the reduction test we decided to place a halo ring and perform continuous skeletal traction in a specially designed wheelchair and bed traction system allowing a translation of the head (Fig. 3). The traction procedure started with 2 kg and was increased every second day based on the patients pain level. The neurologic status of the patient was checked twice a day. Sagittal X-rays were performed every week to document axial reduction of the C0C1 complex. After longitudinal reduction was achieved, the arm of wheelchair including the halo-traction cable, was gradually tilted posteriorly over a period of several weeks to achieve translation of the C0C1 complex (Fig. 4). A few days of halo-traction started, the neurological function improved. Especially the function of the lower extremity improved to Frankel D/E. During the traction period of 6 weeks (3 weeks longitudinal traction and 3 weeks of translation) no neurological or pin-side-associated complication occurred. A satisfactory radiological reduction and good alignment could be established.

Fig. 4 Weekly sagittal X-rays to document axial reduction of the C1C2 complex. Red arrow anterior arch of C1

Fig. 5 Intraoperative fluoroscopy after reduction of C2 with a dissector in the foramen magnum

Fig. 6 Intraoperative image showing posterior instrumentation after resection of the posterior arch of C1

The surgical procedure was planned to complete the translational reduction and to perform posterior instrumentation. Under general anesthesia the halo-ring was detached and the head was positioned in a Mayfield head holder. The patient was placed in a prone position with the head under continuous traction and posterior translation. A midline exposure was carried out from the occiput to C5. The atlas showed congenital malformation with a congenital split between the anterior and posterior arch.

Instrumentation included a Y plate fixed with three bicortical screws to the occiput, lateral mass screws at the levels C3 and C4, and pedicle screws at C2. The C2 pedicles were exposed to identify the medial and lateral cortex. The starting point was marked with a high-speed drill. A dissector was placed at the medial pedicle wall to control for medial perforation and manual drilling was performed under fluoroscopic control. The posterior arch of C1 was removed with a Kerrison rongeur to get access to the

Fig. 7 **a** Postoperative sagittal X-ray showing complete reduction of the C1C2 complex. **b** Postoperative sagittal CT cut showing reduction of C1C2 complex

foramen magnum (Fig. 5). A thick fibrous membrane was identified between the foramen, the bed of the posterior arch of C1 and the lamina of C2. The membrane which was

probably due to the chronic instability was removed completely and the dura was exposed (Fig. 6). Under intraoperative fluoroscopy continuous manual pressure, directed anteriorly, was applied on the spinous process of C2 resulting in a complete translational reduction of C1 in relation to C2.

Two rods were bended 90° and placed into the polyaxial screw heads. A complete reduction could be achieved and was verified fluoroscopically by carefully inserting a dissector in the foramen magnum and under the lamina of C2 (Fig. 5). The occiput, C2 to C4 were decorticated by the high-speed drill and homologous bone graft placed to achieve posterior bone fusion. Suction drains were inserted and the wound was closed.

Outcome and follow up

Neurological deficits started to improve under halo traction. After surgery the motoric function continued to improve. 1 week after surgery the patient was ambulatory with crutches and could walk without assistance with only slight disturbances of coordination which continued to improve in the postoperative course. Postoperative X-rays revealed complete reduction of the C0C1 complex (Fig. 7a). A postoperative CT scan was performed to verify the location of the implants and the position of the odontoid process (Fig. 7b). The CT scan depicted no compression at the cervicomedullary junction of the upper spinal cord at the level of the foramen magnum. The patient was discharged 8 weeks after surgery. Follow-up investigations were performed every 6 months including X-rays and clinical examination. The last follow-up showed normal neurological function of the upper and lower extremity. The radiological follow up showed no loss of correction or loosening of the implants.

Conflict of interest None.

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