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CASE REPORT

Microvascular decompression with partial occipital condylectomy in a case of pediatric spasmodic torticollis

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Abstract

Spasmodic torticollis is a rare, neurologic disorder that is caused by abnormal nerve compression of the 11th cranial nerve by blood vessels or bony protrusions. It is typically treated pharmacologically and, if necessary, with surgical intervention. We report a unique case of spasmodic torticollis in a 15-year-old female that involved abnormal compression of the left 11th cranial nerve (CN) by the left vertebral artery, displaced by a hypertrophic left occipital condyle. After treatment with Botox was unsuccessful, the patient was treated with microvascular decompression and occipital condylectomy that adequately relieved the abnormal compression of CN XI. Mild symptoms persisted, and the patient underwent a partial section of the sternocleidomastoid muscle 1 year later, after which torticollis symptoms resolved.

Keywords Spasmodic torticollis · Torticollis · Microvascular decompression · Accessory nerve

Introduction

Spasmodic torticollis is a rare, painful disorder that results in involuntary contraction of the neck muscles, causing twisting, tilting, and rotation of the head. This disorder is typically caused by abnormal compression of the 11th cranial nerve (CN XI), also known as the accessory nerve, by an adjacent artery [9]. The accessory nerve supplies the sternocleidomastoid muscle, which tilts and rotates the head, and trapezius muscle, which inserts in the scapula and clavicle. Abnormal compression of the accessory nerve causes involuntary recurrent, transient contraction of these muscles. Treatment typically progresses from physical therapy to Botox (botulinum toxin) injections, and, if symptoms continue, surgical intervention [3]. In this article, we report a case of spasmodic torticollis in which the involvement of the occipital condyle necessitated resection to relieve symptoms. We also present a literature review of similar cases and analyze outcomes based on treatment methods.

Case report

In 2014, a 15-year-old female, who had experienced minor shoulder elevation and torticollis since age eight, presented to our facility. The patient was a dancer and underwent MRI investigation for potential diagnosis of primary segmental dystonia after increased trouble with shoulder elevation. MRI revealed a hypertrophic occipital condyle displacing a slightly dilated left vertebral artery into the CN XI and the lateral medulla (Figs. 1 and 2), with obliteration of the cerebrospinal fluid space (Fig. 1). Based on these results, the patient was found to have spasmodic torticollis, manifesting in rotational distortion of the anterior arch of C1 relative to the occipital condyles (Fig. 2), and treated with a single round of Botox.

The patient's symptoms remained unchanged following Botox treatment. Thus, the patient underwent a modified microvascular decompression that included an occipital condylectomy to remove the protruding bony structure (Fig. 1). We used intraoperative neurophysiological monitoring (IONM) of motor and sensory potentials and lower cranial nerve monitoring. The patient was positioned prone with her

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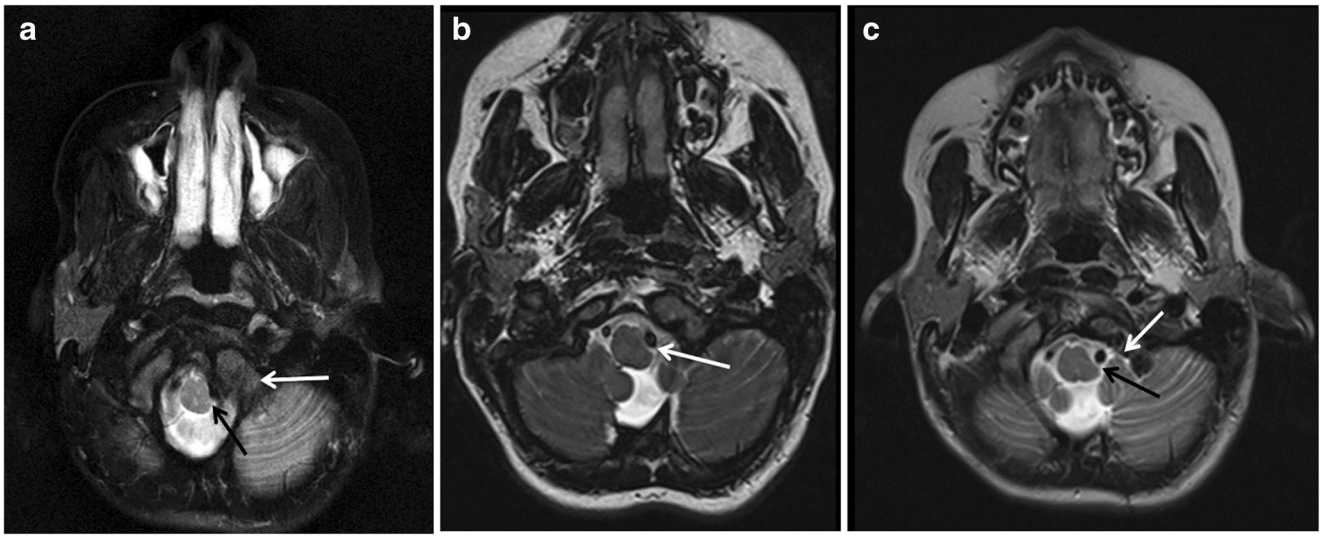

Fig. 1 **a** Axial T2-weighted MRI at the level of the foramen magnum demonstrating obliteration of the cerebrospinal fluid (CSF) space anterolateral to the medulla from an enlarged left occipital condyle (white arrow). The vertebral artery compresses the region of the exiting left spinal accessor nerve (CN11) (black arrow). **b** Axial T2-weighted MRI at the level of the foramen magnum showing more pronounced indentation of the lateral aspect of the medulla as compared with the prior MRI. The dominant left vertebral artery compresses the region of the exiting CN 11 more severely as compared with the prior imaging (arrow), with complete obliteration of the CSF space between the vertebral artery and the medulla. Again noted is the hypertrophic left occipital condyle. **c** Postoperative axial T2-weighted MRI at the level of the foramen magnum shows interval substantial decrease in the size of the left occipital condyle (white arrow). The mass effect of the left vertebral artery has been markedly diminished, now with clear CSF space between the medulla and the left vertebral artery (black arrow)

head turned slightly, and we encountered a slightly dilated vertebral artery compressing the CN XI; the artery was repositioned by dividing the arachnoid bands tethering it in place and then using a suture to bring the artery closer to the dura. We then placed Teflon between the artery and nerve in case of gradual return to the initial position. The occipital condyle was also displacing the artery toward the nerve, and by resecting the medial 2/3 of it, space was created to distance it sufficiently from the CN XI. There were no surgical complications.

The patient was discharged 3 days after surgery following immediate improvement in symptoms. At follow-up, the patient had mild persistent torticollis and, therefore, underwent a partial section of the sternocleidomastoid muscle 1 year after the initial operation. At 2-year follow-up, the patient reported complete resolution of symptoms.

Discussion

This case of spasmodic torticollis in a 15-year-old patient demonstrates the successful utilization of a novel therapeutic approach with good clinical outcome. Occipital condylectomy involves removing a portion of the occipital condyle and is

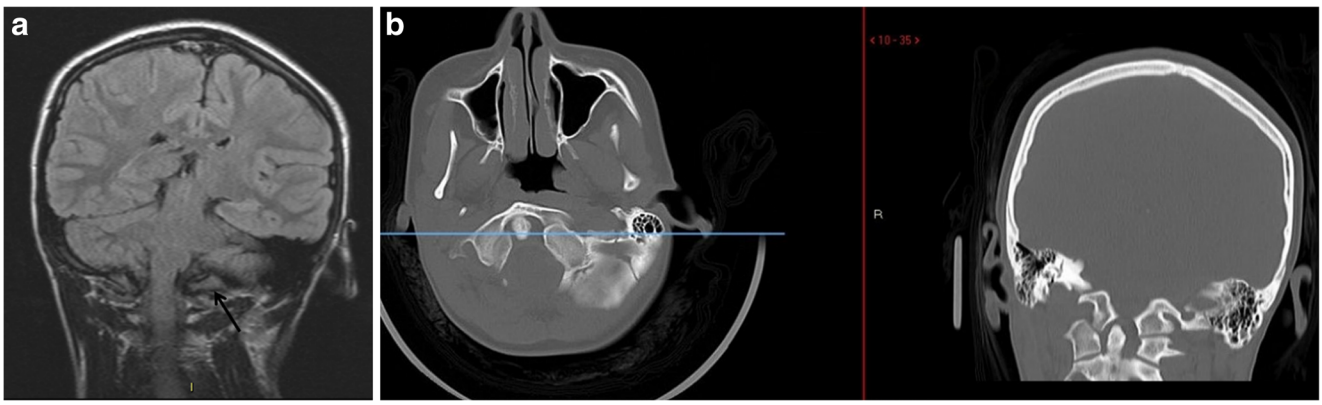

Fig. 2 **a** Coronal FLAIR image showing enlarged left occipital condyle (arrow). **b** Axial and coronal bone images from CT showing typical manifestations of torticollis, including rotational distortion of the anterior arch of C1 relative to the alignment of the occipital condyles. Coronal image shows asymmetry of the spaces between the dens and occipital condyles

traditionally utilized as part of the posterolateral surgical approach for treating intradural ventro-lateral lesions of the cranio-vertebral junction [10]. However, in our case, the patient underwent a partial occipital condylectomy in addition to microvascular decompression to facilitate repositioning of the vertebral artery to successfully relieve abnormal compression of CN XI and its rootlets after Botox injections were unsuccessful. In a literature review of 38 cases of surgically treated spasmodic torticollis, none of the affected patients required an occipital condylectomy or muscle sectioning.

Occipital condylectomy does not come without risks. However, considering the age and pain of the patient and recent biomechanical research that has found no evident clinical instability in patients who underwent removal of 75% of the condyle [6], the risks of treatment were justified.

In a literature review of 38 patients with surgically treated spasmodic torticollis, the pattern of presentation of patients aligned closely with past literature [2], with most cases occurring in females (21/38; 55%) and the average age at diagnosis was 48.25 years [1, 3–5, 7–9]. Diagnosis was made with a combination of MRI, CT, EMG, and angiography, but not specified on an individual case basis (Electronic Resource 1). Symptoms included a combination of tilting, extension, and rotation, and most patients (28/38; 73.7%) experienced head tilting, followed by head rotation (7/38; 18.4%), head extension (3/38; 7.9%), and shoulder elevation (1/38; 2.6%). In addition, there was one patient (2.6%) with antecollis and one patient (2.6%) had no symptoms listed (Table 1). Only two patients (5.3%) had prior treatment specified before ultimate surgical intervention.

All patients (38/38; 100%) underwent surgical microvascular decompression (MVD), also known as the Jannetta procedure, in which a sponge-like material is placed between the involved artery and nerve to ultimately treat the spasmodic torticollis [4]. Cases involved the following arteries: the posterior inferior cerebellar artery (PICA) alone (12/38; 31.6%), PICA and vertebral artery (VA) (3/38; 7.9%), VA alone (1/38; 2.6%), anterior inferior cerebellar artery (AICA) alone (1/38; 2.6%), or the involved artery was not specified (21/38; 55.3%; see Table 2). The involved nerve was the accessory nerve, or CN XI, in 16 (42.1%) cases and was not specified in the remaining cases (22/38; 57.9%; see Electronic Resource 1). Symptoms were resolved in 27 patients (71.1%), improved in 9 patients (23.7%), and did not improve or were unchanged in 2 patients (5.3%). Based on our literature review, microvascular decompression has a good prognosis (36/38; 94.7%) for patients experiencing spasmodic torticollis–induced symptoms.

Table 1 Characterization of cervical dystonia patients based on clinical symptomology

Symptoms	No. of patients (%)
Head rotation	5 (13.2)
Head tilting; shoulder elevation	1 (2.6)
Head tilting	26 (68.4)
Head tilting, rotation	1 (2.6)
Head extension, rotation	2 (5.3)
Head extension, tilting	1 (2.6)
Antecollis	1 (2.6)
Unlisted	1 (2.6)
Total patients	38 (100)

Table 2 Characterization of cervical dystonia patients based on involved artery

Involved artery	Total patients (%)
Anterior inferior cerebellar artery (AICA)	1 (2.6)
Posterior inferior cerebellar artery (PICA)	12 (31.6)
Posterior inferior cerebellar artery, vertebral artery	3 (7.9)
Vertebral artery	1 (2.6)
Not specified	21 (55.3)
Total patients	38 (100)

Conclusion

Spasmodic torticollis, while not life-threatening, can be painfully debilitating. Medical management, usually Botox injections, should be the first attempted treatment because of its low risk, but this treatment fails to resolve symptoms in most cases. If debilitating symptoms persist, microsurgical decompression has been used safely and with good outcomes. Spasmodic torticollis can present with involvement of the occipital condyle, possibly necessitating partial condylectomy, which can be completed with good outcomes. Our case represents the first reported involvement of the occipital condyle; resection of the involved bone, combined with microsurgical decompression and muscle sectioning, successfully resolved our patient's symptoms.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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