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CASE REPORT

A Case Report on Symptomatic Internal Jugular Venous Compression: Clinical and Radiographic Improvement with Bilateral C1 Tubercle Resection

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ABSTRACT

This case report examines internal jugular venous compression necessitating surgical intervention. The focal point is a 48-year-old female patient with Ehlers-Danlos syndrome, who developed intracranial hypertension due to bilateral compression of the internal jugular veins. The patient's symptomatic presentation, marked by persistent pain and pressure localized to retroorbital and occipital regions, accentuated during orthostatic conditions, propelled the investigation that culminated in a comprehensive evaluation via cranial and cervical computed tomography.

The diagnostic revelation of bilateral compression of the internal jugular veins, primarily attributed to C1 tubercles, prompted an elective surgical intervention in March 2023. The meticulous procedure involved bilateral resection of the C1 tubercles, with subsequent postoperative assessments revealing a notable reduction in intracranial pressure. The immediate intraoperative augmentation in the caliber of the jugular vein post-decompression was corroborated through bilateral computed tomographic venography.

In the broader context of neurosurgical interventions for internal jugular venous compression, this case underscores the promising technique of C1 lateral mass resection for alleviating intracranial hypertension, with potential variations based on the clinical significance ascribed to C1 tubercles and the styloid process. The findings presented in this report contribute to the evolving body of evidence supporting surgical approaches in effectively managing internal jugular venous compression and associated intracranial hypertension, thereby enhancing the understanding and treatment of this challenging condition.

Keywords: Ehlers-Danlos Syndrome, Internal Jugular Vein Compression, Intracranial Hypertension, C1 Tubercles.

Introduction

Internal jugular vein (IJV) compression, presenting with symptomatic manifestations, represents a clinical conundrum necessitating precise surgical interventions to alleviate debilitating symptoms[1-13]. The intricate pathophysiology underlying jugular venous outflow obstruction, and its resulting impact on intracranial pressure dynamics[14,15], underscores the complex nature of this condition. Although the mechanisms remains poorly understood, the impingement on jugular venous outflow may cause position-dependent elevation of intracranial pressure (ICP)[16].

Increased ICP manifests with symptoms including cephalalgia, pulsatile tinnitus, vertiginous episodes, visual perturbations, and alterations in mental status, exerting a considerable impact on the overall patient quality of life[17-22]. Timely diagnosis and efficacious intervention for IJV compression are imperative in mitigating symptomatic manifestations and averting potential complications.

IJV compression by C1 tubercle poses a multifaceted challenge in clinical neurosurgery, necessitating a nuanced understanding of its pathophysiological underpinnings and tailored therapeutic intervention[1-3,5-9]. This case report delves into a unique presentation of symptomatic IJV compression, with a specific focus on the involvement of C1 tubercles.

The objective of this case report is to provide a detailed exploration of the diagnostic trajectory, surgical intervention, and subsequent outcomes, thereby contributing to the evolving comprehension of managing IJV compression. Our investigative approach prioritizes meticulous efforts to establish a clear causal association between C1 tubercles and IJV compression, laying the groundwork for a comprehensive understanding of the subsequent surgical resection procedure. In our patient, the successful mitigation of intracranial pressure and the amelioration of associated symptoms was achieved through bilateral C1 tubercle resection.

Case Description:

The case involves a 48-year-old female with Ehlers-Danlos syndrome and tethered cord, with a surgical history of craniocervical fusion and section of the filum terminale. The individual's medical history included hyperadrenergic postural orthostatic tachycardia syndrome, mast cell activation syndrome, and rheumatoid arthritis.

In January 2022, the patient the patient presented with persistent pain and pressure localized to the retroorbital and occipital regions. The symptomatic presentation demonstrated an exacerbation during orthostatic conditions, namely, when assuming a standing or sitting position. Heightened severity was observed upon awakening in the morning. The patient reported a pronounced sensation of excessive cephalic weight, concomitant with a heightened intensity of pain when in an upright seated or standing posture. Consequently, the patient found it necessary employ a rigid cervical collar to provide functional support in lieu of her continuous discomfort.

Additional symptoms within the clinical presentation included muscle spasms, visual disturbances, photosensitivity, pulsatile tinnitus, vertiginous episodes, and

occurrences of fluid exudation from the auricular and nasal regions during severe episodes. Ocular manifestations, inclusive of dynamic fluctuations in blurred and diplopic vision, were observed intermittently. Notably, comprehensive ocular examination conducted by an ophthalmologist revealed an absence of discernible papilledema or optic disc swelling. Furthermore, the patient reported concomitant occurrences of pulsatile tinnitus, with exacerbation noted during periods of physical exertion and heightened severity that coincided with episodes of pronounced vertigo. Consistent vertigo was reported, albeit with fluctuating intensity across different days.

A computed tomographic venogram (CTV) focusing on the cranial and cervical regions showed bilateral compression and severe stenosis of the IJVs, primarily attributable to C1 tubercles (Fig. 1). The identification of IJV compression in imaging prompted an investigation for intracranial hypertension, leading to subsequent findings of elevated intracranial pressure. Preoperative ICP positional testing was conducted, yielding three sets of measurements in supine, sitting, and standing positions. The initial set revealed ICP values of 25 mmHg, 7 mmHg, and 8 mmHg, respectively. The second set demonstrated ICP values of 26 mmHg, 10 mmHg, and 9 mmHg, while the third set exhibited measurements of 20 mmHg, 6 mmHg, and 6 mmHg in the same respective positions.

The collective analysis of symptoms, CTV findings, and ICP measurements substantiates the recommendation for bilateral C1 tubercle resection. The observed symptomatic presentation, coupled with CTV evidence indicating bilateral IJV compression and an associated elevation in supine ICP surpassing 15 mmHg , underscores the potential utility of surgical intervention. The intent of C1 resection aims to ameliorate IJV caliber and flow, thereby addressing the identified pathophysiological factors contributing to the observed clinical manifestations.

In March 2023, the patient underwent an elective surgical resection of the C1 tubercles. Following general anesthesia and endotracheal intubation, the patient was repositioned in the prone orientation, facilitated by bolsters and a pin head holder. After preoperative preparation and draping in adherence to established protocols, the previously employed incision site was infiltrated with a local anesthetic and subsequently incised sharply using a surgical blade. Following a subperiosteal dissection, the procedure involved a comprehensive dissection of the craniocervical fusion site, along with the associated craniocervical fusion hardware. The subperiosteal dissection was then bilaterally extended beyond the confines of the fusion bars, exposing the C1 posterior elements. This extension included the entirety of the transverse foramina and the bilateral C1 tubercles. Following additional dissection, the patient was positioned under the microscope, enabling the complete bilateral excision of the tubercles extending to the transverse foramina. This approach afforded the thorough exposure of the vertebral artery and facilitated the comprehensive decompression of the IJVs bilaterally (Fig. 2). Prior to extubation, an ICP bolt was placed (Fig. 3). The patient was subsequently transferred to the recovery room in stable neurological condition.

Postoperative ICP assessments were conducted in the supine position, with the

head of the bed elevated to zero degrees, during a state of endotracheal intubation under positive pressure ventilation. The postoperative ICP in supine position was recorded at 14 mmHg, denoting a net reduction of 10 mmHg in comparison to preoperative baseline values. An immediate intraoperative augmentation in the caliber of the jugular vein was observed following the surgical decompression procedure. The extent of C1 tubercle resection and surgical decompression of the IJV was confirmed with a post-operative CTV. Subsequently, the patient was discharged from the hospital successfully on postoperative day 4, manifesting an uneventful recovery period following the surgical intervention. Following the osseous decompression, the patient exhibited resolution of headaches, vision, and ear-related manifestations.

Discussion:

Compression of the IJV, resulting in intracranial hypertension, is a multifaceted phenomenon necessitating meticulous assessment[5,11,17,18,23] and judicious therapeutic intervention[1-13,24-26]. IJV compression can arise from several etiologies including bony anatomic compression, neoplasms, or other pathological processes[1,4-6,9]. In instances of IJV compression, impediment of physiological venous drainage from the brain may cause elevated intracranial pressure[27,28] and associated neurological symptoms[18,20,29].

Within the domain of neurosurgical intervention for intracranial hypertension, the prospect of alleviating this condition through IJV decompression via C1 lateral mass resection presents as a promising and innovative technique[1-3,5-9]. For cases where preoperative imaging shows impingment of the IJV by the C1 tubercle, our findings underscore that re-expansion of a compressed IJV can be achieved when the entire C1 tubercle is resected laterally to the transverse foramen. In many cases, IJV compression occurs as the vein passes between the C1 tubercle and the styloid process; the decision to excise either the C1 tubercle exclusively[1-3] or both the C1 tubercle and the styloid process[5-9] depends on the relational anatomy of these two anatomical structures. Pre-operative CTV and/or catheter angiography/venography is necessary to evalute the structural and functional anatomy of the IJV[11,17]. In situations where the jugular vein exhibits evidence of injury and restricted postoperative expansion, our practice is to perform styloid resection to mitigate potential sources of compression. In rare occurrences characterized by a medially positioned jugular vein and perceived inefficacy of C1 resection, isolated styloid process resection may be contemplated as a potential intervention (N.Higgins, personal communication, January 28th, 2023).

Furthermore, we believe it is important to consider the influence of head positioning on jugular vein compression. The IJV serve as the dominant conduit for craniocervical venous drainage in the supine position, but changes in head position have been shown to substantially alter IJV caliber, flow, and function[30,31] . While this facet is not routinely scrutinized, its significance emerges notably in patients manifesting symptoms that occur with specific head positions, as seen in our presented case. In cases of postion-dependent IJV compression, the styloid process may constitute a contributory factor

to compression, a manifestation that may elude detection when patients are subjected to imaging examinations in the supine resting position. In such cases, pre-operative imaging with symptom-provoking head positioning (e.g. extension, lateral rotation) may also be useful in identifying dynamic compression, which may otherwise be undetected in conventional neutral-supine imaging[5]. It is crucial to underscore that the delineated recommendations are grounded in our existing clinical practices and may exhibit variability contingent upon the distinctive characteristics of individual patients.

Limitations and challenges in diagnosing internal IJV compression by C1 tubercle present intricate considerations. Accurate diagnosis relies on a comprehensive evaluation, encompassing clinical presentation, radiographic imaging, and, in some cases, dynamic assessments with position-dependent imaging (Figure 4). Challenges arise in distinguishing symptomatic cases from anatomical variants that may not necessarily contribute to venous compression. Moreover, the dynamic nature of IJV compression, influenced by head positioning, requires meticulous examination, often necessitating specialized imaging protocols. The rarity of this condition further complicates the establishment of standardized diagnostic criteria. Furthermore, differentiating compression-related symptoms from those arising due to other etiologies demands a nuanced approach. While surgical intervention serves as a definitive diagnostic tool and therapeutic measure, identifying cases where surgery might be deferred due to minimal symptomatology or mitigating factors constitutes an essential aspect of the diagnostic challenge. These considerations underscore the need for a individualized approach to the diagnosis of symptomatic IJV compression.

Figure 1. 3D CTV reconstructions showing the compression of the IJV by the ipsilateral C1 tubercle.

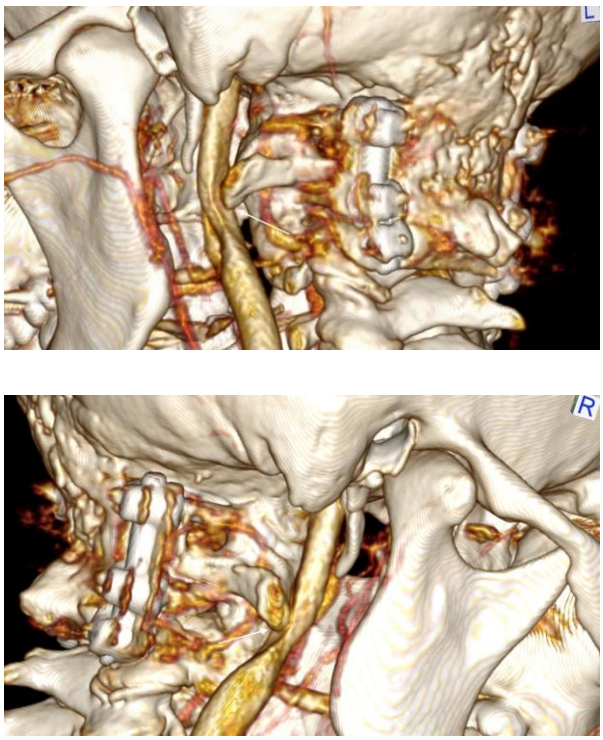

Figure 2. Complete resection of the C1 tubercle exposing the vertebral artery and the IJV (Botton right: C0 pedicle screw, Bottom left: C1 lateral mass screw, Middle across: Dissected vertebral artery, Middle top: Decompressed IJV)

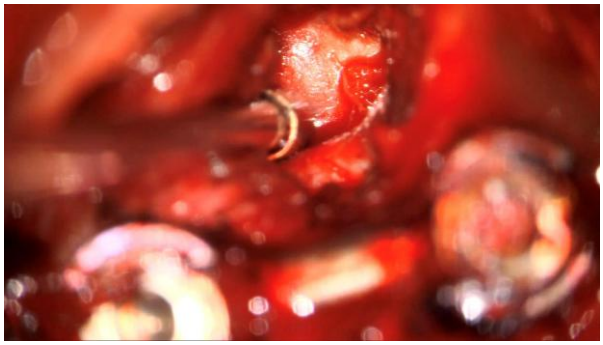

Fig 3. ICP bolt in the supine position, with the head of the bed elevated to zero degrees, during a state of endotracheal intubation under positive pressure ventilation.

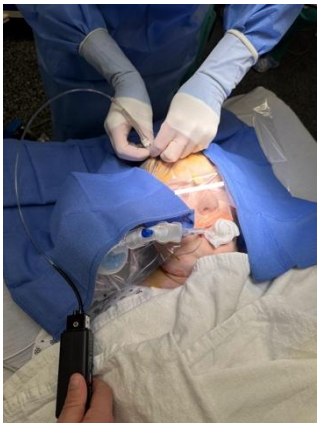

Figure 4: Chronological diagnostic algorithm for internal jugular venous compression attributed to C1 tubercle.

Determining the optimal timing for surgical intervention in cases of IJV compression necessitates a judicious approach. Surgery may be deferred in instances where the symptomatic presentation is minimal, and the impact on the patient's quality of life is limited. Additionally, the decision to defer surgery could be influenced by mitigating factors, such as comorbidities, patient preferences, or the absence of progressive symptoms. Comprehensive preoperative assessment, including dynamic imaging and thorough consideration of clinical symptoms, aids in identifying cases where the risks of surgery may outweigh the potential benefits. The dynamic nature of IJV compression,

influenced by head positioning, further emphasizes the importance of capturing the full clinical picture before deciding on surgical intervention. Ultimately, a multidisciplinary team, including neurosurgeons and specialists familiar with the nuances of IJV compression, can provide valuable insights into determining the optimal timing for surgery or deferring intervention based on individualized patient considerations.

Conclusion:

In conclusion, this case report delves into the complexities of symptomatic IJV compression attributed to C1 tubercles, identified as causative factors for intracranial hypertension. The meticulous evaluation of various therapeutic interventions within the neurosurgical domain, particularly C1 lateral mass resection, highlights a promising avenue for mitigating IJV compression. The multifaceted considerations surrounding the anatomical relationships between C1 tubercles, styloid processes, and the IJV underscore the importance of individualized approaches. Our findings contribute substantively to the evolving body of evidence, shedding light on potential strategies for managing IJV compression and optimizing patient outcomes. Further investigations will be pivotal in refining and validating these strategies for the intricate management of IJV compression.

Conflict of Interest:

None

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Statement of Authorship:

- Paolo A. Bolognese, MD: Dr. Bolognese contributed significantly to the conceptualization of the case report and provided valuable supervision throughout its development.
- Michael Travis Caton, MD: Dr. Caton played an integral role in shaping the conceptual framework of the case report.
- Ilene S. Ruhoy, MD, PhD: Dr. Ruhoy provided essential expertise in the medical review of the case report.
- Jaclyn N. Amaru, MS, PA-C: Ms. Amaru conducted a meticulous retrospective chart review, contributing substantially to the project.
- Sophie Bloom (High School Student): Ms. Bloom actively assisted in retrospective chart review.
- John B. Biggins, PhD: Dr. Biggins made noteworthy contributions to the meticulous preparation of the manuscript.
- Navdeep S. Nayyar, MD, MBA: Dr. Nayyar played a key role in the drafting of the case report.

Disclosures:

None declared

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