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Case Report

Rapid Rostral Progression of Tumor-Associated Syringomyelia Associated with Hemorrhage-Induced Fibrous Septation: A Case Report

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Abstract

Syringomyelia is frequently accompanied by intramedullary spinal ependymomas and is generally attributed to cerebrospinal fluid flow obstruction. However, the mechanisms underlying rapid syrinx progression and acute neurological deterioration remain unclear. A 29-year-old male with C6-C7 intramedullary ependymoma and syringomyelia extending to C3 was initially managed conservatively. Two years later, he presented with progressive quadriparesthesia, left-sided weakness, and respiratory dysfunction. Magnetic resonance imaging revealed rapid rostral extension of the syrinx to the medulla and a newly formed septum at the C4 level. Surgery was performed to remove the tumor and decompress the syrinx. Xanthochromic fluid was observed when the syrinx was opened. The tumor was resected via laminoplasty at the level of C3-C7 and midline myelotomy, and two syringo–subarachnoid shunts were placed. Histopathological examination confirmed a World Health Organization grade II ependymoma. The septal tissue showed fibrosis, granulation, and hemosiderin deposition, indicating repeated hemorrhage. Postoperative magnetic resonance imaging revealed near-total tumor resection and marked reduction of the syrinx, accompanied by significant neurological improvement. Repeated intratumoral hemorrhage may induce fibrous septation within tumor-associated syringomyelia, potentially altering intramedullary fluid dynamics. In this case, the septum may have functioned in a check valve–like manner, contributing to rapid rostral progression of the syrinx and neurological deterioration. Septation recognition may have implications for the timing and management of surgery.

Keywords: Spinal Ependymoma; Syringomyelia; Intramedullary Tumor; Septation

Abbreviations

CSF: Cerebrospinal Fluid; MRI: Magnetic Resonance Imaging.

Introduction

Syringomyelia is frequently accompanied by intramedullary spinal cord tumors, particularly ependymomas [1,2]. The prevailing mechanism involves cerebrospinal fluid (CSF) flow obstruction and pressure dissociation across the spinal cord [3-5]. In most cases, syrinx formation progresses gradually and stabilizes after tumor removal [1,2].

However, rapid rostral extension of tumor-associated syringomyelia, leading to acute neurological deterioration, is uncommon. The roles of intralesional hemorrhage and secondary structural remodeling within the syrinx cavity have not been fully elucidated. Herein, we report a case in which a newly formed fibrous septation within the syrinx appeared to contribute to rapid rostral propagation of the syrinx toward the medulla and respiratory dysfunction.

Case Presentation

The patient was a 29-year-old male who initially developed numbness in the left upper extremity two years prior to presentation. Cervical magnetic resonance imaging (MRI) revealed an intramedullary tumor at the C6-C7 level with associated syringomyelia extending rostrally to the C3 level (Figure 1). Despite these findings, the patient refused further evaluations and treatments. A chronological summary of the clinical course is as follows: initial diagnosis with C6–C7 intramedullary tumor and syrinx extending to C3, two-year period without treatment, followed by rapid neurological deterioration and rostral syrinx extension to the medulla without significant tumor enlargement.

Over the subsequent two years, the patient developed progressive numbness in all four extremities, weakness of the left upper and lower extremities, and respiratory dysfunction. The patient was referred to our department for surgical management. Magnetic resonance imaging (MRI) revealed no significant tumor enlargement; however, marked rostral extension of the syrinx into the medulla oblongata was observed (Figure 2). MRI was performed using a 3.0-T system with T1-weighted, T2-weighted, and contrast-enhanced sequences. The septation was identified as a low-signal linear structure within the syrinx cavity on T2-weighted images. Furthermore, at the C4 level, a new thick septum was observed within the syringomyelia (Figure 2). We considered that the worsening of the neurological symptoms resulted from the aggressive expansion of the syrinx toward the medulla oblongata. The medical history included chronic kidney disease and hypertension. Surgery was performed via C3–C7 laminoplasty using the unilateral, open-door technique. The tumor was microsurgically resected with motor-evoked potential monitoring. First, a posterior midline sulcal incision was made at the C5 level to access the syrinx. The syrinx was drained to decompress the spinal cord. The contents were xanthochromic. We further extended the posterior midline incision to the C7 level and resected the tumor (Figure 3A). The tumor was soft and prone to bleeding; therefore, we proceeded with careful dissection and removal of the tumor from the spinal cord. The anterior surface of the tumor was highly adherent to the spinal cord, resulting in residual tumor tissue. Furthermore, we opened the posterior midline sulcus up to the C3 level and identified the thickened septum (Figure 3B), and removed it as completely as possible to improve CSF flow. Given the extensive rostral extension of the syrinx and the risk of isolation of the rostral and caudal compartments due to fibrous septation, two syringo–subarachnoid shunts were placed to ensure adequate bidirectional drainage (Figure 3C).

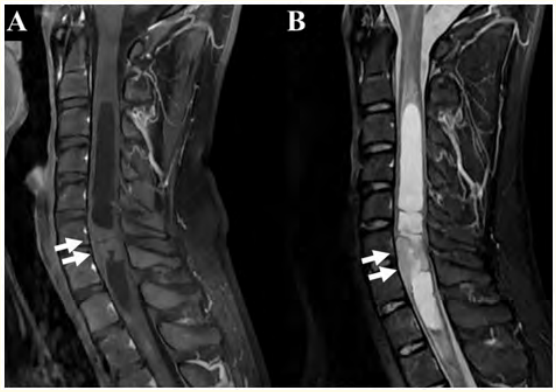

Figure 1: Initial MRI findings at first presentation.

(A) Sagittal T1-weighted image demonstrating an intramedullary lesion at the C6–C7 level (double arrows).

(B) Sagittal T2-weighted image showing an associated syrinx extending rostrally from the tumor to the upper cervical spinal cord. The tumor appears to act as a focal site of cerebrospinal fluid (CSF) flow disturbance.

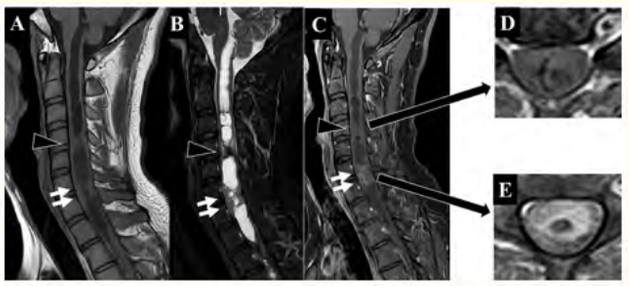

Figure 2: Rapid progression of syringomyelia with newly developed intraluminal septation.

(A) Sagittal T1-weighted image demonstrating the intramedullary tumor at the C6–C7 level (double arrows) and the newly developed intraluminal septation (arrowhead).

(B) Sagittal T2-weighted image showing marked rostral expansion of the syrinx cavity with significant spinal cord distension.

(C) Contrast-enhanced T1-weighted image demonstrating heterogeneous enhancement of the tumor (double arrows).

(D) Axial image at the level of the newly developed intraluminal septation (arrowhead), which was not observed on prior imaging, demonstrating compartmentalization within the syrinx cavity.

(E) Axial image at the tumor level (double arrows), showing the enhancing intramedullary mass and associated cord expansion.

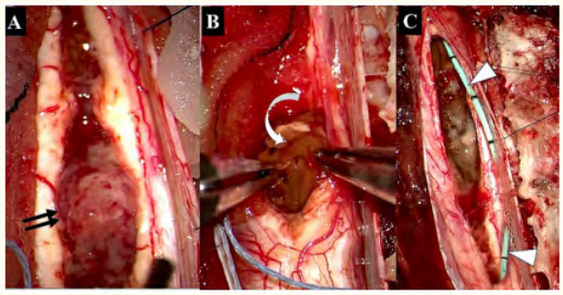

Figure 3: Intraoperative findings supporting a check-valve mechanism.

(A) After midline myelotomy, a well-demarcated intramedullary tumor is identified (double arrows).

(B) Upon opening the syrinx cavity, xanthochromic fluid and fibrous septation are observed (curved arrow), indicating prior hemorrhage and subsequent organization.

(C) Following tumor removal, syringo-subarachnoid shunt (SS shunt) (arrowheads) is are placed to confirm intraluminal continuity and decompression.

Histopathological findings

Hematoxylin and eosin (H&E) staining of the tumor demonstrated a moderately cellular glial neoplasm composed of uniform cells with round to oval nuclei and perivascular pseudorosette formation (Figure 4 A). Immunohistochemically, tumor cells showed a characteristic dot-like perinuclear staining pattern for epithelial membrane antigen (EMA) (Figure 4 B), were negative for Olig2 (Figure 4 C), and retained nuclear expression of H3K27me3 (Figure 4 D). In contrast, the resected septal tissue exhibited dense fibrosis, granulation tissue, and hemosiderin deposition, accompanied by inflammatory cell infiltration, without evidence of neoplastic proliferation (Figure 4 E, F). No histological continuity between the tumor and the septal structure was identified.

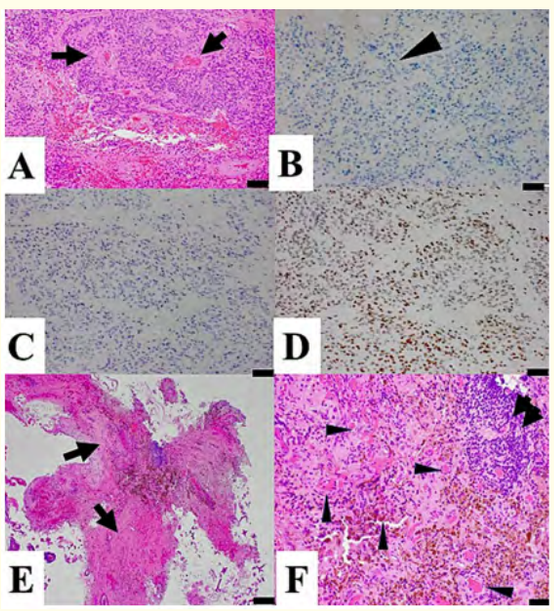

Figure 4: Histopathological findings of the tumor and non-neoplastic hemorrhage-induced septation.

(A) H&E staining of the tumor shows a moderately cellular glial neoplasm composed of uniform cells with round to oval nuclei and perivascular pseudorosette formation (arrows). (scale bar: 50 um).

(B) Immunohistochemistry for epithelial membrane antigen (EMA) demonstrates a characteristic dot-like perinuclear staining pattern (arrowhead). (scale bar: 100 um).

(C) Olig2 immunostaining is negative in tumor cells.

(D) H3K27me3 expression is retained in tumor nuclei. (scale bar: 100 um).

(E) Low-power view demonstrates a dense fibrous structure (arrows) within the spinal cord. (scale bar: 200 um).

(F) High-power view reveals prominent fibrosis with collagen deposition, neovascularization (arrowheads), and lymphocytic infiltration (double arrows). (scale bar: 50 um).

Postoperative magnetic resonance imaging demonstrated near-total resection of the tumor without definite residual enhancement. The syrinx cavity showed marked collapse with restoration of spinal cord configuration and relief of cord distension (Figure 5). These radiographic findings paralleled the clinical course, with early improvement in respiratory dysfunction and subsequent gradual recovery of motor deficits during rehabilitation.

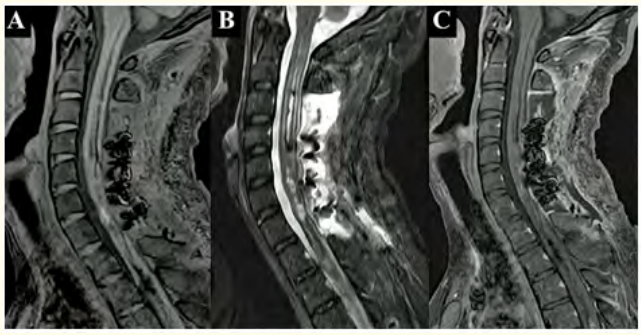

Figure 5: Postoperative cervical magnetic resonance imaging demonstrating surgical outcome.

(A) Sagittal T1-weighted image showing postoperative changes following tumor resection.

(B) Sagittal T2-weighted image demonstrating marked reduction of the syrinx cavity with improvement of spinal cord expansion.

(C) Contrast-enhanced T1-weighted image showing near-total tumor resection without definite residual enhancing lesion.

Discussion

Syringomyelia associated with intramedullary spinal tumors, particularly ependymomas, is commonly attributed to the obstruction of CSF pathways and altered intramedullary pressure dynamics [1-5]. In many cases, the syrinx regresses after tumor removal, supporting a predominantly mechanical mechanism [1,2]. However, the present case demonstrated rapid rostral progression and acute neurological decline that could not be fully explained by static CSF obstruction.

A distinctive feature was the development of a thick septum at the C4 level, which was anatomically separated from the tumor located at the C6-C7. This septum was not present on the MRI obtained two years earlier, indicating an acquired structural change. Intraoperative findings of xanthochromic fluid and histopathological evidence of fibrosis, granulation tissue, and hemosiderin deposition strongly supported repeated intralesional hemorrhage followed by chronic inflammatory remodeling.

Ependymomas are vascular tumors prone to microhemorrhage [6,7]. Biologically active blood degradation products trigger macrophage recruitment, cytokine release, fibroblast activation, and collagen deposition [8,9]. Over time, this cascade may transform a simple fluid cavity into a structurally remodeled compartment with altered compliance. In this context, hemorrhage likely contributed not only to volumetric expansion but also to architectural modification of the syrinx.

The anatomical dissociation between the tumor and septum suggests partial compartmentalization of the syrinx cavity. Incomplete membranes can significantly influence pressure transmission in a pulsatile system driven by cardiac-related CSF oscillations [3,4]. We hypothesized that the septum may have functioned in a check valve–like manner, preferentially permitting rostral pressure propagation while limiting caudal redistribution. Repeated directional pressure loading may have resulted in cumulative headward extension of the medulla oblongata.

This mechanism may explain the rapid neurological deterioration and respiratory dysfunction observed in this patient. The clinical decline appeared to be disproportionate to tumor growth, suggesting that altered fluid dynamics, rather than the mass effect alone, played a central role. Postoperative MRI revealed a marked reduction in the syrinx following tumor removal and shunt placement, accompanied by significant clinical improvement. This radiographic reversibility supported the concept of a dynamically driven process. However, alternative explanations such as subtle tumor progression, changes in CSF dynamics unrelated to septation, or subarachnoid adhesions cannot be completely excluded.

Blood–spinal cord barrier disruption may have further contributed to this. Increased vascular permeability associated with ependymomas can permit the leakage of protein-rich plasma into the cavity [10], potentially elevating oncotic pressure and promoting further fluid accumulation. Therefore, a multifactorial process involving repeated hemorrhage, inflammatory remodeling, altered compliance due to septation, pulsatile pressure asymmetry, and osmotic forces likely underlies the observed rapid progression.

Although tumor enlargement and subarachnoid adhesions remain alternative considerations, the temporal development of septation, pathological evidence of hemorrhage-induced fibrosis, and rapid postoperative regression favor a dynamic intraluminal remodeling mechanism.

This case suggests that tumor-associated syringomyelia should not be regarded as a static fluid collection. Structural changes, such as newly formed septation, may represent markers of evolving instability and the risk of rapid neurological deterioration. Early recognition of these changes may influence surgical decision-making. Although a definitive causal relationship cannot be established in a single case, the temporal evolution of septation, pathological evidence of hemorrhage-induced fibrosis, and rapid postoperative radiographic regression collectively support a dynamic intraluminal remodeling mechanism. This case highlights that structural remodeling within the syrinx, rather than tumor size alone, may play a critical role in rapid neurological deterioration.

Conclusion

Here, we describe a case of cervical ependymoma with rapidly progressive rostral syringomyelia associated with newly acquired fibrous septation. Repeated hemorrhages likely induced septal formation, which may have altered intramedullary fluid dynamics and contributed to rapid neurological deterioration. Understanding the dynamic and potentially reversible nature of tumor-associated syringomyelia may aid in optimizing the timing and management of surgery.

Competing Interests

The authors declare that they have no conflict of interest.

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The authors did not receive support from any organization for the submitted work.

Ethics Approval

All procedures used in this research were approved by the Ethical Committee of International University of Health and Welfare.

Submission Statement

This manuscript is original and has not been submitted elsewhere in part or in whole.

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Bibliography

1. Samii M and Klekamp J. "Surgical results of 100 intramedullary tumors in relation to accompanying syringomyelia". *Neurosurgery* 35.5 (1994): 865-873.

2. Klekamp J. "Treatment of intramedullary tumors: analysis of surgical morbidity and long-term results". *Neurosurgery* 19.1 (2013): 12-22.

3. Williams B. "On the pathogenesis of syringomyelia: a review". *Journal of the Royal Society of Medicine* 73.11 (1980): 798-806.

4. Oldfield EH., *et al*. "Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment". *Journal of Neurosurgery* 80.1 (1994): 3-15.

5. Batzdorf U. "Syringomyelia: current concepts in diagnosis and treatment". Baltimore, Williams and Wilkins, (1991).

6. Sonneland PRL., *et al*. "Myxopapillary ependymoma: a clinicopathologic and immunocytochemical study". *Cancer* 56.4 (1985): 883-893.

7. Terao T., *et al*. "Spontaneous Hemorrhage of a Spinal Ependymoma in the Filum Terminale Presenting with Acute Cauda Equina Syndrome: A Case Report". *NMC Case Report Journal* 3.3 (2016): 91-95.

8. Kumar V., *et al*. "Robbins & Cotran pathologic basis of disease". 9th ed. New York, Elsevier, (2015).

9. Schaer DJ., *et al*. "Hemolysis and free hemoglobin revisited: exploring hemoglobin and heme-mediated toxicity". *Blood, The Journal of the American Society of Hematology* 121. 8 (2013): 1276-1284.

10. Jin L-Y., *et al*. "Blood-Spinal Cord Barrier in Spinal Cord Injury: A Review". Journal of Neurotrauma 38.9 (2021): 1203-1224.