Occipitocondylar hyperplasia and syringomyelia presenting with facial pain

Case report

JAMES S. WALKDEN, M.B.CH.B.(HONS), M.R.C.S.(Ed),
RICHARD A. COWIE, M.B.CH.B., F.R.C.S.(SN)(Ed),
AND JOHN A. THORNE, M.B.CH.B., F.R.C.S.(SN)

Department of Neurosurgery, Royal Manchester Children’s Hospital, Manchester, United Kingdom

The authors describe a unique presentation and long-term management of a rare craniovertebral abnormality in a patient presenting to their institution. This 10-year-old girl presented with right-sided facial pain and subjective dysesthesia of the chest wall without evidence of cervical myelopathy. She was found to have extensive cervicothoracic syringomyelia secondary to compression at the foramen magnum by hypertrophic occipital condyles. Posterior decompression and medial condylectomy was performed, with significant radiological and clinical improvement over the next 5 years of follow-up. The authors discuss the clinical pathophysiology and operative techniques used.

KEY WORDS • occipitocondylar hyperplasia • craniovertebral junction • syringomyelia • atypical facial pain • congenital anomaly

The bony anomalies of the craniovertebral junction (CCJ) and their underlying embryology are well described in the surgical literature. We noted only one published case report of occipitocondylar hyperplasia that was associated with myelopathy and previous spinal cord injury. We describe a previously unreported presentation involving facial pain and chest dysesthesia secondary to extensive syringomyelia associated with this rare craniovertebral junction abnormality. We describe the presentation, investigation, and long-term management of this unusual condition.

Case Report

Presentation and Examination. This 10-year-old girl was referred from her general practitioner because of a short history of right-sided facial pain over the previous 3 months. The pain was described as burning in nature and was nearly constant over the right V2 and V3 distributions of the trigeminal nerve. This pain had remained unresponsive to simple analgesia over this time, and gabapentin administration was being considered. There was absolutely no history of preceding trauma and there was no relevant family history. On review at our institution we also elicited a history of associated dysesthesia over the right face, neck, and chest, which had been intermittent over the previous 3 months. The patient absolutely denied any neck pain or limb symptoms such as weakness or sensory disturbance. There was a history of intermittent occipital headaches but these were not associated with position or straining.

On examination the patient had fully intact cranial nerve function, although she did report subjective dysesthesia over the right face, neck, and chest down to the T-4 level. Despite this she was able to discriminate light touch, pinprick, and temperature sensations completely on objective testing. Results of her limb examination were entirely normal and she had normal gait including heel-toe and Romberg testing. The right knee reflex was slightly brisk but otherwise all limb reflexes including Babinski responses were entirely normal, with no clinical evidence of cervical myelopathy.

Neuroimaging Investigation. Admission MRI sequences of the brain and whole spine demonstrated cervicomедullary compression at the level of the foramen magnum, associated with extensive cervicothoracic syringomyelia extending to the T-12 level. Grossly hypertrophied occipital condyles were noted, and there was no associated hydrocephalus (Figs. 1 and 2).

We performed urgent CT and CT angiography studies with 3D reconstruction to further characterize the lesion and any associated vertebral artery (VA) anomalies that are described with CCJ abnormalities. We also obtained CT scans for image guidance intraoperatively (Figs. 3 and 4).
Operation. Following extensive discussion with the patient and her family we proceeded to decompress the cervicomedullary junction from a dorsal midline approach. This was performed with the patient prone in Mayfield pins and with neuroimaging studies (Brainlab, Inc.) to help guide the extent of condylar resection and aid arterial preservation. The foramen magnum lip and C-1 arch were exposed laterally to the VAs on both sides. A suboccipital extradural decompression was first performed to enable improved retraction and to prevent any undue medullary compromise during the drilling stage. The VAs were skeletonized and retracted superiorly to expose the medial hypertrophied condyles, which were drilled from a medial to lateral direction using a handheld, high-speed diamond bur.

The image-guidance system was used continuously to avoid disrupting the atlantooccipital joint and possible secondary instability in the future. We encountered difficulty resecting the most inferior part of the occipital condyles behind the arch of C-1 from above the VAs, and elected to then retract them inferiorly for safe completion of the drilling inferiorly. Prior to closure the degree of decompression and dural expansion was excellent visually and radiologically on image guidance, and the atlantooccipital joints appeared intact, with no vascular complications encountered. The muscle and fascia were closed in layers by using subcuticular sutures to the skin.

Postoperative Course. The patient sustained no neurological deficits in the postoperative period and remained hemodynamically stable throughout her stay. There were no wound complications and the patient was discharged on the 7th postoperative day in a soft cervical collar following physiotherapy assessment. We performed CT and dynamic radiographic imaging prior to discharge to en-

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**Fig. 1.** Axial T2-weighted (A), coronal T1-weighted (B), and sagittal T1-weighted (C) MRI demonstrating cervicomedullary compression by hypertrophied occipital condyles.

**Fig. 2.** Sagittal T2-weighted MRI studies demonstrating extensive cervical (A) and thoracic (B and C) syringomyelia from the C-2 to T-12 vertebral levels.
Sure satisfactory decompression and to confirm that there was no atlantooccipital instability (Fig. 5).

**Long-Term Course.** At first follow-up 12 weeks postoperatively the patient had noted complete improvement in her facial pain and chest dysesthesia. There was still some residual pain around the operative site associated with occasional occipital headaches, but on neurological examination her nerve function remained intact (Fig. 6).

We continued to review the patient on a 6-month basis and performed further MRI sequences at 6 months and 1 year, along with dynamic radiography. At 6 months there was complete resolution of the presenting symptoms along with the postoperative wound pain and headaches. Serial MRI, as shown, demonstrated gradual reduction in the syrinx cavity; on 3- and 5-year imaging sequences there is only a small cavity remaining at the level of the C-5 vertebral body. At the 5-year follow-up visit the patient remains asymptomatic with no symptoms of neck pain or suggestive of dynamic instability. The yearly dynamic radiographs up to 5 years postoperatively have shown no radiological evidence of instability (Fig. 7).

**Discussion**

The CCJ is a complex anatomical area and may be associated with a wide spectrum of clinical presentations due to degenerative, compressive, or vascular conditions. The embryological basis of craniocervical disorders is well described and occipitocondylar hyperplasia is linked to malformation of the proatlas and the 4th occipital sclerotome.9,10 This is a very rare malformation, with only single case reports of bilateral occipitocondylar hyperplasia described.5,7,9 Whereas in all these cases the clinical presentation involved symptoms and signs of myelopathy, this unique case did not have any such features and is the first to describe facial pain in isolation.

Unilateral trigeminal neuralgia and facial pain has been described rarely as a presentation of Chiari I malformation in a small number of case reports.1,3,11,13–17 It has been more commonly linked to syringobulbia associated with a variety of hindbrain malformations.8 As demonstrated in the imaging in this particular patient, there was no evidence of syringobulbia or hydrocephalus to explain the symptoms of facial pain. The most likely diagnostic possibilities are of disruption to the trigeminal spinal tract fibers, either at the foramen magnum or cervical level. As demonstrated in Fig. 1, there was significant ventrolateral compression of the lower medulla bilaterally at the foramen magnum, which could have resulted in compression of the laterally located trigeminal spinal tract and nucleus. Indeed, this compression was visually evident at surgery and supported by the immediate and complete improvement in pain postoperatively. However, there was no evidence of myelopathy or spinothalamic tract disturbance clinically, which one may have expected with significant lateral brainstem compression.

The other possible location of trigeminal tract disturbance may be within the upper cervical spine related to the extensive syrinx cavity. The subnucleus caudalis located in the upper cervical spine has been linked to neuropathic pain mechanisms in the trigeminal distribution.13 However, this theory is not supported by the clinical or radiological findings, in particular because the symptoms were strictly unilateral with no other sensory modalities involved and the syrinx was midline on imaging (Fig. 1). We therefore suspect that the most likely site of trigeminal tract disruption was lateral compression of the caudal medulla by the hyperplastic occipital condyles. This theory is also more likely to explain the improvement in occipital headaches following surgery.

We opted to perform a dorsal midline decompression to achieve suboccipital decompression of the distorted medulla as well as lateral decompression. Our secondary aim was to maintain the integrity of the atlantoaxial joint and avoid instrumented fixation if possible. Preoperative assessment of the VA anatomy demonstrated a safe dor-
operative trajectory and could be used in conjunction with image guidance to reduce the chance of vascular injury. This approach has proved successful in preserving atlantooccipital joint function over the 5-year follow-up as well as improving the spinal canal CSF dynamics.

Occipitocondylar hyperplasia needs to be distinguished from the equally rare condylus occipitalis (median occipital condyle) because this condition may also present with myelopathy secondary to ventral compression. We could identify 3 reported cases of condylus occipitalis and 1 postmortem finding in the published literature. In all of the live cases described, the clinical presentation was of cervical myelopathy, and in the majority of cases this was associated with atlantoaxial instability.

In the condition of condylus occipitalis the compressive bony lesion is completely midline, unlike occipitocondylar hyperplasia, which is ventrolateral as shown above. There also appears to be a high association with transverse ligament insufficiency in this condition, which is not apparent in the few reported cases of occipitocondylar hyperplasia. In both conditions, however, the majority of authors describe a posterior approach to relieve cervicomedullary compression, although fusion was required in the majority of condylus occipitalis cases. In the condylus occipitalis case described by Kotil and Kalayci, an anterior surgical approach was proposed; however, 2 later authors achieved good outcomes by using posterior decompression with fixation. In the only other operative description of occipitocondylar hyperplasia, posterior decompression without fusion was also performed, with a good outcome. The importance of preserving the atlantooccipital joints during surgery to avoid unnecessary fusion is therefore emphasized when faced with this rare condition.

Conclusions

This unusual case demonstrates further the wide spectrum of presentations arising from craniovertebral junction disorders. Early investigative imaging was critical in allowing prompt surgical management prior to the onset of possible myelopathy or permanent neurological sequelae. Preoperative assessment of the VAs and use of image guidance was extremely useful in surgical planning and intraoperative preservation of the atlantooccipital joints. A dorsal midline approach with sparing of the

Fig. 5. Serial axial CT scans obtained on the 3rd postoperative day demonstrating satisfactory removal of the medial condyles without disrupting the atlantooccipital joints.

Fig. 6. A and B: Postoperative MRI studies demonstrating significant reduction in the cervicothoracic syrinx cavity at 3 months postoperatively. C and D: Dynamic radiographs obtained at 3 months demonstrating no evidence of atlantooccipital instability.
atlantooccipital joints has proven successful in preventing spinal instability as well as sustained clinical improvement over long-term follow-up. Regular interval imaging in which MRI and dynamic radiographs are used to identify development of any secondary instability or altered CSF dynamics is also key to managing the extremely rare condition of occipitocondylar hyperplasia.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Drafting the article: Walkden. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Walkden. Administrative/technical/material support: Walkden.

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