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Case report

A MODIFIED PIN AND WIRING TECHNIQUE FOR OCCIPITOCERVICAL FUSION IN A CHILD WITH OCCIPITALIZATION OF THE ATLAS: A CASE REPORT

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Abstract

A rare case of occipitalization of the atlas with spinomedullary compression was reported. The child presented with neck pain and quadriplegia. Radiographic images showed occipito-atlantal instability. Posterior stabilization was performed using a contoured pin modified in Maharaj Nakorn Chiang Mai Hospital, wiring, and strut bone grafts between the occiput and C4 vertebra. Use of the modified pin and wiring technique, as described herein, achieved solid bone fusion and maintained stable constructs. The patient has shown symptomatic improvement and no remarkable complications after 5 years postoperatively. **Chiang Mai Medical Journal 2010;49(1):35-39.**

Keywords: occipitalization of atlas, occipitocervical fusion, occipito-atlantal instability, contoured pin

Occipitalization of the atlas is congenital bony fusion of the atlas to the base of the occiput. The congenital malformation is also defined as assimilation of the atlas or atlanto-occipital fusion. The reported prevalence of occipitalization of the atlas is 1.4-2.5 per 1,000 children.[1-3] Due to the rarity of this condition, no definite incidence has been reported in Thailand.

Congenital fusion of the first cervical vertebra with the base of the occiput is caused by a failure of segmentation during embryonic development. Hypoplasia of the odontoid might be associated with occipitalization of the atlas and may lead to instability. Due to its proximity to the spinomedullary area, there is an approximate 50% rate of subsequent atlantoaxial instability with development of neurological deficit.[4]

A child with occipitalization of the atlas, with atlantoaxial instability, may not have neurological deficits until their second or third decade of life. Routine plain films of the cervical spine are difficult to interpret for congenital anomalies. In a child, occipitocervical stabilization for restoring stability has always been a surgical challenge because of the small bony structure and anatomical complexity in this area. In 1927, Forester initially reported occipitocervical fusion by using a fibular graft between the occiput and C1 to stabilize a progressive atlantoaxial dislocation after an odontoid fracture. Since then, more advanced occipitocervical stabilization techniques have been steadily developed.

At present, occipitocervical stabilization, with rigid internal fixation techniques, is the standard

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procedure in adults, but these methods cannot be achieved easily in pediatric patients, due to their smaller anatomy. In addition, the costs of modern implants are very expensive. To promote bony fusion of the occipitocervical area, most surgeons use corticocancellous graft harvested from the iliac crest. Attachment of the graft may be performed with rigid wiring or heavy suture combined with various methods of postoperative immobilization. Kumar R, et al described a technique for occipitocervical fusion using a contoured rod, sublaminar wire fixation and onlay bone grafts in a case of assimilated atlas, and reducible C1-C2 dislocation.[5]

We present the case of a child with occipitalization of the atlas and odontoid hypoplasia, in which we performed occipitocervical fixation with a contoured Steinmann pin modified in Maharaj Nakorn Chiang Mai Hospital.

Case report

A ten-year-old boy had a three-month history of neck pain. The pain radiated intermittently into his right shoulder and arm. He started to feel weakness in the right upper extremity two months before seeking treatment. Later, he had weakness in the left upper extremity. Two weeks prior to hospitalization, he had muscular weakness in both lower extremities and there was paresthesia of his legs. His gait had become impaired because the weakness involved his lower extremities. There was no bowel or bladder dysfunction, and no significant preceding trauma or past medical history.

Physical examination revealed weakness in all four limbs, while muscle strength was greater in the lower (grade III-IV) than the upper extremities (grade I-II). Decreased sensation of deep pain below both knees was observed. His deep tendon reflexes were hyperactive bilaterally throughout his upper and lower extremities.

The plain radiographs of the cervical spine showed an assimilated posterior arch of the atlas. The images suggested occipito-atlantal instability, with forward displacement of the anterior arch of the atlas and a decreased antero-posterior spinal canal diameter at this level (Fig. 1). MR images of the cervical spine revealed bony fusion of the posterior arch of the atlas, as well as the occiput and presence of a focal hyperintense signal of the cord on the T2-weighted image with spinal cord compression at the level of the atlas. Hypoplasia of the odontoid process was also noted (Fig. 2).

We performed a posterior midline approach to expose the occiput and upper cervical spine. A stainless steel Steinmann pin (4.0 mm diameter) was preoperatively contoured, corresponding to the skull shape of our patient. The contoured Steinmann pin, modified in Maharaj Nakorn Chiang Mai Hospital, was fixed to the occiput and the upper cervical spine (Fig. 3). The strut

Figure 1. Lateral radiograph of craniocervical region. The posterior arch of the atlas is fused to the occiput, and hypoplasia of the odontoid process can be identified. Note; superior protrusion of dens into the foramen magnum.

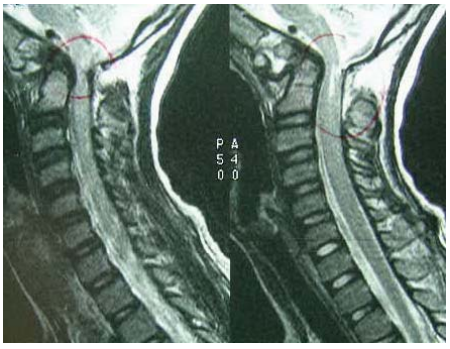

Figure 2. Preoperative magnetic resonance imaging shows constriction of the dural sac at the cervicovertebral junction and hyperintense change in the spinal cord on a T2-weighted MR image.

Figure 3. The picture shows the U-loop contoured Steinmann pin (4.0 mm), which was modified at Maharaj Nakorn Chiang Mai Hospital.

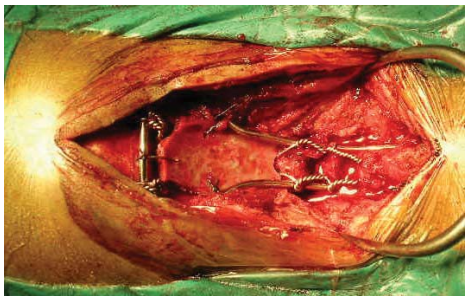

Figure 4. Operative view of occipitocervical fusion using a U-loop contoured pin secured with wiring and onlay iliac crest autograft.

grafts were harvested from the posterior iliac crest and placed bilaterally along the occipito-cervical region for arthrodesis. Stainless steel wires were applied to fix the contoured pin to the occiput and cervical spine. Reduction and alignment of the occiput and cervical spine were controlled by positioning the head with skull traction and appropriate tightening of the wires. After acceptable alignment had been confirmed, morsellized iliac autograft was packed into the arthrodesis site (Fig. 4), and the wound was closed in layers. Skull traction was applied and maintained for 4 weeks after surgery. Then the patient was allowed to ambulate with an SOMI brace.

In the first week of the postoperative period, the patient had improvement in deep pain sensations. In addition, muscle power improved in both the upper and lower extremities. At 8 weeks, his sensory loss was almost completely resolved. The intrinsic power in his hands, and muscle power of his legs had significantly improved by the time he was discharged from hospital. He was fully ambulatory without walking aids, but still had some impairment in movement of his hands.

Twelve months after surgery, the sensory disturbances had disappeared, and there was no identifiable weakness in the extremities. Also, the patient no longer suffered neck and shoulder pain. He had some limitation in craniovertebral junction mobility, but his neck motion was compensated in the lower cervical spine. Radiographs of the cervical spine showed the incorporated graft and a bony union at the fusion site. The spine was also stable in flexion and extension.

At 5 years after surgery, the patient had no pain, neurologic symptoms, or limitation of the neck, which had interfered with his activities of daily living. He had no pain where the implant was fixed to the occiput, and no wound breakage of the surgical scar. No broken wire or arthrodesis failure was observed (Fig. 5).

Discussion

One of the most common congenital malformations at the occipito-cervical region is occipitalization of the atlas, in which there is a partial or complete bony fusion between the atlas and

Figure 5. At 5 years follow-up, plain radiograph shows stable fixation with bony fusion from the occiput to C4, without breakage of the constructs.

base of the occipital bone.[6,7] Commonly associated anomalies include congenital cervical fusion, hypoplasia of the axis, and Arnold Chiari malformations type I.[8] The symptoms are usually affected by compression from the occipito-cervical canal stenosis or instability.

Detection and treatment of this deformity are important, as it often causes cervical spine instability and basilar impression. Special imaging such as CT scan, three-dimensional CT scan, or MRI should be obtained before surgery. The goal of the treatment is to solve the problems and stabilize the cervical spine. The recommended treatment for occipitocervical instability is occipito-cervical fusion, followed by stabilization in this area using either a semirigid or rigid instrument.[9-11]

Our patient presented with pain of the right shoulder and arm, weakness of all extremities, and gait impairment. MRI images revealed assimilated posterior arch of the atlas with the occiput. Instability and spinal cord compression of this area was also observed. Stabilizing the bony structures of this area was quite challenging, when selecting an implant for the patient, because the small bony structures were irrelevant to rod-screw implants, and the patient had financial problems.

Given the cheap cost and availability of the pin (approximately 400 baht), we were able to modify the implant by contouring a conventional 4.0 mm Steinmann pin and applying it with sublaminar wires to improve sagittal alignment. Although the implant might not be as strong as a modern one (like rod screw fixation), the pin plus wires were usable and easy to apply. Autologous strut grafts, harvested from the posterior iliac crest, and a cancellous allograft were used for occipito-cervical arthrodesis. We needed to immobilize the patient in traction postoperatively for several weeks and apply an SOMI brace until bony union.

After 5 years postoperatively, no breakage of metal, pressure sore from the modified pin at the occiput area, or secondary deformity of the cervical spine were seen. The patient had limited craniovertebral junction mobility, but no difficulty in performing his activities of daily living. The patient can compensate by motioning the lower cervical levels.

Conclusion

Although several implant designs for occipitocervical stabilization have been developed, they may not be appropriate for small bony structures in children. In this report, we demonstrated good results from an economical contoured Steinmann pin, which was modified in our hospital and then secured with wires for occipitocervical stabilization in a young patient.

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รายงานผู้ป่วยเด็กที่มีความผิดปกติของกระดูกสันหลังส่วนคอ Atlas เชื่อมติดกับกระโหลกศีรษะส่วน occiput ซึ่งได้รับการผ่าตัดรักษาเชื่อมกระดูกโดยวิธีใช้ลวดโลหะที่ถูกดัดแปลงในโรงพยาบาลมหาราชนครเชียงใหม่

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ภาควิชาออร์โทปีดิคส์ คณะแพทยศาสตร์ มหาวิทยาลัยเชียงใหม่

บทคัดย่อ

รายงานผู้ป่วยที่มีภาวะความผิดปกติของกระดูกสันหลังส่วนคอ atlas เชื่อมกับกระโหลกศีรษะส่วน occiput ซึ่งมีการกดทับประสาทไขสันหลังส่วนต้นคอ เด็กมาพบแพทย์ด้วยอาการปวดต้นคอ และแขนขาอ่อนแรง ภาพถ่ายทางรังสีพบความไม่มั่นคงบริเวณกระดูกกระโหลกศีรษะ occiput และกระดูก atlas ผู้ป่วยได้รับการผ่าตัดรักษาโดยใช้ลวดโลหะที่ถูกดัดแปลงในโรงพยาบาลมหาราชนครเชียงใหม่ ให้เข้ารูปกับกระดูกบริเวณนี้ และเชื่อมกระดูกกระโหลกศีรษะกับกระดูกสันหลังส่วนคอ ถึงกระดูกคอชิ้นที่ 4 พบว่าการผ่าตัดโดยใช้โลหะแบบนี้สามารถให้ความมั่นคงกับกระดูกได้ และทำให้มีการเชื่อมกันของกระดูกได้ดี ผู้ป่วยมีอาการทางคลินิกดีขึ้นชัดเจน และไม่พบผลแทรกซ้อนจากการผ่าตัดด้วยวิธีนี้ หลังจากติดตามผู้ป่วยนานถึง 5 ปี **เชียงใหม่เวชสาร 2553; 49(1):35-39.**

คำสำคัญ: ความผิดปกติของกระดูก atlas การเชื่อมกระดูกกระโหลกและกระดูกสันหลังส่วนคอ กระดูกสันหลังส่วนคอไม่มั่นคง การดัดลวดเหล็ก