Factors related to surgical outcome after posterior decompression and fusion for craniocervical junction lesions associated with osteogenesis imperfecta

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Abstract Treatment for craniocervical junction lesions associated with osteogenesis imperfecta (OI) has been described, but there are divergent views on operative procedures and preoperative and postoperative therapies due to the small number of cases. It has been suggested that a major procedure such as combined anterior and posterior surgery with concomitant ventriculoperitoneal (VP) shunting is required for OI associated with basilar impression (BI). However, here we report a case with a good outcome after posterior decompression fusion only. The patient was a 29-year-old woman with OI (Sillence type-IA) who had neurological symptoms of vertigo, nausea, and shaking during walking. Diagnostic imaging revealed hydrocephalus, severe BI, and Chiari type-II malformation. Preoperative Halo traction led to improvement in symptoms, and posterior decompression fusion from the occipital bone to C6 was subsequently performed. Lateral mass screws and Nesplon cables as sublaminar wiring for reinforcement for fusion were used in the operation. The patient wore a Halo vest for 4 weeks postoperatively. She experienced no symptoms postoperatively. Bone fusion and improved hydrocephalus were clear on images at 3 years after surgery, and the postoperative course has been good. In craniocervical junction lesions associated with OI, instability with compression of the

nerve and bone fragility in multiple sites can become problematic. Anterior odontoid resection and posterior fusion are required for OI with BI to give ideal decompression on images. However, the results of this case suggest that a good postoperative outcome can be achieved by performing not the combination of anterior odontoid resection and VP shunting, but only with posterior decompression fusion, especially for OI cases of Sillence type-I.

Keywords Osteogenesis imperfecta · Basilar impression · Hydrocephalus · Posterior decompression fusion · Anterior odontoid resection

Introduction

Osteogenesis imperfecta (OI) is an inheritable bone disorder caused by abnormal synthesis of type-I collagen. OI is classified into four types (type I, mild; II, lethal; III, severe; IV, moderate) based on clinical, radiographic, and genetic characteristics and bone fragility [1]. Recently, the Sillence Classification is expanded to include OI V–VIII, as proposed by Rauch and Cabral [2, 3]. OI has previously been associated with basilar impression (BI) [4, 5]. However, OI is relatively rare (1/20,000–30,000) and there are many asymptomatic cases; thus, there are few case reports of surgery for OI and optimal operative procedures have not been defined [5–7]. A combination of anterior and posterior surgery with ventriculoperitoneal (VP) shunting has been proposed to be required [5], but it is unclear if such a major operation is needed for all cases of OI with BI. We experienced a case of OI associated with BI, as well as Chiari type-II malformation and hydrocephalus that led to serious neurological symptoms. A good outcome was obtained

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after surgery with posterior decompression fusion, and without anterior decompression. To the knowledge, surgery for a case of OI with three complications and associated pathological conditions has not been reported previously. Thus, we describe the case and evaluate the factors associated with a good surgical outcome with reference to the literature.

Case report

The patient was a 29-year-old woman who was a housewife. She had a history of fractures in four limbs due to OI (Sillence type-IA) and had undergone right femoral distraction osteogenesis at the age of 28. Her major complaints were vertigo, nausea, episodes of apnea, shaking during walking, double vision, and perioral numbness. She had been diagnosed with hydrocephalus and BI at the Department of Neurosurgery in another hospital, and had been followed up since her chief complaint was mild headache only at that time. The symptoms described as her major complaints had developed 6 months before she visited the department. Her neurosurgeon had told her that surgical treatment was impossible because the risk was too high, and had recommended continued follow up since the size of hydrocephalus had not increased. However, her symptoms did not improve, and she visited the department.

At the initial visit, independent gait with stagger was possible and she had no bladder disturbance or rectal disorder. Numbness was in perioral regions only, and the four limbs had no symptoms. Alalia was not detected. A pathophysiological examination showed horizontal nystagmus

and hyperreflexia in the four limbs, but no muscle weakness.

Cervical spine plain radiographs did not show instability, but C1 and C2 vertebrae could not be identified on lateral radiographs, indicating severe BI (Fig. 1). In CT, BI and C1 occipitalization were clear on multiplanar reconstruction CT images (Fig. 2). In 3D-CT, the C1 posterior arch could not be identified visually, even in views from underneath (Fig. 2). T2 weighted MRI (T2 WI) showed hydrocephalus, severe BI, and Chiari type-II malformation, but syringomyelia was not detected (Fig. 3).

Regarding the patient's double vision, an ophthalmologist had confirmed that her eyesight was normal. Since there was no otological disease, vertigo was suspected to be of a central nervous origin. The Department of Neurosurgery had diagnosed hydrocephalus, but the relationship between hydrocephalus and her symptoms was unclear. We suspected these symptoms were due to compression on the brain stem and cerebellum, and scheduled an operation.

Since the level of BI was severe, Halo wheel chair traction was tried preoperatively. Taking bone fragility due to OI into account, Halo traction was started at 2 kg and gradually increased to 12 kg with careful monitoring of neurological symptoms and pain. After traction, the distance between the tip of the dens and Chamberlain's line (the line from the hard palate to the posterior margin of the foramen magnum) had slightly decreased by 4 mm and the clivoaxial angle had slightly improved from 85° to 95° (Fig. 4). MRI after traction also showed a slight improvement in the level of BI, in accord with the odontoid forward shift. Improvement of vertigo, nausea, and perioral numbness also occurred after Halo traction for 3 weeks,

Fig. 1 Plain radiograph at the initial visit. C1 and C2 were invaginated in the skull base, and only C3 and lower could be identified

Fig. 2 CT images at the initial visit. On MPR images, BI and C1 occipitalization were clear. On 3D-CT images, the C1 posterior arch was difficult to identify

Fig. 3 MRI T2WI image showing hydrocephalus, severe BI, and Chiari type-II malformation

although the changes on images were slight. The improvement in symptoms may have been due to an effect of fixing the occipital bone and cervical spines.

Preoperatively, we discussed the surgical plans with the patient and her family. First, we suggested that foramen magnum decompression (FMD) might be necessary for Chiari type-II malformation. Given the effects of preoperative Halo traction, we recommended posterior fusion from the occiput to the cervical spine. We discussed a point of

difficulty with this surgery regarding the need for concomitant anterior decompression or VP shunting. Combined odontoid resection is clearly beneficial for BI because it directly removes pressure on the brain stem. However, odontoid resection has many complications, including infection, and combined anterior and posterior surgery is a major and particularly invasive operative procedure. Also, the patient's symptoms had improved with preoperative Halo traction, and she expected to have postoperative improvement after posterior decompression and fusion without odontoid resection. VP shunting is recommended based on the literature, but shunting has problems including obstruction, and hydrocephalus may be improved after FMD and posterior fusion only. We discussed the possibility of concomitant anterior odontoid resection and VP shunting as the preferred procedure in this case, but the patient and her family selected posterior decompression fusion only after considering the benefits and risks. We also explained the possibility that anterior decompression, VP shunting or a reoperation related to OI might be required if her symptoms failed to improve or recurred after surgery.

The operation was performed with the patient in a prone position through a posterior approach under spinal cord monitoring and intraoperative Halo traction at 10 kg. The occipital bone was gradually abraded with an air drill, and FMD and C1 laminectomy were performed. Lateral mass screws (OASYS System, Stryker Spine, Allendale, NJ) were used for C4, C5, and C6. All pedicles were narrowed and pedicle screws could not be applied. We were concerned about bone fragility due to OI, and the ultra-high molecular weight polyethylene tapes (3-mm in width); Nesplon cable (NESPLON Cable System, Alfresa Pharma,

Fig. 4 CT MPR images before and after Halo traction. With traction at 12 kg, the distance between the tip of the dens and Chamberlain's line slightly decreased by 4 mm, the clivoaxial angle improved from 85° to 95°, and the odontoid shifted slightly forward

Fig. 5 Plain radiograph and MRI T2WI image 3 years postoperatively. Bone fusion was obtained and hydrocephalus was alleviated

Osaka, Japan) were used for C2, C3, C4, and C5 as sublaminar wiring to reinforce the fusion (Fig. 5). The left iliac crest bone graft was performed. The operation time was 6 h and 12 min, and the total blood loss was 270 ml. After the operation, the patient wore a Halo vest and her respiration was controlled with endotracheal intubation. The tube was extubated on the next day and her breathing stabilized. On the third day after the operation, she started walking by standing while wearing the Halo vest. She had no symptoms and the Halo vest was removed 4 weeks after the operation. On postoperative CT images the clivoaxial angle was 94°, and traction was maintained. BI remained on MRI, but was slightly improved in accord with the odontoid forward shift. The Chiari malformation was improved by FMD. At 3 years after the operation, bone fusion has been completed and hydrocephalus has also alleviated on MRI (Fig. 5).

Discussion

The rate of OI in association with BI ranges from 25 to 71% [4, 5], and the rate of OI and BI in association with

Chiari malformation has been reported to be 28% [5]. OI and BI have also been associated with hydrocephalus [8–10], but many of these cases were asymptomatic [1, 10, 11]. Thus, there are few reports of surgery for OI associated with BI and, to the knowledge, no detailed report on a patient with OI and BI associated with Chiari type-II malformation and hydrocephalus that caused neurological symptoms and required surgery.

The problematic points in surgery in the case were the severe compression of the nerve in multiple sites and the associated bone fragility. Previous reports of surgery for OI associated with BI have included FMD + laminectomy with poor surgical results [12, 13]. Transoral odontoidectomy has concern about postoperative aggravation [14–16]. Thus, Sawin et al. [5] suggested that VP shunting should first be performed in cases of BI associated with hydrocephalus, followed by Halo traction. Posterior decompression fusion could then be performed for cases with reducible BI, whereas combined odontoid resection followed by posterior fusion was proposed for irreducible BI. However, we suggest that this may not necessarily be the first choice of operative procedure, since VP shunting often causes obstruction: in a multicenter study, 39% of patients

experienced complications in 5 years after this operation [17]. Combined odontoid resection is clearly beneficial for BI because it directly removes pressure on the brain stem. However, odontoid resection has many complications, including infection. In addition, anterior decompression by odontoid resection can result in aggravation of postoperative instability, especially in cases of OI, and posterior fusion is then required. Thus, patients undergo major surgical intervention in such combined operations. We considered the possibility of anterior odontoid resection and VP shunting as the preferred procedure in this case, but a discussion with the patient and her family about the merits and risks led to selection of a less invasive operative procedure.

There is a report of a patient with OI and BI associated with syringomyelia who underwent posterior fusion without posterior fossa decompression and had a good postoperative course [6]. In contrast, Noske et al. [7] reported that a patient who underwent Halo traction and achieved improvement in symptoms, and then underwent posterior decompression and fusion, showed improvement for 2 years, but hydrocephalus and other symptoms then worsened and VP shunting was required. This led to the conclusion that symptoms and signs of raised intracranial pressure and hydrocephalus caused by BI complicating OI can be treated by axial halo traction with subsequent posterior solid fusion only, but additional VP shunt placement may also be needed. However, we believe that there are two differences between this case and our case. First, the Sillence classification was type IV in Noske, while the present case was type I, indicating milder OI. Second, the patient in the Noske case was a 22-year-old man who did head spins on the ground without using his hands, as part of a Brazilian martial arts group. He gave up martial arts postoperatively, but his daily activity level was probably significantly greater than the patient in the case. For these reasons, the Noske case might have shown greater disease progression postoperatively. Therefore, we do not think the case will need VP shunting in the immediate future, since bone fusion and improved hydrocephalus have been obtained. To date, the relationship between the surgical method and OI classification has not been examined because of the small number of cases, but the disease severity of OI should be taken into account. However, in either case, OI is a progressive disorder and the long-term prognosis remains uncertain. Careful follow up is required to recognize recurrence of symptoms and signs of hydrocephalus. The case indicates that a good surgical outcome can be achieved with posterior decompression and fusion for BI complicating OI, especially in a patient with type I OI and relatively moderate levels of daily activity.

Posterior decompression fusion for a patient with the four conditions of OI, BI, Chiari type-II malformation, and

hydrocephalus has not been reported previously. In this respect, we were concerned that improvement of symptoms might be mainly from FMD for Chiari type-II malformation because of the mild imaging improvement of BI compared to the good outcome of FMD. This may be somewhat true, but cannot account for the major improvement of symptoms preoperatively with Halo traction before FMD. The improvement in hydrocephalus and absence of recurrence of symptoms also support the importance of the effects of posterior decompression and fusion on BI. Furthermore, the improved symptoms with Halo traction, despite the slight changes on images, strongly suggest not only a decompression effect with traction but also an effect of fixation of the occipital bone and cervical spines in this case. This outcome suggests that posterior decompression and fusion may be useful for treatment of OI lesions. A good surgical outcome requires evaluation of the extent of improvement in symptoms under preoperative Halo traction, and postoperative solid bone fusion without aggravation. Augmentation of fusion using intraoperative sublaminar cables and a postoperative Halo vest are also important.

Conclusion

A good surgical outcome was obtained with posterior decompression fusion only in a case of OI and BI associated with Chiari type-II malformation and hydrocephalus. Thus, VP shunting and odontoid resection may not be needed in all cases of OI and BI, if improvement in symptoms with preoperative Halo traction is obtained. These should be considered only when improvement in symptoms is not obtained or symptoms worsen after posterior decompression and fusion. As the important factors related to good surgical outcome, use of variable anchors and fixation with a Halo vest postoperatively can be used to promote solid bone fusion. Furthermore, severity of OI and the level of activities of daily living may be associated with postoperative aggravation of BI or hydrocephalus.

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

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