Occipito - C2 laminar screws fixation for treatment of chronic C1-2 instability - Case report

Fixação occipito - C2 com parafusos de lâmina para tratamento de instabilidade crônica C1-2 – Relato de Caso

RESUMO

Introdução: As doenças da transição crânio-cervical (CVJ) abrangem inúmeras afeções, como a instabilidade C1-2. O tratamento cirúrgico é necessário em alguns casos, especialmente frente à presença de déficits neurológicos. Neste contexto, quando indicado um procedimento cirúrgico, inúmeras técnicas cirúrgicas de fixação são descritas para esta região, variando conforme as características individuais de cada caso.

Métodos: Apresentamos o caso de uma jovem de 12 anos com severo comprometimento neurológico (não deambulante, com tetraparesia grave de instalação progressiva) secundário à subluxação C1-2 crônica, com compressão neural. Submetida a tratamento cirúrgico por via posterior, através de fixação occipito-cervical com instrumentação da lâmina do áxis, com concomitante descompressão posterior, ocorreu importante melhora neurológica após o procedimento.

Discussão: O tratamento das doenças que acometem a junção crânio-cervical é complexo, requerendo o domínio de inúmeras técnicas de fixação, tendo em vista as variações anatômicas amplas presentes na região. O conhecimento destas técnicas é necessário para se atingir bons resultados. Uma vez que o tratamento das doenças desta região deve ser individualizado conforme as características de cada paciente.

INTRODUCTION

Anomalies of the craniovertebral junction (CVJ) include many conditions, as assimilation of the atlas and other segmental defects, basilar invagination, atlantoaxial instability and other malformations. They can be isolated or part of clinical conditions such as achondroplasia or Down syndrome. Congenital anomalies of the CVJ can be asymptomatic for years or present with severe neurological symptoms and sudden death\(^1\),\(^4\),\(^20\).

Although controversies exist in the literature\(^2\), in cases with important neurological symptoms, surgical treatment is generally indicated\(^2\),\(^3\). This paper discusses surgical strategies and fixations options for a pre-adolescent patient with a C1-2 chronic instability and severe neurological deficits.

CASE REPORT

A 12-year-old girl presented with a 3-month history of progressive gait impairment, leading to inability to walk two months before admission. The neurological examination revealed global muscle strength grade 2/4 and upper motor neuron findings (spasticity, hyperreflexia, clonus and Babinski signs). She also had bladder and bowel dysfunction. This patient had a history of multiple bone and joints deformities with no definitive diagnosis. She had undergone bilateral femur osteotomies and maxillofacial surgery due to congenital hip dysplasia and difficulties in mouth opening.

A previous magnetic resonance imaging (MRI) of the cervical spine at the age of four (Fig. 1A) showed a C1-C2 subluxation and upper cervical stenosis, probably linked to an os odontoideum, although clinical symptoms were not reported by her parents. A new MRI showed severe worsening of the upper cervical stenosis, the C1-2 subluxation with cranio- cervical kyphosis and a clivo-canal angle of roughly 90o degrees (Fig. 1B).

A flexion-extension computed tomography (CT) of the CVJ failed in showing significant change in subluxation, probably due to its chronicity. Manual traction also failed in determining radiological changes. Careful neurological monitoring during preoperative dynamic CT is advised and symptom arousal provides information on intraoperative risks related to positioning; however, this patient did not present neurological symptoms even during maximal flexion (Fig. 2).

She was positioned in mild extension and distraction. We favor this position for occipito-cervical fusion because it allows the use of intraoperative radiological antero-posterior x-rays\(^21\). A wide occipito-cervical exposure was performed. C1 posterior arch was resected and the posterior margin of foramen magnum opened. Due to the inability to reduce the C1-2 subluxation, a selected C1-2 fusion was not considered once insertion of C1 lateral mass screws was challenging. The subaxial (C3-6) spine had also very small lateral masses, which could support just less than 10 mm lateral mass screws. Based on this, an occipito-C2 laminar fusion was performed. Laminar screws were chosen because her axis anatomy precludes the use of pedicular screws, as shown in Figure 3. The laminar screws used were 4 mm width and 20 to 22 mm length each, resulting in a good bone purchase. After system fixation, rib graft was added (Figure 4). The patient had no complications and was discharged home at postop day 3, wearing a rigid cervical collar.

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Relato de caso

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Three months later, the patient started walking with assistance, global muscular strength increased to a grade 3-4/4 and she recovered bladder control.

DISCUSSION

When discussing atlanto-axial dislocation, the main question to be answered is about its reducibleness. Lesions are considered fixed or irreducible when no reduction of the dislocation is demonstrated on full neck extension or after cervical traction. Many authors consider transoral decompression and subsequent craniocervical posterior fixation. However, Goel et al. proposed that transoral decompression is unnecessary, once an isolated posterior approach is possible, treating the C1-2 joints using an interbody spacer to distract them, followed by C1-2 fusion.

When C1-2 fixation is necessary, the most important point to be considered is the specific anatomic patterns of this region, described elsewhere in detailed. Although the use of C1 lateral mass can be feasible in almost all patients, in severe anterior C1 subluxation, as in our case reported, this can be extremely challenging. Morphological individual variations of C2 anatomy demand the knowledge of several possible fixation techniques to achieve fusion.

Although methods of atlantoaxial fixation using wiring techniques have historical value - like the Brooks or Gallie techniques - their performance ask for complete integrity of the posterior elements, rigid immobilization after surgery is required and high nonunion rates (up to 30%) have been reported. Several techniques using C2 screws have been described, specially for trauma. From a biomechanical view point, transarticular C1-2 screws and pars, pedicle and laminar screws are superior to wiring.

However, transarticular, pars and pedicle C2 screws can sometimes be precluded due to anatomical variations, including small pars in children or a medially located vertebral artery located (in about 20% of the patients). In such cases, as in our patient, the use of laminar screws can be a safe option, or maybe the only one. This technique allows for immobilization of the axis without risking the vertebral artery and can be used in almost all patients, including children. Cadaveric studies suggest that, although pedicle screws had the highest insertional torque, lamina screws appeared to provide stronger fixation than pars screws. Long term follow-up of the different techniques of axis instrumentations, assessing complications, such as adjacent level degeneration and hardware failure, are necessary to define the optimal technique to be chosen in C2 fixation.

CONCLUSION

Management of occipito-cervical malformation is complex. An extensive knowledge of fixation techniques available for occipito-C1-2 fusion is needed due to frequent anatomical variations. These techniques allow for good clinical outcomes without added morbidity, when properly used in carefully selected cases.

REFERENCES


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