Published in final edited form as:

World Neurosurg. 2020 July ; 139: 337–342. doi:10.1016/j.wneu.2020.04.095.

Mobile Myxopapillary Ependymoma with Associated Filum Terminale Cyst

Panagiotis Mastorakos, Isaac Jonathan Pomeraniec, Smit Shah, Alireza Shoushtarizadeh, Martha M. Quezado, John Heiss

Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Bethesda, Maryland, USA

Abstract

BACKGROUND: Intradural ependymal cysts are benign, fluid-filled cysts usually situated along the ventral surface of the spinal cord. There are previous reports of 19 intradural cysts in the literature, including one cyst of the filum terminale. Here, we report for the first time the presence of a radiographically occult filum terminale cyst associated with a myxopapillary ependymoma. We propose that mobility of the tumor may provide indirect evidence of the presence of a cyst.

CASE DESCRIPTION: A 65-year-old male patient presented with a homogeneously enhancing ovoid mass measuring 25 mm × 10 mm within the thecal sac at the L3 through L4 levels. Repeat magnetic resonance imaging demonstrated migration of the tumor 12 mm rostrally. Following the L2 through L4 laminectomy and resection of the intradural tumor, we identified a filum terminale ependymal cyst superior to the tumor, which was also resected.

CONCLUSIONS: Ependymal cysts associated with spinal tumors are rare and may be radiographically occult. The change in cyst size may explain tumor mobility. Complete resection of the cyst and histopathologic analysis is recommended to differentiate between ependymal cyst and cystic tumor tissue.

Keywords

Cyst; Filum terminale; Mobile tumor; Myxopapillary ependymoma

INTRODUCTION

Intradural ependymal cysts are rare, benign, fluid-filled cysts that are developmental in origin and may occur throughout the spinal cord. These cysts occur by sequestration of neuroectodermal cells during embryogenesis and are lined with ependymal cells.^{1–3} There are 19 reported cases of intramedullary ependymal cysts,^{1–6} and there is only 1 reported case of an ependymal cyst arising from the filum terminale.⁷

To whom correspondence should be addressed: Panagiotis Mastorakos, M.D., Ph.D., [panos.mastorakos2@nih.gov].

Conflict of interest statement: This study was funded by the National Institute of Neurological Disorders and Stroke of the National Institutes of Health.

Myxopapillary ependymomas (MPEs) are World Health Organization grade I tumors arising from ependymal cells of the filum terminale.⁸ These tumors often are associated with cystic degeneration.⁹ There are no reported cases of ependymal cysts of the filum terminale associated with MPE. A filum terminale cyst, without confirmation of ependymal origin, associated with a paraganglioma of the cauda equina region has previously been reported.⁷

We present a case of a MPE with an associated filum terminale ependymal cyst. The present case demonstrates the prospect of a radiographically occult tumor-associated cyst and further suggests that mobility of the tumor over serial imaging may be an indirect indication of cyst presence. We propose the indication for excisional biopsy of the cyst to rule out underlying neoplastic tissue.

CLINICAL PRESENTATION

A 65-year-old male patient presented with 1 month of progressively worsening low back and bilateral lower-extremity pain that was worse at night. Physical examination revealed decreased range of motion of the right ankle secondary to a remote ankle fracture. The neurologic examination was otherwise without focal deficit. Magnetic resonance imaging (MRI) of the spine demonstrated an intradural, homogeneously enhancing ovoid mass measuring 25 mm × 10 mm at the L3 through L4 levels. The patient was enrolled in our protocol for treatment of neurosurgical diseases and underwent a whole-axis MRI to rule out dissemination of the tumor, given the likely diagnosis of ependymoma. The repeat imaging demonstrated rostral translocation of the tumor (Figure 1). We performed a laminectomy of L3 and L4 with intradural exploration to resect the tumor. The bony dissection was extended to the L2 level in to remove the filum terminale cyst.

Operative Details

After we performed laminectomies at the L3 and L4 levels, intraoperative ultrasonography was performed through the dura, which demonstrated an intradural mass surrounded by cauda equina nerve roots. After we opened the dura, dorsal nerve roots were separated to either side, revealing a dark violet mass attached on either end to the filum terminale. The tumor extended superiorly to the L3 level. A distended, xanthochromic cyst with a translucent wall was discovered in the filum terminale superior to the tumor. The filum was transected rostral and caudal to the tumor. The filum at the rostral end of the tumor was cut between 2 metal clips to prevent leakage of cyst fluid. The tumor was resected en bloc and submitted for frozen section. Intraoperative pathology was consistent with MPE. Following this finding, an L2 laminectomy was performed to expose the superior border of the cyst. The filum terminale proximal of the cyst was cut to release the superior segment of the filum containing the cyst (Video 1).

Postoperative Findings

The patient remained neurologically intact after surgery. Repeat MRI demonstrated complete resection of the tumor (Figure 2A). Grossly we identified a violet mass measuring 2.5 × 1.0 × 0.5 cm with a portion of attached filum terminale (Figure 3A). Histopathologic analysis of tumor tissue was consistent with MPE: strong glial fibrillary

acidic protein positivity composed of epithelioid cells disposed in sheets, papillary structures, and pseudo-rosettes. Microcysts with myxoid material and hyalinized blood vessel walls were identified. The proliferative index was low, with MIB1 staining about 2% of tumor cells (Figure 3B–D). Analysis of the cystic lesion showed a cystic wall lined with cuboidal cells strongly positive for glial fibrillary acidic protein, consistent with an ependymal cyst (Figure 3E–F). The patient continued to make a strong recovery without evidence of symptom or tumor recurrence at 1-year follow-up (Figure 2B).

DISCUSSION

We present a case of a radiographically occult ependymal cyst that was identified intraoperatively during resection of an MPE. Resection and histopathologic analysis of the cyst ensured that the lesion was not neoplastic. This is a rare case that highlights the possibility of identifying a congenital cyst during intrathecal surgery at the level of the cauda equina. We also propose that mobility of a cauda equina lesion between images obtained at different times may provide an indirect indication of presence of a cyst.

This case represents the first report of an ependymal cyst confirmed by histopathology that was associated with a cauda equina ependymoma. Multiple mechanisms have been proposed as to how a cauda equina tumor can induce syringomyelia.¹⁰ Ependymomas of the cauda equina can have a cystic component or associated syringomyelia due to the cavitation of part of the tumor.¹¹ Also, extra-axial tumor may cause cerebrospinal fluid flow block, leading to syringomyelia.¹² In this case, although the filum terminale cyst may have developed as a result of the ependymoma affecting cerebrospinal fluid flow, venous drainage, or edema, the filum terminale cyst was not in direct connection with the ependymoma and the pathologic examination was most consistent with an ependymal cyst, suggesting a separate process.

Ependymal cysts can be found throughout the neuraxis,^{1–6} but there is only one report of an ependymal cyst below the conus medullaris.⁷ In Table 1,^{1–7,13–22} we summarize the reports of spinal ependymal cysts. These lesions are developmental in origin and lined with ependymal cells.^{1–3} The approaches to treatment of these cysts span from simple fenestration or complete resection to placement of a cystosubarachnoid shunt. A filum terminale cyst associated with a paraganglioma in the cauda equina region was previously reported.²² The authors identified a nonenhancing intradural cystic lesion that extended cranially to the tumor on preoperative imaging. Similar to the present case, the cyst was attached, but nonadherent to the conus medullaris and nerve roots, allowing for complete resection.²²

During preoperative evaluation, repeat imaging (MRI) of the lumbar spine after 1 week from the index findings demonstrated no change in tumor growth. However, the tumor had migrated 12 mm in the rostral direction. In retrospect, this finding may be used as an indirect indication of the presence of a cyst. A change in cyst volume may explain the translocation of the tumor within the spinal canal. Previous reports have described mobile tumors of the cauda equina (Table 2).^{23–37} This rare phenomenon is primarily observed in schwannomas^{24,30,36,37} and to a lesser extent in ependymomas.^{23,33,38} The proposed mechanisms include nerve root laxity,^{23,30,37} positional changes,^{23,25,26,33} Valsalva

maneuver,^{30,33} or intrathecal contrast administration.²⁴ We propose that mobility of the cauda equina tumor may constitute an indirect indication of the presence of an ependymal cyst and that change in cyst volume may be associated with translocation of the cyst.

We report a case of a radiographically occult ependymal cyst associated with a MPE. We review the clinical presentation, presumed pathophysiology, and relevant literature. Ependymal cysts associated with spinal tumors are rare and may be radiographically occult. Complete resection of the cyst and histopathologic analysis is recommended to differentiate between ependymal cyst and cystic tumor tissue. We also propose a potential mechanism of cauda equina tumor mobility.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Abbreviations and Acronyms

MPE	Myxopapillary ependymoma
MRI	Magnetic resonance imaging

REFERENCES

1. Iwahashi H, Kawai S, Watabe Y, et al. Spinal intramedullary ependymal cyst: a case report. *Surg Neurol.* 1999;52:357–361. [PubMed: 10555841]
2. Saito K, Morita A, Shibahara J, Kirino T. Spinal intramedullary ependymal cyst: a case report and review of the literature. *Acta Neurochir (Wien).* 2005; 147:443–446 [discussion: 446]. [PubMed: 15666187]
3. Takci E, Sengul G, Keles M. Spinal intramedullary ependymal cyst and tethered cord in an adult. Case report. *J Neurosurg Spine.* 2006;4:506–508. [PubMed: 16776364]
4. Chhabra R, Bansal S, Radotra BD, Mathuriya SN. Recurrent intramedullary cervical ependymal cyst. *Neurol India.* 2003;51:111–113. [PubMed: 12865539]
5. Nagano S, Ijiri K, Kawabata R, et al. Ependymal cyst in the conus medullaris. *J Clin Neurosci.* 2010; 17:272–273. [PubMed: 20036544]
6. Park CH, Hyun SJ, Kim KJ, Kim HJ. Spinal intramedullary ependymal cysts: a case report and review of the literature. *J Korean Neurosurg Soc.* 2012;52:67–70. [PubMed: 22993683]
7. Kato M, Nakamura H, Suzuki E, et al. Ependymal cyst in the lumbar spine associated with cauda equina compression. *J Clin Neurosci.* 2008;15: 827–830. [PubMed: 18407500]
8. Chen X, Li C, Che X, Chen H, Liu Z. Spinal myxopapillary ependymomas: a retrospective clinical and immunohistochemical study. *Acta Neurochir (Wien).* 2016;158:101–107. [PubMed: 26577638]
9. Choudhury M, Rautela A, Nain M. Myxopapillary ependymoma: lesser known cytomorphologic features. *J Cytol.* 2015;32:42–45. [PubMed: 25948946]
10. Barnett HJM, Foster JB, Hodgson P. *Syringomyelia and Tumors of the Nervous System.* London: Saunders; 1973.
11. Nagahiro S, Matsukado Y, Kuratsu J, Saito Y, Takamura S. Syringomyelia and syringobulbia associated with an ependymoma of the cauda equina involving the conus medullaris: case report. *Neurosurgery.* 1986;18:357–360. [PubMed: 3703197]
12. Blaylock RL. Hydrosyringomyelia of the conus medullaris associated with a thoracic meningioma: case report. *J Neurosurg.* 1981;54:833–835. [PubMed: 7241193]
13. Dharker SR, Kanhere S, Dharker RS. Intramedullary epithelial cyst of the spinal cord. *Surg Neurol.* 1979;12:443–444. [PubMed: 524259]

14. Rousseau M, Lesoin F, Combelles G, Krivosic Y, Warot P. An intramedullary ependymal cyst in a 71-year-old woman. *Neurosurgery*. 1983;13:52–54. [PubMed: 6877566]
15. Fortuna A, Mercuri S. Intradural spinal cysts. *Acta Neurochir (Wien)*. 1983;68:289–314. [PubMed: 6880882]
16. Findler G, Hadani M, Tadmor R, Bubis JJ, Shaked I, Sahar A. Spinal intradural ependymal cyst: a case report and review of the literature. *Neurosurgery*. 1985;17:484–486. [PubMed: 4047361]
17. Sharma BS, Banerjee AK, Khosla VK, Kak VK. Congenital intramedullary spinal ependymal cyst. *Surg Neurol*. 1987;27:476–480. [PubMed: 3563862]
18. Pagni CA, Canavero S, Vinattieri A, Forni M. Intramedullary spinal ependymal cyst: case report. *Surg Neurol*. 1991;35:325–328. [PubMed: 2008649]
19. Robertson DP, Kirkpatrick JB, Harper RL, Mawad ME. Spinal intramedullary ependymal cyst. Report of three cases. *J Neurosurg*. 1991;75: 312–316. [PubMed: 2072172]
20. Kumar R, Nayak SR, Krishnani N, Chhabra DK. Spinal intramedullary ependymal cyst. Report of two cases and review of the literature. *Pediatr Neurosurg*. 2001;35:29–34. [PubMed: 11490188]
21. Lalitha AV, Rout P, F DS, Shailesh, Rao S. Spinal intramedullary neuroepithelial (ependymal) cyst. A rare cause of treatable acute para paresis. *Indian J Pediatr*. 2006;73:945–946. [PubMed: 17090911]
22. Adam D, Moisesescu C, Iftimie D, Burdusa G. Filum terminale paraganglioma with associated cyst: a case report. *Rom Neurosurg*. 2018;XXXII:217–223.
23. Wortzman G, Botterell EH. A mobile ependymoma of the filum terminale. *J Neurosurg*. 1963;20: 164–166. [PubMed: 14192087]
24. Tavy DL, Kuiters RR, Koster PA, Hekster RE. Elusive tumor of the cauda equina. Case report. *J Neurosurg*. 1987;66:131–133. [PubMed: 3783245]
25. Isu T, Iwasaki Y, Akino M, Nagashima M, Abe H. Mobile schwannoma of the cauda equina diagnosed by magnetic resonance imaging. *Neurosurgery*. 1989;25:968–971. [PubMed: 2601828]
26. Namura S, Hanakita J, Suwa H, Mizuno M, Ohtsuka T, Asahi M. Thoracic mobile neurinoma. Case report. *J Neurosurg*. 1993;79:277–279. [PubMed: 8331413]
27. Varughese G, Mazagri R. Mobile tumours in the lumbar spinal canal: a diagnostic problem. *Can J Surg*. 1997;40:59–63. [PubMed: 9030087]
28. Iizuka H, Iida T, Akai T. Mobile neurinoma of the cervicothoracic junction. *Surg Neurol*. 1998;50: 492–493. [PubMed: 9842879]
29. Friedman JA, Wetjen NM, Atkinson JL. Utility of intraoperative ultrasound for tumors of the cauda equina. *Spine (Phila Pa 1976)*. 2003;28:288–290 [discussion: 291]. [PubMed: 12567033]
30. Marin-Sanabria EA, Sih IM, Tan KK, Tan JS. Mobile cauda equina schwannomas. *Singapore Med J*. 2007;48:e53–e56. [PubMed: 17304380]
31. Kim SB, Kim HS, Jang JS, Lee SH. Mobility of intradural extramedullary schwannoma at spine: report of three cases with literature review. *J Korean Neurosurg Soc*. 2010;47:64–67. [PubMed: 20157382]
32. Khan RA, Rahman A, Bhandari PB, Khan SI. Double migration of a schwannoma of thoracic spine. *BMJ Case Rep*. 2013;2013 bcr2012008182.
33. Kotani T, Okawa A, Akazawa T, Sakuma T. Mobile ependymoma diagnosed with cine MRI. *BMJ Case Rep*. 2014;2014 bcr2013202984.
34. Terada Y, Toda H, Yokote A, Iwasaki K. A mobile schwannoma of the cervical spinal cord: case report and review of the literature. *Neurosurgery*. 2016;78:E156–E159. [PubMed: 26287552]
35. Toscano DT, Felbaum DR, Ryan JE, Sayah A, Nair MN. Mobile schwannoma of the lumbar spine: a case report and review of the Literature. *Cureus*. 2016;8:e715. [PubMed: 27610287]
36. Onishi FJ, Salu S, Cavalheiro S. Mobile cauda equina schwannomas: how to deal with this rare event and avoid surgical complications. *Surg Neurol Int*. 2018;9:125. [PubMed: 30034916]
37. Hamabe F, Soga S, Imabayashi H, Matsunaga A, Shinmoto H. Mobile spinal schwannoma with a completely cystic appearance. *Am J Case Rep*. 2019; 20:859–863. [PubMed: 31204384]
38. Moon K, Filis AK, Cohen AR. Mobile spinal ependymoma. *J Neurosurg Pediatr*. 2010;5:85–88. [PubMed: 20043741]

Figure 1.

Preoperative magnetic resonance imaging (MRI) of the lumbar spine. Sagittal view (A) T1-weighted noncontrast, (B) T1-weighted postcontrast, and (C) T2-weighted MRI demonstrating an intradural, homogeneously enhancing ovoid mass measuring 25 mm × 10 mm at the L3/L4 level. (D) T2-weighted of repeat MRI 1 week following initial imaging, demonstrating a 12-mm rostral translocation of the tumor.

Figure 2.

Postoperative magnetic resonance imaging (MRI) of the lumbar spine. **(A)** T1-weighted noncontrast, T1-weighted postcontrast, and T2-weighted sagittal postoperative MRI demonstrating complete resection of tumor and cyst. **(B)** T1-weighted noncontrast, T1-weighted postcontrast, and T2-weighted sagittal 1-year follow up MRI demonstrating no evidence of recurrence.

Figure 3.

Histopathologic analysis. (A) Grossly identified violetered mass measuring $2.5 \times 1.0 \times 0.5$ cm with part of the filum terminale attached. (B–C) Hematoxylin and eosin stain of tumor tissue demonstrating epithelioid cells disposed in sheets, papillary structures and pseudo-rosettes. Microcysts with myxoid material and hyalinized blood vessel walls were identified. (D) Glial fibrillary acidic protein (GFAP) stain demonstrating strongly GFAP-positive epithelioid cells. (E) Hematoxylin and eosin stain of cystic lesion demonstrating cystic wall lined with cuboidal cells. (F) GFAP stain of cystic lesion demonstrating strongly GFAP-positive cuboidal cells.

Cases of Ependymal Cysts Reported in the Literature

Table 1.

Study	Age, years/Sex	Level	Location	Symptoms	Surgery	Postoperative Symptoms
Dharker et al., 1979 ¹³	38/F	T6	Intramedullary	Paraparesis, LBP	Partial resection + marsupialization	No improvement
Rousseau et al., 1983 ¹⁴	71/F	T2	Intramedullary	Right-sided abdominal pain	Partial resection + marsupialization	No improvement
Fortuna and Mercuri, 1983 ¹⁵	67/F	T12	Intramedullary	Intermittent paraparesis	Partial resection + marsupialization	Requiring walking-aid
	57/F	T12	Intramedullary	Medullary pain	Partial resection + marsupialization	Symptom resolution
Findler et al., 1985 ¹⁶	6/M	C7–T1	Intramedullary	Spastic paraparesis and numbness	Partial resection + marsupialization	Symptom resolution
Sharma et al., 1987 ¹⁷	7/M	T4	Intramedullary	Paraparesis and urinary incontinence	Complete resection	Symptom resolution
Pagni et al., 1991 ¹⁸	39/M	C5–6	Intramedullary	LUE paresthesias	Complete resection	Requiring walking-aid
Robertson et al., 1991 ¹⁹	48/F	T12–L1	Intramedullary	Paresthesias	Partial resection + marsupialization	Partial improvement
Iwahashi et al., 1999 ¹	18/F	T6	Intramedullary	BLE weakness/myelopathy	Partial resection + marsupialization	Symptom resolution
Kumar et al., 2001 ²⁰	5/M	T4–6	Intramedullary	Gait instability	Complete resolution	Symptom resolution
	4/M	C1–5	Intramedullary	Neck pain + BUE weakness	Partial resection + marsupialization	Partial resolution
Chhabra et al., 2003 ⁴	15/M	C2–3	Intramedullary	Quadriparesis	Partial resection + marsupialization followed by complete resection	Symptom resolution
Saito et al., 2005 ²	44/F	T11–L1	Intramedullary	RLE weakness	Partial resection + marsupialization + shunt	Symptom resolution
Takci et al., 2006 ³	33/M	L3	Intramedullary	Numbness BLE + LBP	Partial resection + marsupialization	Symptom resolution
Lalitha et al., 2006 ²¹	8/F	T2–3	Intramedullary	BLE pain + gait instability	Complete resection	Symptom resolution
Kato et al., 2008 ⁷	31/F	L2–3	Cauda equina	BLE numbness/LBP	Complete resection	Symptom resolution
Nagano et al., 2010 ⁵	64/F	T11–L1	Conus medullaris	BLE pain and numbness	Partial resection + marsupialization	Remaining BLE numbness
Park et al., 2012 ⁶	46/F	T11–12	Intramedullary	LLE paresthesias	Partial resection	Symptom resolution
Adam et al., 2018 ²²	46/F	L3	Conus medullaris	BLE paresthesias	Complete resection	Symptom resolution

F, female; LBP, low back pain; M, male; LUE, left upper extremity; BLE, bilateral lower extremities; BUE, bilateral upper extremities; LLE, left lower extremity.

Table 2.

Cases of Mobile Spine Tumors Reported in the Literature

Study	Age, years/Sex	Location *	Tumor Type	Size	Underlying Etiology
Wortzman and Botterell, 1963 ²³	49/F	L3 → L2	Ependymoma	NA	Postural change
Tavy et al., 1987 ²⁴	NA	Cauda equina	Schwannoma	NA	Not indicated
Isu et al., 1989 ²⁵	51/F	T11–L2 → T12–L1	Schwannoma	NA	Postural change
	52/M	L1 → L1–L2	Schwannoma		
	42/M	T12 → L1	Schwannoma		
Namura et al., 1993 ²⁶	51/M	T4–T5 → T9–T10	Schwannoma	NA	Postural change
Varughese et al., 1997 ²⁷	65/M	L5 → L4	Schwannoma	NA	Redundancy of cauda equina nerve root
	78/M	L3 → L2	Schwannoma		
Iizuka et al., 1998 ²⁸	48/M	C7–T1 → T1–T2	Schwannoma	NA	Elongation of nerve root involved by tension
Friedman et al., 2003 ²⁹	28/M	L4 → L3	Schwannoma	NA	Increased length and mobility of the nerve root involved
	33/M	L5 → L4–L5	Schwannoma	NA	
	41/F	L2 → L1–L2	Schwannoma	NA	
Marin-Sanabria et al., 2007 ³⁰	27/M	L1 → L2	Schwannoma	3 cm × 1 cm × 1 cm	Not indicated
	41/M	L2–L3 → L1–L2	Schwannoma	1.5 cm × 2.7 cm × 1.3 cm	Strained significantly at micturition preoperatively (Valsalva)
Kim et al., 2010 ³¹	45/M	L3–L4 → L2–L3	Schwannoma	1.6 cm × 3.0 cm	Not indicated
	32/M	T10–T11 → T11	Schwannoma	1.0 cm × 1.6 cm × 2.0 cm	
	27/M	L3–L4 → L2–L3	Schwannoma	1.4 cm × 1.2 cm	
Khan et al., 2013 ³²	52/M	T10–T11 → T7–T8	Schwannoma	4.0 cm × 0.75 cm	Dilatation of the subarachnoid space by extramedullary tumor
Kotani et al., 2014 ³³	12/M	L1 → T12	Ependymoma	NA	Postural change
Terada et al., 2016 ³⁴	68/M	C5–C7 → C6–T1	Schwannoma	NA	Not indicated
Toscano et al., 2016 ³⁵	40/M	T11–L2 → T12–L1	Schwannoma	NA	Not indicated
Onishi et al., 2018 ³⁶	49/M	L3 → L4–L5	Schwannoma	3.0 × 1.5 cm	Not indicated
Hamabe et al., 2019 ³⁷	48/F	L4–S1 → L2–L4	Schwannoma	5.1 cm (diameter)	Laxity of nerve roots

F, female; NA, not reported; M, male.

* Indicates the 2 discrete locations identified for each mobile tumor.