# **Case Report**

# Unusual association of cervical arachnoid cyst and idiopathic intracranial hypertension

Shirin H. Alokayli, MBBS, Sarah A. Maghrabi, MBBS, Fawaz S. Almotairi, MD, PhD, Sherif M. Elwatidy, MD, FRCS-SN.

## ABSTRACT

تعتبر الأكياس العنكبوتية شائعة داخل الجمجمة بالمقارنة مع تواجدها في العمود الفقري، وعادةً ما تكون غير مصحوبة بأعراض. في هذا التقرير، نقدم حالة نادرة تظهر ارتباطًا بين الأكياس العنكبوتية العنقودية وارتفاع ضغط الدم في الجمجمة غير المفسر. المريض، البالغ من العمر 71 عامًا، قدم بشكوى من آلام في كلا الكتفين وآلام في الجزء الأيسر من الجسم، ورغم أن الفحص السريري لم يكشف عن أي مؤشرات سريرية واضحة، فقد أظهرت الصور الاشعاعية وجود كيس عنكبوتي على الجانب الأيسر من الفقرات الجامسة والسادسة. وعلى الرغم من الحاولات الجراحية لاستئصاله الا ان المريض احتاج زراعة تحويلة البروتونية عدة مرات. لاحقا تم تشخيص المريض بارتفاع ضغط الدم في الجمجمة غير المفسر، وقررنا اللجوء إلى إجراء تحويلة بارتفاع ضغط الدم في الجمجمة غير المفسر، وقررنا اللجوء الى إجراء تحويلة بطينية لتخفيف الأعراض. يؤكد هذا التقرير على أهمية التشخيص المبكر لارتفاع ضغط الدم في الجمجمة غير المفسر من خلال إجراء الثقب القطني في بطينية لتخفيف الأعراض. يؤكد هذا التقرير على أممية التشخيص المبكر لارتفاع سغط الدم في الجمجمة غير المفسر من خلال إجراء الثقب القطني في مولاتها الأكياس العنكبوتية العنقودية، والذي يساعد على بدء العلمي وفي مارتما مغط الدم في الجمجمة غير المفسر من خلال إجراء التقب القطني في موالات الأكياس العنكبوتية العنقودية، والذي يساعد على بدء العلاج بمراحل

Arachnoid cysts (ACs) are more commonly seen intracranially rather than intraspinally, with most being asymptomatic. This case report presents a rare association between symptomatic AC and idiopathic intracranial hypertension (IIH). In a 71-year-old man who exhibited long-standing bilateral shoulder pain and severe left brachialgia despite an unremarkable physical examination. Radiologic investigations revealed a left C5-6 cervical arachnoid cyst, and during treatment, the patient was diagnosed with IIH. Surgical excision of the cyst failed, so the patient was treated with a lumbar puncture (LP) shunt that required several revisions. During these revisions, IIH was diagnosed, leading to the insertion of a ventriculoperitoneal (VP) shunt, which improved the symptoms. Early diagnosis of IIH through lumbar puncture in cases of spinal arachnoid cysts allows for earlier treatment with cerebrospinal fluid (CSF) diversion via a VP shunt, reducing repeated hospital admissions and surgical interventions.

#### Neurosciences 2024; Vol. 29 (4): 284-287 doi: 10.17712/nsj.2024.4.20240005

From the Division of Neurosurgery, Department of Surgery, King Saud University, Riyadh, Kingdom of Saudi Arabia.

Received 27th January 2024. Accepted 2th July 2024.

Address correspondence and reprint request to: Dr. Shirin H Alokayli, College of Medicine, King Saud University, Riyadh, Kingdom of Saudi Arabia. E-mail: Shirin.H.Alokayli1@gmail.com ORCID ID: https://orcid.org/0000-0002-5266-6037

Intraspinal arachnoid cysts (ACs) are uncommon compared to other locations in the spine.<sup>1</sup> Most arachnoid cysts are clinically silent and have a benign course; however, some cranial ACs can enlarge, causing neurological symptoms and signs of raised intracranial pressure and local mass effects. On the other hand, some cases of cervical AC can present with upper extremity pain, paresthesia, stiffness, and weakness. Physical examination shows a decrease in tone, power, and reflexes. These symptoms may be unilateral or bilateral.<sup>1,2</sup>

In the case of symptomatic AC, treatment is indicated to prevent further spinal injury. The MRI is the gold standard for identifying lesions and their extent.<sup>3</sup> According to the Nabors classification spinal cysts are classified as either extra- or intradural cysts.<sup>4</sup> The etiology of AC is categorized as idiopathic, posttraumatic, or postinfectious, and in some cases congenital.<sup>5</sup>

In the present case, the patient had a cervical intraspinal arachnoid cyst associated with signs and symptoms of increased intracranial pressure. The literature is devoid of similar cases.

**Case Report.** *Patient information.* In this case report no identifiers were collected, and data were taken from the medical record system.

A 71-year-old male patient, with a medical history including hypertension, dyslipidemia, hypothyroidism, and coronary stent placement a decade ago, in addition

**Disclosure.** The authors declare no conflicting interests, support or funding from any drug company.



to a prior left donor nephrectomy.

*Clinical findings.* The patient initially presented in 2011 with bilateral shoulder and arm pain, which progressed over time and caused neurological symptoms including weakness. After treatment, recurrent symptoms for 3 months included persistent bilateral shoulder and arm pain in 2019 and progressive headache, dizziness, and altered consciousness in 2022 and 2023. These symptoms prompted diagnostic evaluations and therapeutic interventions to address the underlying pathology.

**Diagnostic assessment.** Diagnostic imaging findings revealed the presence of a cervical arachnoid cyst in 2011 which he underwent a surgical evaluation and treatment (Figure 1). Subsequent MRI scan in 2019 showed cyst progression within the cervical spine. With symptoms occurring again imaging studies in 2022 confirmed bilateral subdural collections and features consistent with intracranial hypotension, prompting further diagnostic evaluations and therapeutic



Figure 1 - Preoperative MRI scan T2series, sagittal (A) and axial (B) images showing the arachnoid cyst (white arrows) occupying the left half of the body of C5 and expanding the left C5-6 intervertebral foramen.

interventions. In 2023, a cervical MRI scan revealed progressive enlargement of the arachnoid cyst, guiding to use a different treatment approach.

**Therapeutic intervention.** The patient's treatment involved several interventions over the years. Initially, in 2011, surgical excision of anterior C5 corpectomy, cyst excision and fusion of C4-C6 (Figure 2), followed by placement of lumboperitoneal shunt to treat the persistent postoperative CSF leak from the cervical wound. In 2019, a revision of the lumboperitoneal shunt was performed due to recurrent symptoms and cyst progression (Figure 3). Subsequently, in 2023, a right frontal ventriculoperitoneal shunt with a mediumpressure valve was inserted to address progressive cyst enlargement and alleviate symptoms. This treatment aimed to establish an alternative CSF diversion pathway while minimizing the risk of complications associated with the previous shunt.

*Follow-up and outcomes:* Postoperatively, the patient's clinical condition was marked by an improvement in symptoms, with resolution of headache, dizziness, and alterations in consciousness observed. Subsequent follow-up evaluations, including neurological examinations and radiographic imaging studies, affirmed the stability of neurological function and documented regression of the cervical cyst, further substantiating the efficacy of the therapeutic interventions implemented.

**Discussion.** The pathogenesis of spinal arachnoid cysts (AC) remains unclear, with theories suggesting the involvement of diverticula in the septum posticum or ectopic arachnoid granulations. Nonetheless, many AC cases are idiopathic, lacking an identifiable underlying cause.<sup>2</sup> Various mechanisms have been proposed for AC growth: CSF production from cyst walls containing choroid plexus remnants, an increase in osmotically active proteins in the cystic fluid, selective



Figure 2 - CT scan after C5 corpectomy and C4-6 fusion and fixation with titanium plate and screws (red arrow), sagittal (A) and axial (B) images showing bony changes (yellow arrow) involving the body, pedicle and lamina of C5 vertebra



Figure 3 - MRI scan T2 series, sagittal A) and axial (B) images showing enlargement of the residual AC (white arrow) after ligation of the LP shunt.

Dates	Relevant past medical history and interventions A 71-year-old male patient, with a medical history including hypertension, dyslipidemia, hypothyroidism, and coronary stent placement a decade ago, in addition to a prior left donor nephrectomy.		
2011			
Dates	Summaries from initial and follow-up visits/events	Diagnostic testing	Interventions
2011	Initial visit: Bilateral shoulder and arm pain.	Clinical and radiological assessments reveal cervical arachnoid cyst	Anterior C5 corpectomy, cyst excision, and C4–C6 vertebrae fusion
2011	Postoperative event	MRI: shows CSF leakage	Lumboperitoneal (LP) shunt insertion to manage CSF leakage
2019	Follow-up visit: Recurrence of symptoms (bilateral shoulder and arm pain for 3 months)	MRI: Showed cyst progression	Revision of LP shunt due to elevated CSF opening pressures (>25 cm)
2019	Postoperative: Improvement in symptoms	Postoperative imaging (MRI): Indicated cyst regression	Follow up
2022	Follow-up visit: Recurrent symptoms (headache, dizziness, altered consciousness)	MRI and CT scans: Confirmed subdural collections and intracranial hypotension due to LP overshunting	LP shunt Clamping
	Follow-up visit: Persistent symptoms (shoulder and arm pain, headache, dizziness)	MRI: cyst progression	Insertion of a right frontal ventriculoperitoneal (VP) shunt with a medium-pressure valve under
2023			laparoscopic guidance
2023	Follow-up visit: Significant improvement in symptoms (resolution of headache, dizziness)	Follow-up imaging: Documented regression of the cervical cyst	Follow up
2023	Follow-up visit: Stability in neurological function	N/A	Confirmed success of VP shunt intervention

#### Table 1 - Timeline for events and follow-ups of the patient.

or active transport mechanisms from cyst-lining cells, or mechanical fluid entrapment within the cyst.<sup>6</sup> Most arachnoid cysts are asymptomatic and follow a benign course, although some may enlarge and cause neurological symptoms.<sup>5,6</sup>

In most reported cases, surgical excision of the cyst has been completed, preventing neurological symptoms. However, some studies have noted an association between recurrent AC and the development of idiopathic intracranial hypertension (IIH). The IIH is characterized by increased intracranial pressure without hydrocephalus or mass lesions and with normal CSF composition tested by lumbar puncture.<sup>6-8</sup>

Surgical resection is the treatment for symptomatic arachnoid cysts, whether cranial or spinal. Cervical ACs typically follow a posterior approach via laminectomy, especially for thoracic and lumbar cysts, while some studies have described an anterior approach for cervical ACs.<sup>9</sup> Some AC has resolved spontaneously and has not caused a mass effect symptoms.

For recurrent ACs, a lumboperitoneal (LP) shunt may be used. In our case, the patient initially underwent a cervical corpectomy and cyst excision, complicated by CSF leakage. Literature suggests using a ventriculoperitoneal (VP) shunt rather than an LP shunt in patients with cyst hemorrhages.<sup>3,9</sup>

Our patient experienced repeated LP shunt malfunctions, leading to CSF accumulation and increased pressure within the cervical cyst. Each LP shunt revision revealed elevated CSF opening pressure. Additionally, the patient developed low intracranial pressure symptoms from the LP shunt, necessitating temporary clamping, which he could not tolerate due to rapid pressure build-up in the cyst and subsequent arm symptoms. Consequently, we replaced the LP with a VP shunt, opting for a medium-pressure valve with an anti-siphon mechanism connected to a ventricular catheter in the right frontal horn of the lateral ventricle, minimizing occlusion risk by the choroid plexus and facilitating easy tapping of the frontal horn.

In this case, we documented IIH in association with symptomatic cervical AC, confirmed by consistently elevated CSF opening pressure (>250 mm  $H_2O$ ) during LP shunt revisions. The patient's symptoms persisted post-primary surgery for cervical AC excision and improved only after CSF diversion. The relationship between arachnoid cysts and IIH remains unclear, with insufficiently documented cases to establish causality. Further research is needed to understand the primary pathology and optimal management for patients with both conditions.

*Conclusion.* Clinicians should be aware of the association between symptomatic arachnoid cysts and

IIH, particularly when the symptoms of AC recur with malfunction of the shunt (LP/VP). Early diagnosis of IIH by lumbar puncture, in cases of spinal arachnoid cyst, allows earlier treatment with CSF diversion (preferably VP shunt) and eliminates repeated hospital admissions and surgical interventions. We prefer the insertion of a medium pressure valve through the right frontal horn of the lateral ventricle.

**Acknowledgement.** We would like to thank Scribendi (www.scribendi.com) for English language editing.

### References

- Sadek A-R, Nader-Sepahi A. Spinal Arachnoid Cysts: Presentation, management and pathophysiology. *Clinical Neurology and Neurosurgery* 2019; 180: 87–96.
- Manzo M, D'Urso PI. Craniocervical junction arachnoid cyst. Surgical Neurology International 2020; 11: 34.
- Houlihan LM, Marks C. Cerebrospinal fluid hydrodynamics in arachnoid cyst patients with persistent idiopathic intracranial hypertension: a case series and review. *Surg Neurol Int* 2020;11.

- 4. Abdelhameed E, Morsy AA. Surgical outcome of primary intradural spinal arachnoid cysts: a series of 10 cases. *The Egyptian Journal of Neurology, Psychiatry and Neurosurgery* 2021; 57.
- Cuoco JA, Muthukumar S, Rogers CM, Entwistle JJ, Patel VM, Olasunkanmi AL, et al. Spinal Intradural Arachnoid Cysts in Adults: An Institutional Experience and Literature Review. *Neurosurgery/Neurosurgery Online* 2022; 92: 450-463.
- Ahmed AK, Cohen AR. Intracranial arachnoid cysts. *Child's Nervous System* 2023; 39: 2771-2778.
- 7. Houlihan LM, Marks C. Cerebrospinal fluid hydrodynamics in arachnoid cyst patients with persistent idiopathic intracranial hypertension: A case series and review. *Surgical Neurology International* 2020; 11: 237.
- 8. Barkatullah AF, Leishangthem L, Moss HE. MRI findings as markers of idiopathic intracranial hypertension. *Current Opinion in Neurology* 2020; 34: 75-83.
- Epstein NE. Review/Perspective On the Diagnosis and Surgical Management of Spinal Arachnoid Cysts. Surgical Neurology International 2022; 13: 98.