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A CASE OF OCCIPITO ATLANTAL GANGLION CYST MIMICKING CYSTIC SCHWANNOMA- A RARE LOCATION

Neurosurgery

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ABSTRACT

Background: Ganglion cysts at the cranio-cervical junction or occipito-atlantal joint represent a rare entity in neurosurgical practice. These benign cystic lesions originate from the joint capsule or tendon sheath. Within the spine, they are predominantly located in the lumbar region (88–99%), with less frequent occurrences in the thoracic spine (up to 8%) and the cervical spine (1%–4%). To date, no reported case has described a ganglion cyst at the occipito-atlantal joint extending into the hypoglossal canal with intradural extension, making this case exceptionally rare. **Case-description:** A 67-year-old male presented with rightward deviation of the tongue for 1.5 years, leg stiffness for six months, and tingling sensations in both upper and lower limbs for one month. Examination revealed tongue atrophy on the right with deviation on protrusion, increased tone in the left lower limb, and bilateral plantar flexion. MRI revealed a well-demarcated multiloculated cystic lesion, hyperintense on T2 and isointense on T1 sequences, with peripheral contrast enhancement. The lesion was located in the foramen magnum, extending into the left hpoglossal canal and intradurally into the foramen magnum, compressing the cervicomedullary junction. Surgical excision via a posterior midline suboccipital approach led to resolution of symptoms and no remaining neurological deficits. **Conclusion:** Given the progressive yet reversible nature of symptoms caused by cervical ganglion cysts, surgical removal is the preferred treatment in cases presenting with neurological compromise. Selection of the surgical approach should aim to minimize invasiveness while considering the patient's symptoms, spinal stability, and lesion characteristics such as size and location.

KEYWORDS

Ganglion cyst, occipito-atlantal joint, hypoglossal canal, cervicomedullary junction

INTRODUCTION

Ganglion cysts are degenerative, cystic lesions originating from joint tissue that can cause mass effect and myelopathy. These lesions are most commonly located in the lumbar spine (88–99%), followed by the thoracic spine (up to 8%) and the cervical spine (1–4%).[1] Approximately two-thirds of cervical ganglion cysts are found below the C2 level, with up to 40% occurring at the atlantoaxial joint.[2] To our knowledge, there have been no previously reported cases of ganglion cysts at the occipito-atlantal junction.

Surgical management options include posterior approaches (such as laminectomy and suboccipital craniectomy) and anterior approaches (notably the transoral route). The anterior approach presents challenges due to its proximity to the airway and its potential to compromise spinal stability. Conversely, the posterior approach may offer limited surgical exposure, which can lead to incomplete or subtotal lesion resection.

We report our first case of an occipito-atlantal ganglion cyst with extension into the hypoglossal canal and intradural involvement. The patient was treated via a midline suboccipital craniotomy, foramen magnum decompression, excision of the posterior arch of C1, and complete resection of both the intradural and extradural components of the cyst.

Case Report

A 67-year-old man presented to us with complaints of deviation of tongue to the right (1 and half years), stiffness of both legs (6months) and paraesthesia of all 4 limbs (1 month). Examination revealed tongue atrophy on the right side with deviation to right on protrusion(LMN type), hypertonia of left lower limb and bilateral plantar extensors. Power in muscle groups of upper and lower extremities were 5/5. There were no bowel or bladder symptoms.

Magnetic resonance imaging (MRI) of Brain (Fig1) showed a well defined T2 hyperintense and T1 isointense multiloculated cystic lesion with peripheral rim of contrast enhancement in the foramen magnum region extending intracranially into right hypoglossal canal and intra-durally into the foramen magnum compressing cervicomedullary junction. Intraoperatively (Fig2A), a cystic lesion at the cervico-medullary junction noted on the right side with some solid component extending through the dura as an extradural component. Cyst was compressing the cervico-medullary junction. The solid component was fibrous and moderately vascular and it was abutting the right vertebral artery. The wall of the cyst was extending upto the hypoglossal canal which was drilled and removed.

Post operatively, spasticity improved significantly.

Histopathology (Fig 2B) showed mucoid/ myxoid material with bland appearing spindle cells floating in the mucoid material consistent with ganglion cells suggestive of Ganglion cyst.

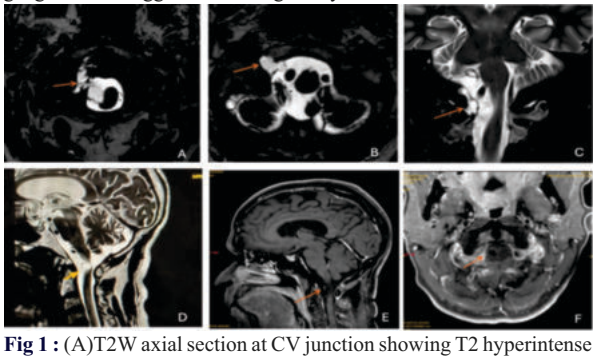

Fig 1 : (A)T2W axial section at CV junction showing T2 hyperintense cystic intradural lesion pushing the cord to the left and the orange arrow indicates the extradural component, (B) T2DRIVE axial section at CVJ showing the extradural portion of lesion extending into hypoglossal canal, (C) T2MRI coronal section showing the extra axial lesion pushing the cord to the left, (D) MRI T2W mid sagittal section of CVJ showing T2 hyperintense lesion at cervico-medullary junction (yellow arrow), (E) sagittal section and (F) axial section of CVJ post contrast images showing minimal peripheral rim enhancement

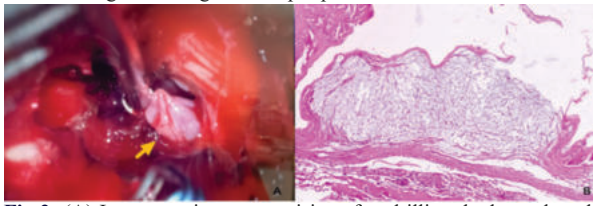

Fig 2: (A) Intraoperative cyst excision after drilling the hypoglossal

canal (B) Hematoxylin and Eosin stained slides showing the cyst wall and a blend of myxoid material and spindle cells

DISCUSSION

Ganglion cysts are benign, cystic lesions that arise from the joint capsule or tendon sheath, and are most commonly observed in the joints of the limbs. Their occurrence in the spine is uncommon, and involvement of the cervical region is particularly rare.[4] Most reported spinal cases are located at the C7–T1 level, with a few near the odontoid process—approximately 50 such cases have been documented. A literature search on PubMed did not reveal any previously reported cases of a ganglion cyst originating from the occipito-atlantal joint, underscoring the exceptional rarity of our case.

The proposed mechanisms underlying the pathogenesis of ganglion cysts include trauma, repetitive microtrauma due to joint instability, and mesenchymal cell proliferation. These cysts are more frequently observed in elderly patients, with a predilection for the L4–L5 level—the most mobile segment of the spinal column. Their common association with spondylolisthesis supports the hypothesis that joint instability plays a key role in cyst development. Accordingly, the leading theory suggests that repeated microtrauma to joint tissue contributes significantly to cyst formation.[5] Additionally, juxtafacet cysts have been reported as a consequence of postoperative instability following spinal surgery.

In our case, there was no history of trauma, rheumatologic disease, facet joint degeneration, or spondylolisthesis. Given the unique biomechanics of the occipito-atlantal joint—distinct from the subaxial cervical spine or lumbar intervertebral joints—it is likely that the etiology in such cases differs from the conventional mechanisms described above.

Cervical juxtafacet cysts typically present as well-defined, hypointense lesions on T1-weighted imaging and hyperintense on T2-weighted sequences, often demonstrating mild rim enhancement following contrast administration. However, the imaging characteristics can vary, potentially due to the presence of hemorrhage, blood products, or calcifications within the cyst. In our case, the lesion demonstrated both an intracanalicular extension into the hypoglossal canal and an intradural component. This necessitated consideration of differential diagnoses, including schwannoma, meningioma, arachnoid cyst, perineural cyst, and epidural abscess. In many instances, definitive diagnosis is established through intraoperative assessment and histopathological analysis.

Surgical excision remains the standard treatment for patients presenting with neurological deficits. In our patient, the lesion was located laterally on the right side. Therefore, a midline suboccipital approach was undertaken, including foramen magnum decompression, excision of the posterior arch of C1, and resection of both the intradural and extradural components of the cyst. The initial extradural approach posed challenges, such as increased traction on nerve roots and bleeding from epidural venous structures. Unfortunately, the displaced vertebral artery was injured during the procedure and had to be clipped and sacrificed.

We subsequently performed a transdural approach to trace and resect the intradural extension of the cyst under direct microscopic visualization of the medulla oblongata and spinal cord, thereby minimizing neural injury. As there was no radiological or intraoperative evidence of instability, spinal fusion was not deemed necessary.

CONCLUSION

Ganglion cysts are more common in the elderly and are generally benign, with low recurrence rates even after partial excision. While conservative treatments—such as bed rest, analgesics, orthoses, or CT-guided aspiration—may be effective for lumbar cysts, they are typically inadequate for cervical cysts due to the risk of progressive myelopathy. Surgical excision is usually necessary to relieve spinal cord compression and restore neurological function. In cases of spinal instability, stabilization may also be required. Therefore, the surgical approach should be tailored to minimize invasiveness while considering clinical presentation, cyst location, size, and spinal stability.

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