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Primary Extramedullary Cervical Spinal Ependymoma: Case Report and Literature Review

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ABSTRACT

Background: Intradural spinal ependymomas are rare neoplasms derived from ependymal cells that typically arise within the spinal cord parenchyma. Extramedullary localization is exceptionally uncommon and often not considered in the differential diagnosis of spinal tumors. The tanicytic subtype, in particular, represents a distinct histological variant that further complicates clinical and radiological diagnosis.

Objective: This study aims to present a rare case of primary extramedullary cervical spinal ependymoma and to review the current literature on its clinical features, diagnostic strategies, histopathological characteristics, surgical management, and outcomes.

Methods: We conducted a comprehensive literature review and present a detailed case report of a 52-year-old male patient with a primary extramedullary cervical ependymoma. A descriptive research design based on published scientific data was used to contextualize the clinical presentation, diagnostic findings, surgical treatment, and postoperative outcome.

Results: The patient presented with a one-year history of cervical pain and progressive left-sided brachialgia. Magnetic resonance imaging revealed a well-defined, hyperintense intradural extramedullary lesion at the C5–C6 level. Surgical excision was performed under intraoperative neuromonitoring, achieving total removal of the tumor. Histopathological examination confirmed a tanicytic ependymoma (WHO Grade II). Postoperative recovery was uneventful, with complete resolution of radicular symptoms and no neurological deficits.

Conclusion: Primary extramedullary spinal ependymomas are exceedingly rare entities that should be considered in the differential diagnosis of intradural extramedullary lesions. Complete surgical resection remains the gold standard for treatment, often resulting in excellent neurological outcomes. Further studies are required to elucidate the pathogenesis, optimal management, and long-term prognosis of these tumors.

Keywords

Spinal tumors, Ependymoma, Tanicytic ependymoma, Intradural-extramedullary, Cervical spine.

Introduction

Intramedullary spinal cord neoplasms account for approximately 4–10% of all central nervous system (CNS) tumors and 2–4% of glial neoplasms, with ependymomas representing up to 60% of all lesions in this location [1]. These tumors originate from ependymal cells lining the central canal of the spinal cord and are typically confined within the medullary parenchyma. Extramedullary occurrence is exceptionally rare, with the

exception of myxopapillary ependymomas, and is seldom included in the differential diagnosis of spinal extramedullary lesions [2].

The histogenesis of intradural extramedullary ependymomas remains uncertain. It is hypothesized that these tumors may originate from heterotopic ependymal cell remnants trapped during neural tube closure, a theory supported by various clinical and pathological observations [3]. Despite their rarity, extramedullary ependymomas can mimic more common intradural extramedullary lesions such as schwannomas, meningiomas, or even arachnoid cysts, both clinically and radiologically [4].

Ependymomas generally exhibit slow growth and a broad age distribution, with peak incidence occurring between the third and fifth decades of life. Clinical presentation is often insidious, characterized by localized pain, radiculopathy, and occasionally motor or sensory deficits, which can delay diagnosis. Magnetic resonance imaging (MRI) remains the gold-standard diagnostic tool, providing critical anatomical and signal characteristics. However, definitive diagnosis relies on histopathological and immunohistochemical analysis, which typically demonstrates perivascular pseudorosettes, ependymal rosettes, and immunoreactivity for glial fibrillary acidic protein (GFAP) and S100 protein [5].

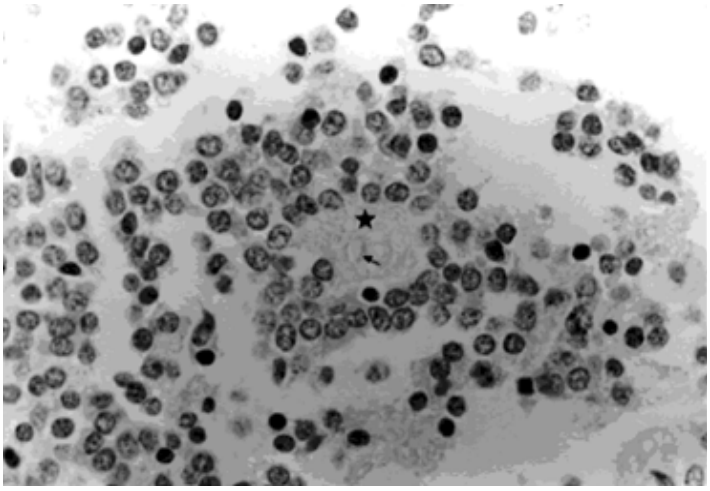

Figure 1: Ependymal rosettes, represented by columnar cells arranged around a central lumen, and perivascular pseudorosettes.

Therapeutic strategies for spinal ependymomas are centered on complete surgical excision, which is associated with favorable outcomes and low recurrence rates. Adjuvant radiotherapy is considered in cases of high-grade (anaplastic) tumors or subtotal resection. Prognosis is largely dependent on the extent of resection and histological grade, with complete excision offering the best chance for long-term disease control [1].

In this paper, we present a rare case of primary extramedullary cervical tanicytic ependymoma and provide a comprehensive review of the current literature regarding its clinical presentation, diagnosis, histopathology, surgical management, and prognosis.

Methods

This study was designed as a descriptive, qualitative investigation combining a comprehensive literature review with a detailed single-patient case report. The objective was to contextualize a rare presentation of primary extramedullary cervical ependymoma within the broader scientific understanding of spinal ependymomas, their clinical features, diagnostic approach, histopathological characteristics, and therapeutic strategies.

Literature Review

A systematic search of the scientific literature was performed using databases such as PubMed, SciELO, and Google Scholar. Keywords included “spinal tumors,” “ependymoma,” “tanicytic ependymoma,” “intradural-extramedullary,” and “cervical spine.” Articles published over the last two decades were prioritized. Relevant data were extracted regarding incidence, clinical presentation, radiological findings, surgical management, histopathology, adjuvant therapy, and outcomes. Special attention was given to previously reported cases of primary intradural extramedullary ependymomas, which remain exceptionally rare.

Case Selection and Data Collection

We report the case of a 52-year-old male patient treated surgically for a primary extramedullary cervical ependymoma at our institution. Clinical records, imaging studies, intraoperative findings, and histopathological reports were reviewed in detail. All patient data were anonymized, and informed consent for the publication of clinical details and images was obtained.

Results-Case Presentation

A 52-year-old male presented with a one-year history of progressive cervical pain and a one-month history of worsening left-sided brachialgia. Neurological examination revealed positive signs of radicular irritation in the left upper limb without motor weakness or clinical evidence of myelopathy.

Radiological Findings

Magnetic resonance imaging (MRI) of the cervical spine revealed a well-circumscribed, oval-shaped intradural extramedullary lesion located posterolaterally to the spinal cord at the C5–C6 level. The lesion appeared hyperintense on both T1- and T2-weighted sequences and was associated with focal spinal cord edema (Figure 2). These findings suggested a benign, slow-growing lesion but did not permit a definitive preoperative differential diagnosis, which included schwannoma, meningioma, and arachnoid cyst.

Surgical Procedure

The patient underwent microsurgical resection under general anesthesia with intraoperative neuromonitoring to preserve neural function. A midline posterior cervical incision was made, followed by limited laminectomies of C5, C6, and C7. Upon durotomy, a well-encapsulated lesion was identified posterolaterally, adherent to the pia mater but without dural or root attachments. The tumor was carefully dissected and removed en bloc without the need for ultrasonic aspiration.

Postoperative Course

The patient's postoperative course was uneventful. He experienced complete resolution of brachialgia and remained free of new neurological deficits. Follow-up MRI confirmed total tumor resection with no residual or recurrent lesion (Figure 3).

Histopathological Findings

Histopathological examination demonstrated numerous perivascular pseudorosettes and ependymal rosettes composed of elongated cells with oval nuclei arranged in a radial pattern (Figure 4). Immunohistochemistry showed focal positivity for glial fibrillary acidic protein (GFAP) and diffuse positivity for S100 protein, consistent with a diagnosis of tanicytic ependymoma (WHO Grade II).

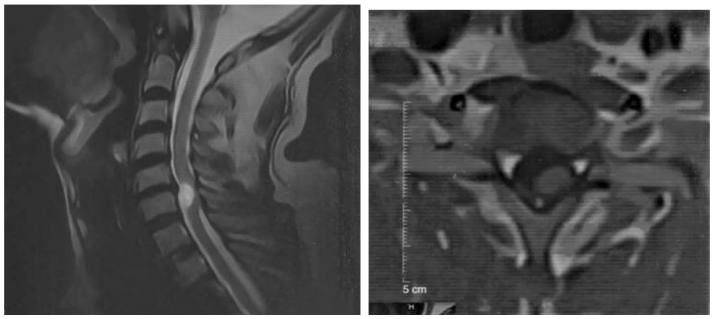

Figure 2: Preoperative magnetic resonance imaging of the cervical spine: (A) sagittal T2-weighted image and (B) axial T1-weighted image showing a hyperintense, oval-shaped intradural extramedullary lesion opposite C5–6, located posterolateral to the spinal cord.

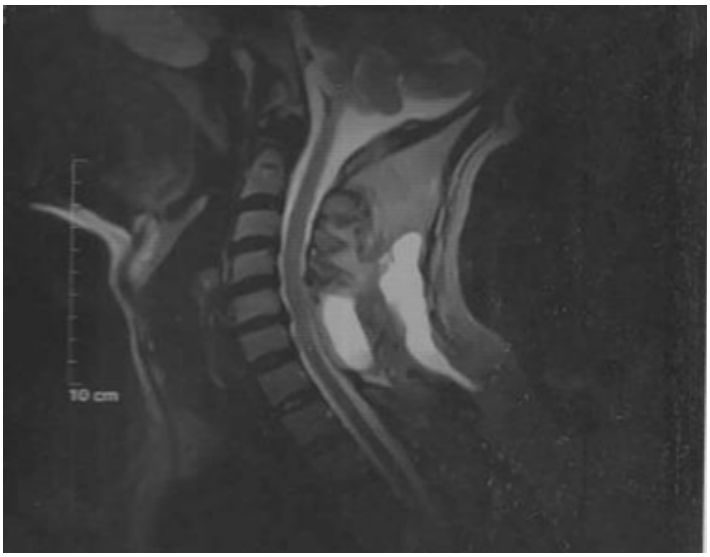

Figure 3: Postoperative MRI of the cervical spine showing complete excision of the lesion with no radiological abnormalities of the spinal cord.

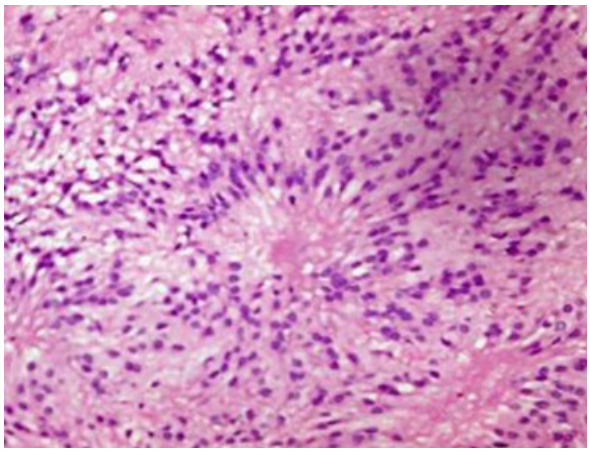

Figure 4: Histological images stained with hematoxylin and eosin: (A) Photomicrograph of a tanycytic ependymoma showing numerous pseudorosettes; H&E ×100. (B) Photomicrograph of a tanycytic ependymoma showing numerous ependymal canals and rosettes; H&E ×100. (C) Photomicrograph of a tanycytic ependymoma showing elongated cells with oval nuclei arranged in a rosette pattern; H&E ×400.

Discussion

Pathogenesis and Histogenesis

Ependymomas are glial neoplasms that originate from ependymal cells lining the ventricular system and central canal of the spinal cord. They account for up to 60% of intramedullary spinal cord tumors in adults [1]. However, their occurrence in an intradural extramedullary location is exceedingly rare, with fewer than 10 primary cases described in the literature to date [4]. The pathogenesis of extramedullary ependymomas remains incompletely understood.

One widely accepted theory suggests that these lesions arise from heterotopic ependymal cell rests displaced from the central canal during neural tube closure. These misplaced cells, which retain their proliferative potential, may later give rise to ependymal tumors in extramedullary locations [3]. Another proposed mechanism involves migration of neoplastic cells from intramedullary origins along perivascular spaces or via subpial extension. The hormonal hypothesis—based on the slightly higher incidence of extramedullary ependymomas in women—remains speculative and requires further investigation [6].

Clinical Presentation and Diagnostic Challenges

The clinical manifestations of extramedullary ependymomas are nonspecific and primarily reflect the tumor's location and mass effect rather than its histological nature. The most common presenting symptoms include localized spinal pain, radiculopathy, paresthesia, and motor deficits. Because of their slow growth and indolent course, symptoms often progress over months or even years, leading to delayed diagnosis [5].

In our case, the patient presented with progressive cervical pain and unilateral brachialgia—symptoms commonly associated

with benign extramedullary lesions such as schwannomas or meningiomas. The absence of myelopathy and lack of specific radiological features further complicated preoperative diagnosis.

Magnetic resonance imaging (MRI) remains the diagnostic modality of choice. Typical imaging findings include isointense or hyperintense signals on T1- and T2-weighted images and homogeneous enhancement after gadolinium administration [7]. However, these findings are not pathognomonic and overlap significantly with those of more common intradural extramedullary tumors. Definitive diagnosis relies on histopathological and immunohistochemical analysis.

Differential Diagnosis

The differential diagnosis of intradural extramedullary spinal lesions primarily includes schwannomas, meningiomas, and less commonly arachnoid cysts or metastatic lesions. Schwannomas typically arise from dorsal sensory roots and may show cystic degeneration or a "dumbbell" configuration extending into the foramina. Meningiomas often exhibit a dural attachment and a "dural tail" sign on MRI. In contrast, extramedullary ependymomas are usually well circumscribed, lack dural attachment, and may exhibit a homogeneous signal pattern [2].

Intraoperatively, the absence of dural or root involvement, combined with a well-encapsulated tumor surface, can help distinguish ependymomas from meningiomas or schwannomas. However, histological confirmation remains essential. Classic pathological features include perivascular pseudorosettes, ependymal rosettes, and immunopositivity for GFAP and S100, as observed in our case.

Surgical Considerations and Management Strategies

Surgical resection remains the gold standard for treatment of spinal ependymomas, including the rare extramedullary variant. The primary goal is gross total resection while preserving neurological function. Most reported cases, including ours, have achieved complete excision with favorable outcomes [1].

Intraoperative neuromonitoring is recommended to minimize neurological morbidity, especially when the tumor is adherent to the pia mater or closely associated with neural elements. In some cases, strong adhesion to the pia may limit the extent of resection, and subtotal excision may be necessary. However, incomplete resection is associated with higher recurrence rates and poorer prognosis [8].

The role of adjuvant radiotherapy remains controversial. It is generally reserved for high-grade (WHO Grade III, anaplastic) ependymomas or cases with residual tumor. Some authors advocate for postoperative radiotherapy even after complete excision of anaplastic lesions to reduce recurrence risk [9]. Chemotherapy has no established role in the management of spinal ependymomas.

Prognosis and Outcomes

The prognosis of extramedullary ependymomas is generally favorable, particularly when gross total resection is achieved. Reported recurrence rates are low, and long-term neurological outcomes are excellent [10]. However, rare cases of malignant transformation and cerebrospinal fluid dissemination have been reported. These findings underscore the importance of long-term follow-up with periodic MRI surveillance.

Our patient experienced complete resolution of radicular symptoms and remained neurologically intact following surgery. No tumor recurrence was detected on postoperative imaging. These outcomes align with previously reported cases in which total excision was associated with excellent prognosis and low recurrence rates.

Conclusion

Primary intradural extramedullary ependymomas of the cervical spine are exceedingly rare entities that pose significant diagnostic and therapeutic challenges. Due to their nonspecific clinical presentation and radiological resemblance to more common spinal lesions such as schwannomas and meningiomas, they are often not considered in the initial differential diagnosis. Histopathological examination remains essential for definitive diagnosis, typically revealing perivascular pseudorosettes, ependymal rosettes, and characteristic immunohistochemical profiles.

Complete microsurgical excision remains the cornerstone of treatment and is associated with excellent neurological outcomes and low recurrence rates. Adjuvant radiotherapy should be considered in cases of subtotal resection or high-grade histology, whereas the role of chemotherapy remains limited.

Given the scarcity of reported cases, long-term follow-up and additional studies are necessary to further elucidate the biological behavior, molecular characteristics, and optimal management strategies for this rare tumor type. Expanding our understanding of extramedullary ependymomas will contribute to improved diagnostic accuracy, surgical planning, and patient outcomes.

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