Occipitocondylar hyperplasia: an unusual craniovertebral junction anomaly causing myelopathy

Case report

CHIMA OHAEBULAM, M.B., B.S., ERIC J. WOODARD, M.D., AND MARK PROCTOR, M.D.

Department of Neurosurgery, Children’s Hospital, Brigham and Women’s Hospital, Harvard Medical School, Boston, Massachusetts

Osseous anomalies of the CCJ have a wide spectrum, and have been characterized extensively in the past 25 years.1 We report a rare craniovertebral junction anomaly, bilateral occipital condyle hyperplasia, not previously described in the literature.

Case Report

Presentation and Examination. This 10-year-old Argentinian girl was referred to our institution for treatment 1 year after she suffered an SCI after a fall during judo class. She suffered a left hemiparesis, which completely resolved over the ensuing 3 months.

Plain radiographs of her neck at the time were interpreted as being normal. Magnetic resonance images of her brain and cervical spine were obtained (Fig. 1) and demonstrated severe bilateral condylar hyperplasia with ventral compression of her CMJ. A CT scan was also obtained that more clearly demonstrated the bone anatomy in this area (Fig. 2).

Although she had no further neurological decline, the child continued to have neck pain, which waxed and waned with exertion while surgical opinions were sought by the family. Both a transoral approach for decompression and a bilateral far-lateral decompression with stabilization had been recommended prior to the patient’s presentation to our group.

At the time of her initial assessment at our institution, the patient demonstrated full strength in all her extremities. She was hyperreflexic in her lower extremities and had normal reflexes in her upper extremities. She had positive Babinski reflexes bilaterally. Her neck had normal range in flexion and extension, but she was able to achieve no more that approximately 60° of rotation in either direction.

In addition to the previously noted findings on her MR images and CT scans, we believed her to have some T2-weighted signal hyperintensity, probably reflecting an old SCI. Flexion–extension cervical spine radiographs demonstrated no evidence of instability.

A purely dorsal approach was recommended to the patient and her family, in an effort to avoid the morbidity aspects of a transoral approach. It was believed that a midline approach would accomplish the goal of drilling down the hypertrophied medial portion of the occipital condyles while staying out of the atlantooccipital joints.

Surgical Procedure. The patient and her family elected to pursue the dorsal approach, which was accomplished through a standard midline suboccipital incision. The arch of C-1 was identified and dissected out laterally on both sides as far as the joint capsule of the atlantooccipital joint. The vertebral artery was skeletonized and mobilized superiorly. A diamond burr was used internally to decompress the medial occipital condyles bilaterally, then a curette was used to crack the residual cortical bone into the cavities thus created. The anterior–posterior extent of the decompression was assessed using an intraoperative lateral radiograph. The joint capsule remained intact throughout.

Abbreviations used in this paper: CCJ = cranio cervical junction; CMJ = cervicomedullary junction; CT = computerized tomography; MR = magnetic resonance; SCI = spinal cord injury.
inspection confirmed the excellent extent of decompression achieved. The wound was closed in a standard fashion.

Postoperative Course. Postoperatively, the patient did very well and had no neurological deficits. A CT scan confirmed the extent of decompression and demonstrated no subluxation suggestive of instability (Fig. 3). She was discharged from the hospital on the 2nd postoperative day.

The patient has subsequently gone on to do well, with no neck pain, improved neck range of motion, and resolved signs of myelopathy at her 1-year follow-up visit. A follow-up MR imaging procedure performed at this time demonstrated excellent decompression and alignment (Fig. 4). We plan to follow the patient over time to ensure that any subsequent atlantooccipital instability is identified.

Discussion

Although CCJ anomalies have been recognized for cen-
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Fig. 4. Sagittal (left), coronal (center), and axial (right) T2-weighted MR images of the CCJ obtained 1 year postoperatively, demonstrating the absence of compression at the CMJ.

turies, they have only been thoroughly characterized to encourage a systematic approach to their treatment in the past 25 years.1

During development, the proatlas is the fourth occipital sclerotome.3 The neural arch component of the proatlas divides into rostral ventral components and caudal dorsal portions. The anterior, U-shaped margin of the foramen magnum is formed by the rostral ventral component; this structure also forms the occipital condyles. A third condyle may be present in the midline.2 The caudal dorsal division of the neural arch of the proatlas forms the lateral atlantal masses and the superior portion of the posterior arch of the atlas.3

The excessive growth in the relevant portion of the proatlas is likely the origin of the condylar prominence in this patient. The radiographic cord compression and clinical manifestations of myelopathy and prior SCI in our patient made it imperative to decompress the upper cervical cord to prevent further injury. Although transoral approaches to the CCJ have revolutionized the treatment of pathological entities in this area,1 they are not trivial undertakings by any means.

Because the source of compression was ventrolateral to the thecal sac (rather that directly ventral) in this patient’s case, we decided that a midline dorsal approach would allow enough access so that a satisfactory decompression could be performed. It would also be possible to stay outside the atlantoccipital joint and avoid any instability that might result from disruption of these structures. At surgery, these goals were achieved without difficulty.

Image-guidance systems might have assisted in more precisely ensuring that preoperatively determined limits for resection of the condyles were not exceeded. No image-guidance system that might have permitted this was available for our use at the time, however, and to our knowledge, no such application of these systems in a similar context has been described in the literature.

Conclusions

This unusual case demonstrates the complexity of development of the craniovertebral junction and illustrates the features of a previously unreported abnormality in this area. It also serves to highlight some of the factors to be considered in deciding between anterior and posterior approaches to congenital pathological entities in the area.

References


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Address reprint requests to: Chima Ohaegbulam, M.B., B.S., Department of Neurosurgery, Bader-3, 300 Longwood Avenue, Children’s Hospital, Boston, Massachusetts 02115. email: cohaegbulam@partners.org.