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CASE REPORT

Complete Remodeling After Conservative Treatment of a Severely Angulated Odontoid Fracture in a Patient With Osteogenesis Imperfecta

A Case Report

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Study Design. Case report.

Objective. This is the first case report describing successful healing and remodeling of a traumatic odontoid fracture that was dislocated and severely angulated in a patient with osteogenesis imperfecta who was treated conservatively.

Summary of Background Data. Osteogenesis imperfecta (OI) is a rare genetic disorder resulting in a low bone mass and bone fragility, predisposing these patients to fractures that often occur at a young age. Although any bone in the body may be involved, odontoid fractures are uncommon in this population. Because of a very high fusion rate, conservative management is accepted as a safe and efficient treatment of fractures of the odontoid in children. Several authors, however, recommend surgical treatment of patients who have failure of conservative treatment and have severe angulation or displacement of the odontoid.

Methods. A 5-year-old female, diagnosed with OI type I, presented with neck pain without any neurological deficits after falling out of a rocking chair backward, with her head landing first on the ground. Computed tomography confirmed a type III odontoid fracture without dislocation and she was initially treated with a rigid cervical orthosis. At 1 and 2 months of follow-up, progressive severe angulation of the odontoid was observed but conservative treatment was maintained as the space available for the spinal cord was sufficient and also considering the patient's history of OI.

Results. Eight months postinjury, she had no clinical symptoms and there was osseous healing of the fracture with remodeling of the odontoid to normal morphology.

Conclusion. Even in patients with OI, severely angulated odontoid fractures might have the capacity for osseous healing and complete remodeling under conservative treatment.

Key words: osteogenesis imperfecta, cervical spine fracture, cervical trauma, odontoid fracture, Anderson and D'Alonzo, angulation, displacement, cervical kyphosis, advanced imaging, remodeling, conservative management, cervical orthosis.

Level of Evidence: 5

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Osteogenesis imperfecta (OI) is a rare genetic disorder characterized by low bone mass, increased bone fragility, and susceptibility to fractures.[1,2] In the majority of cases, it is caused by a genetic defect in the biosynthesis of collagen type I. Although any bone in the body may be involved, odontoid fractures are uncommon in this population.[3,4]

Because of a very high fusion rate, conservative management is accepted as a safe and efficient treatment of certain odontoid fractures in children.[5,6] Several authors, however, recommend surgical treatment of patients who fail conservative treatment or have severe angulation or displacement of the odontoid.[6,7]

We describe a case of a young patient with OI with a type III fracture of the odontoid according to Anderson and D'Alonzo's classification,[5] who developed severe angulation and subsequent remodeling during conservative treatment.

CASE REPORT

History and Examination

A 5-year-old female with OI type I, which was diagnosed shortly after birth and treated with intravenous bisphosphonates, was transferred to the emergency department after

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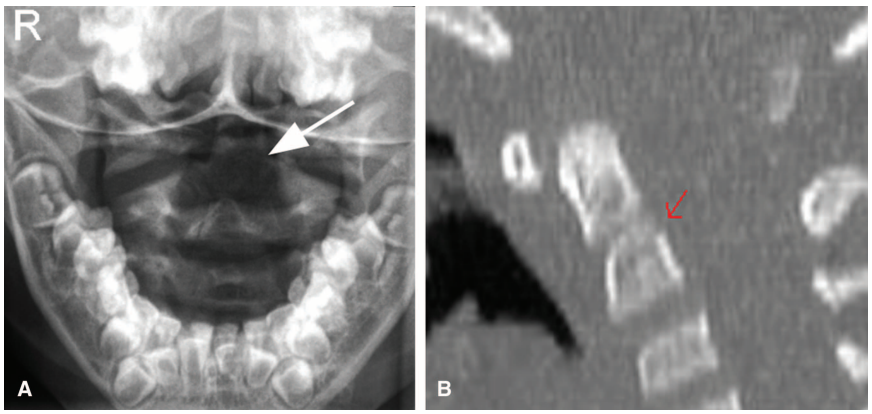

Figure 1. A, Initial odontoid view radiograph obtained at the emergency department is suggestive for a C2 fracture, depicted by the arrow. **B,** Computed tomographic scan confirmed a type III odontoid fracture without angulation or displacement.

falling out of a rocking chair backward, headfirst to the ground. Initially, she complained of only neck pain, provoked by anteflexion of the neck.

Physical examination revealed high cervical neck pain in the midline and paravertebrally on palpation without pain in the thoracic or lumbar spine. No neurological deficits or other clinical symptoms were found. Initial conventional imaging of the cervical spine suggested a fracture of C2 (Figure 1A). Additional computed tomographic (CT) scan confirmed a type III odontoid fracture without angulation or dislocation (Figure 1B). She was treated with a rigid Philadelphia collar (pediatric size) to stabilize her cervical spine.

At serial outpatient follow-up visits, she had a full range of motion without any pain or other complaints. At 1-month follow-up, computed tomography of the cervical spine was repeated, demonstrating kyphotic angulation (29°) and anterior displacement (2 mm) of the odontoid (Figure 2). She was then referred to our academic hospital, and a decision to continue with the conservative rigid collar treatment was made. At 2 months, CT scan revealed incomplete healing of the fracture, residual displacement, and sufficient space available for the spinal cord. At 3 months, follow-up CT scans with flexion-extension views showed progressive healing and no instability of fracture elements (Figure 3A, B), and the rigid collar was switched to a soft collar, which was continued during school days for 2 more months. A restriction for certain sports and strenuous activities was advised for 1 year after the initial trauma. Eight months postinjury, she had no complaints and CT scan showed further healing and remodeling of the odontoid (Figure 4).

DISCUSSION

This is the first case report describing successful conservative treatment of an isolated type III odontoid fracture with gradual kyphotic angulation and subsequent successful healing with adequate remodeling in a young patient with OI. Only a few cases of patients with OI with C2 fractures, mostly Hangman fracture type, have been described in the literature.[2,3,8]

Odontoid fractures are relatively common fractures among all upper cervical spine injuries in the pediatric population younger than approximately 7 years of age because that is the age when the synchondroses (specifically the dentocentral) of the axis close.[7] Some authors have also named these fractures traumatic epiphysiolysis or synchondrotic slips.[7] Initially, however, our patient did not have any acute displacement, and in combination with an adequate traumatic event, neck pain, characteristic CT image, angulation at follow-up, and bony fusion at follow-up, this was described a type III odontoid fracture.

There is no clear consensus about the best treatment of odontoid fractures in young children and most recommendations are based on experience with older children and adults.[4,9] A variety of treatment strategies has been proposed based on fracture type, degree of displacement and kyphotic angulation, age, and comorbidities.[9–11] Tokunaga *et al*[11] recently suggested that malunited odontoid fractures in children younger than 3 years have the capacity for remodeling to

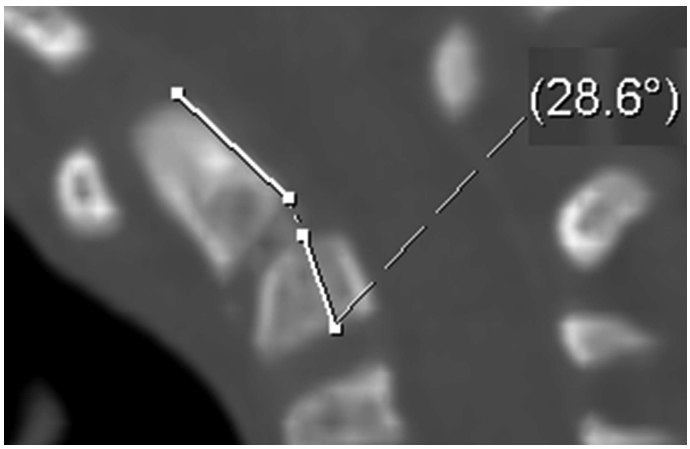

Figure 2. Computed tomographic scan of the cervical spine showing kyphotic angulation (28.6°) and anterior displacement (2 mm) of the odontoid at 1 month postinjury. Kyphotic angulation was defined as the angle between a tangent line drawn along the posterior aspect of the fractured odontoid fragment and the posterior aspect of the C2 body. The degree of (sagittal) displacement was defined as the distance between a tangent line along the anterior aspect of the odontoid fragment and the anterior aspect of the C2 body.[8] Both parameters were measured using PACS EasyVision Systems (Philips B.V., Best, the Netherlands).

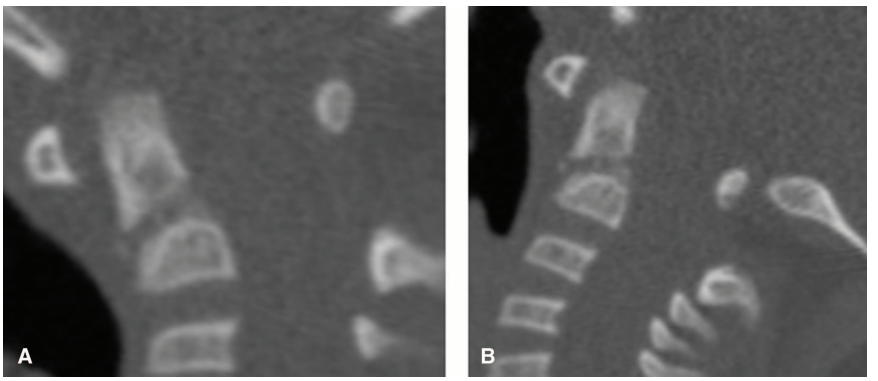

Figure 3. (A), Flexion and **(B)** extension computed tomographic scans showing a stable odontoid at 4 months postinjury.

normal morphology if the angulation of the odontoid at the time of bony union is no more than around 30°. Advanced imaging techniques play an important role in evaluating these fractures. Although magnetic resonance imaging provides relevant information about soft-tissue involvement and ligamentous and spinal cord injury, computed tomography provides good spatial resolution and the possibility for dynamic examination of the spine, avoiding the necessity of sedation in the young child.

For type III odontoid fractures, both conservative management and operative management have been applied, with fusion rates of 84% and 100%, respectively.[8,12] It has been suggested that nonoperative treatment can be safely applied in most cases of type III odontoid fractures with minimal dislocation and angulation and in those without instability and severe or progressive neurological symptoms.[8,9,11,12] However, there are no such data or evidence for the OI population, neither are there any sufficient data available to suggest superiority of any conservative treatment method, that is, rigid collar, Minerva brace, or halo vests.[6] We decided to continue the conservative rigid collar treatment that was already initiated at the outside hospital, also since significant (local) complication rates up to 68% (although not specifically for children with OI) have been reported with the use of halo vests, and the decreased bone quality in OI could hamper pin fixation in the skull.[13] Also, halo pin fixation might require a CT scan of the skull to evaluate skull thickness, increasing radiation exposure even more. Our report demonstrates that, even in the presence of progressive angulation, a displaced odontoid fracture in a young patient with OI can successfully be treated by conservative methods. However, careful consideration for each individual patient regarding the best treatment option is necessary because of the poor bone quality and ligamentous laxity seen in OI.

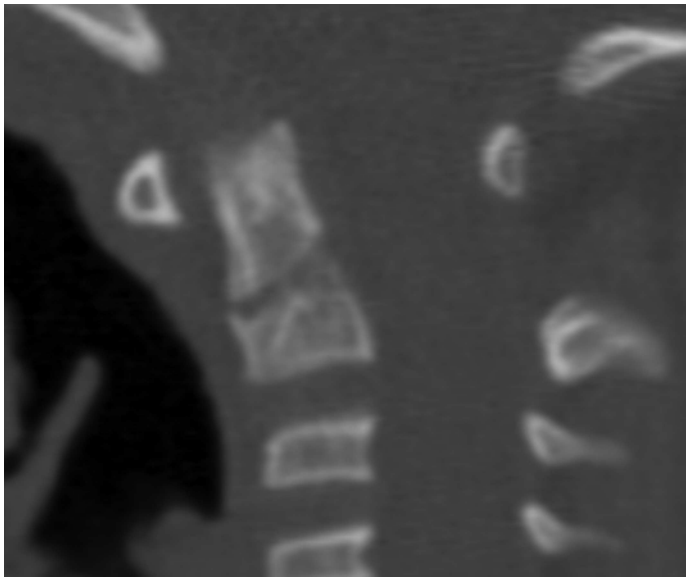

Figure 4. Remodeling of the odontoid toward normal morphology at 8 months postinjury.

CONCLUSION

Because of the rarity of the combination of OI and odontoid fractures, it is unclear how patients with this combination should be treated. This unique case indicates that type III odontoid fractures in patients with OI can be treated conservatively, despite gradual worsening of the position of the fracture fragments. Significant remodeling capacity became apparent in the course of the follow-up.

➢ Key Points

- ❑ Osteogenesis imperfecta predisposes patients to fractures; however, odontoid fractures are very rare in this population.
- ❑ This is the first case report describing an isolated traumatic type III odontoid fracture in a young child with OI, who developed progressive angulation and displacement over the course of conservative treatment.
- ❑ Despite the angulation and displacement of the fracture fragment, treatment with external immobilization combined with close clinical and radiographical follow-up resulted in complete healing and remodeling to normal morphology of the odontoid in this patient with OI.

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