

Resection of disseminated recurrent myxopapillary ependymoma with more than 4-year follow-up: operative nuance for prolonged prone position. Illustrative case

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BACKGROUND Symptomatic disseminated myxopapillary ependymoma (MPE) in a young person presents a daunting challenge because the risks of prolonged prone positioning and spinal cord injury may outweigh the likelihood of attaining the benefit of gross total resection.

OBSERVATIONS The authors reported the case of a 15-year-old girl with five discrete recurrent spinal cord ependymomas. The patient received a 25-hour surgical procedure for gross total resection of the tumors and fusion over an approximately 33-hour period. She experienced complete resolution of all preoperative neurological symptoms and subsequently received adjuvant radiation therapy. At 52 months after surgery, she was still experiencing neurologically intact, progression-free survival. This case illustrated one of the most extensive recurrent tumor resections for MPE with prolonged disease-free survival reported to date. It may also represent the longest prone position spinal case reported and was notable for a lack of any of the complications commonly associated with the prolonged prone position.

LESSONS The authors discussed the complexity of surgical decision-making in a symptomatic patient with multiple disseminated metastases, technical considerations for resection of intradural and intramedullary spinal cord tumors, and considerations for avoiding complications during prolonged positioning necessary for spinal surgery.

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KEYWORDS ependymoma; prone surgical positioning; spinal cord; tumor

Myxopapillary ependymomas (MPEs) are rare neoplasms comprising 0.5% of all ependymomas¹ or approximately 13% of ependymomas arising in the spinal cord. MPEs most frequently develop in the lumbosacral region at the filum terminale and conus medullaris and have an annual incidence of 1 per 1 million people in the United States.²

MPEs were formerly classified as World Health Organization (WHO) grade I, but further appreciation for recurrence potential and capacity for dissemination has resulted in reclassification of MPEs as WHO grade II in the most recent WHO Classification of Tumors of the Central Nervous System.³ Most cases of MPE are localized at initial presentation. However, there are reports of disseminated disease at first diagnosis.⁴ When dissemination is present at the time of initial diagnosis without previous surgery, the mechanism for tumor spread is thought to be due to

spontaneous capsule rupture and bleeding.^{5,6} Dissemination is also a recognized potential complication after resection.^{7,8}

Despite absence of definitive guidelines, resection is the first-line treatment for MPE.⁹ Because of the risk of dissemination after resection, gross total resection (GTR) is most desirable and portends the best prognosis.¹⁰ Ideally, GTR is achieved through an en bloc resection,¹¹ but often piecemeal GTR or even subtotal resection (STR) is performed due to inability to safely mobilize tumor from neural structures. Although overall survival does not appear to be impacted by extent of resection,^{11,12} Kraetzig et al.¹⁰ showed a 2.5-fold decrease in recurrence when GTR is achieved compared to STR. If residual tumor cannot be resected, the use of adjuvant radiation therapy (RT) may be considered. Mixed results associated

ABBREVIATIONS EBL = estimated blood loss; FFP = fresh frozen plasma; GTR = gross total resection; LR = lactated Ringer's; MPE = myxopapillary ependymoma; MRI = magnetic resonance imaging; POVL = perioperative vision loss; RBC = red blood cell; STR = subtotal resection; RT = radiation therapy; WHO = World Health Organization.

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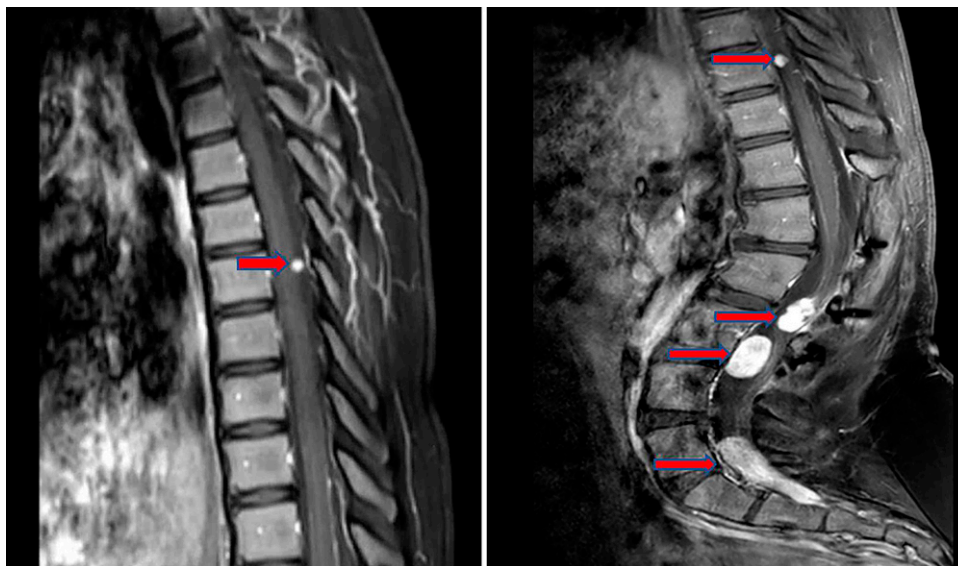


FIG. 1. Preoperative MRI with gadolinium contrast demonstrating five contrast-enhancing lesions (arrows) at the levels of T7 (left), T10, L2, L3, and S1–3 (right) in a 15-year-old girl with a history of MPE resection 3 years earlier who is now presenting with back pain and bilateral lower-extremity paresthesia.

with adjuvant RT may be due to differential outcomes based on patient age¹³ and dose of RT.¹⁴ Agbahiwe et al.¹⁵ found that adjuvant RT yields improved local control compared to surgery alone in a retrospective study of 16 pediatric patients. Several other reports indicate a benefit of adjuvant RT even in adult patients.^{14,16,17} Understanding the potential benefits and limitations of RT is important because this therapy has well-recognized risks. Patients with MPE are likely to achieve long-term survival, especially in the pediatric population, so potential late effects of RT must be considered. Compared to traditional photon radiation, proton radiation may serve as an effective alternative with an improved safety profile in the pediatric population with spinal ependymoma.¹⁸

Recurrent and disseminated MPE further complicates management, and there is a paucity of evidence to guide treatment planning. Here we present a case of recurrent MPE in the form of disseminated disease along the neuraxis and discuss the complex decision-making involved prior to attempting GTR as well as the keys to avoiding complications of prolonged positioning.

Illustrative Case

Initial Presentation

A patient initially presented at 12 years old with symptoms of L2 and L3 radiculopathy. Magnetic resonance imaging (MRI) of the spine demonstrated a single gadolinium-enhancing intradural lesion that was radiographically consistent with an ependymoma. The patient received an uncomplicated piecemeal tumor resection and laminoplasty at the L1–3 levels with a different surgeon. The diagnosis of MPE was confirmed on analysis of specimens obtained at the time of surgery. No further adjuvant treatment was used at that time.

Disseminated Recurrence

Three years after the patient's initial surgical procedure, when she was 15 years old, she returned to our clinic with reports of back pain. The patient endorsed worsening of her back pain at night and during

flexion of the spine while tying her shoes. Furthermore, she noted bilateral lower extremity paresthesias. She also reported difficulty with ascending stairs. Physical examination demonstrated full strength in the bilateral lower extremities. The patient had patchy paresthesia throughout the lower extremities and diminished reflexes at the patella and Achilles tendon. Standing radiographs demonstrated mild thoracolumbar kyphosis. Gadolinium-enhanced MRI demonstrated five intradural tumors at T7, T10, L2, L3, and S1–3, suggestive of disseminated recurrence of her prior MPE (Fig. 1, right). We conducted extensive discussion with the patient and her parents regarding surgical and nonsurgical options. The low likelihood of prolonged disease-free survival was discussed. The patient and her family elected to proceed with a staged attempt at resection followed by fusion to address the thoracolumbar kyphosis.

Procedure

Extensive preoperative planning was carried out to facilitate GTR of the lesions and reduce the risk of complications, especially given the length of the procedure and known risks of prolonged prone positioning.^{19,20} The patient was placed prone on a Jackson table with Wilson attachment on the first day and without the Wilson frame to attain improved lordosis on the second day. Her head was maintained in a Mayfield to avoid pressure on the cranium. Neuro-monitoring of both upper and lower extremities was performed to avoid neurapraxia. GTR of all five tumors and fusion from T7 to the pelvis was accomplished in a 2-day staged procedure. Removal of all tumors was accomplished with laminectomy, opening of the dura at the midline, and GTR in a piecemeal fashion. Direct stimulation of nerve roots was used during resection of tumor that had infiltrated the nerve roots, particularly in the S1–3 area, where the nerve sheaths had to be opened to remove tumor. Sharp dissection without cautery was performed to minimize thermal injury. An MRI was performed intraoperatively after tumor resection on day 1 (Fig. 2, left) and prior to fusion (Fig. 2, right). Lastly, neuronavigation was used to perform an instrumented fusion from T7 to the sacrum (Fig. 3). In total, the



FIG. 2. Postoperative MRI demonstrating complete resection of the five intradural lesions immediately after resection on operative day 1 (left) and prior to starting fusion on operative day 2 (right).

patient was in the prone position from 8:45 AM on operative day 1 until 11:15 PM and then from 7:30 AM to 6:00 PM for day 2 of the staged procedure (a cumulative operative time of 25 hours in a 33.25-hour period). The total time for anesthesia was 28 hours.

Outcome and Adjuvant Therapy

Postoperatively, the patient experienced full resolution of her neurological deficits and retained full bowel and bladder function as well as lower extremity strength and sensation. MRI was performed to monitor for indications of adjuvant RT. A residual, stable 2-mm nodular focus of intradural/extramedullary enhancement at the level of T4–5 was identified on MRI (performed with metal artifact reduction protocol).

She received subsequent adjuvant radiation several weeks postoperatively. For radiation planning, the postoperative MRIs were fused to the planning software, and the clinical treatment volume was defined as the entire spinal canal and nerve roots extending from C2 to the terminus of the thecal sac. She was treated on TomoTherapy with 6 MV photons using an intensity modulated radiotherapy technique using daily image guidance. The field extended from C2 to the bottom of the thecal sac and was treated at 150 cGy/day to a total cumulative dose of 4,200 cGy. Given the MRI evidence of a 2-mm



FIG. 3. Two postoperative radiographs showing spinal fusion with instrumentation from T7 to the sacrum and improvement of thoracolumbar kyphosis compared to preoperative imaging. Imaging captures the thoracic levels (left) and the lumbar to sacral levels (right).

nodular focus of intradural/extramedullary enhancement at the level of T4–5, using a concomitant boost technique, the area at T4–5 and below L2 received a higher dose of 180 cGy/day for a total cumulative dose of 5,040 cGy.

The patient is currently 59 months out from her staged tumor resection and fusion and remains neurologically intact. On the most recent follow-up contrasted MRI, at 52 months after the recurrent tumor resection, there is no evidence of tumor progression. Her thoracolumbar rods have broken bilaterally, but because she is not overtly symptomatic from this, we are managing that finding nonoperatively.

Discussion

Observations

Our case demonstrates prolonged asymptomatic, disease-free survival after GTR and RT of symptomatic, disseminated, recurrent MPE. We reviewed the literature for cases of disseminated MPE with a PubMed database search of “disseminated myxopapillary ependymoma,” which yielded 191 results. After applying inclusion criteria of confirmed MPE; dissemination, which was defined as multiple neuraxis lesions, drop metastases, or extraneural dissemination at initial presentation and/or at time of recurrence; and treatment data, we identified 24 reports of 75 distinct patients (Table 1).^{4–45} In the setting of disseminated recurrence, there were five reported cases of GTR in four different patients. RT was also used in one of these cases. Abdallah et al.²¹ reported GTR of one drop metastasis, whereas Lee et al.²² reported GTR of a maximum of two masses at the time of surgery. Lastly, Abdu et al.²³ described a case in which one mass was removed by GTR. Thus, to the best of our knowledge, our illustrative case with GTR of five lesions is the greatest number of lesions removed by GTR in the setting of disseminated MPE at the time of recurrence. Our case also demonstrates the longest period of asymptomatic, disease-free survival after reoperation for symptomatic, recurrent disseminated MPE among reported cases of GTR of recurrent disseminated MPE.

Lessons

Surgical Decision-Making

The reported overall survival for MPE is high, with rates of 90% to 100% at 5 years.^{10,14,16,24} However, the concern with this condition is a loss of spinal cord function at or below the spinal level with the greatest tumor burden. Surgical decision-making should be carefully considered for treatment of MPE, and patients ought to be counseled that surgical and/or adjuvant RT has not demonstrated improvement in overall survival.^{14,16,24}

Consideration must be given to the constellation of symptoms a patient is experiencing. Ultimately, the risks of surgery must be weighed against the likelihood of tumor progression. As in our case, metastatic seeding of the thecal sac can occur after resection of WHO grade II MPE.^{8,10} Treatment failure rates have been reported in approximately 30% of patients.^{14,16} Kraetzig et al.¹⁰ reported distant metastases in 57.9% (11/19) of patients with MPE; 36% (4/11) of cases of disseminated MPE were identified at the time of initial diagnosis, whereas 64% (7/11) were identified during the follow-up period with a median time to recurrence of 20 months. Interestingly, 72.7% (8/11) of these patients remained asymptomatic with no evidence of progression over a median follow-up period of 32 months. The authors concluded that close follow-up is an appropriate option for asymptomatic patients with metastatic disease.¹⁰

TABLE 1. Patient and treatment information for cases of disseminated MPE at initial presentation or as recurrent disease

Authors & Year	Age, Sex	Initial Presentation				Time to Recurrence	Recurrence		
		Local/Disseminated	1° Treatment	2° Treatment	Local/Disseminated		1° Treatment	2° Treatment	
Vongsfak et al., 2020 ²⁹	13, M	Disseminated	STR	RT	No recurrence at 2-mo follow-up				
Straus et al., 2014 ⁴	63, M	Disseminated	NTR	RT	No recurrence at 6-mo follow-up				
Mishra et al., 2021 ³⁰	18, M	Disseminated	Biopsy	RT	No recurrence at 18-mo follow-up				
Toktaş et al., 2015 ³¹	26, M	Disseminated	STR		None, no follow-up data provided				
Abdallah et al., 2020 ²¹	11, M	Local	GTR (2 segments)		75 mos later	Disseminated	GTR		
	13, F	Local	GTR (2 segments)		45 mos later	Disseminated	RT		
	12, F	Local	STR	RT	12 mos later	Disseminated	Biopsy	RT	
Looi et al., 2021 ³²	15, M	Local	GTR		16.3 mos median time to recurrence	Disseminated	STR		
	8, M	Local	STR			Disseminated	STR		
	12, M	Disseminated	GTR			Disseminated	STR		
	8, F	Local	GTR			Disseminated	RT		
	8, F	Disseminated	STR	RT		Residual LM disease	None		
	10, F	Local	GTR			Disseminated	RT		
	14, F	Local	STR			Disseminated	STR		
	13, M	Disseminated	NTR	RT		Small residual	None		
	7, F	Disseminated	NTR	RT		Small residual	None		
	16, F	Disseminated	GTR			Disseminated	STR		
	21, M	Disseminated	NTR	RT		Small residual	None		
	18, M	Disseminated	GTR	RT	No recurrence	None			
Abdulaziz et al., 2015 ¹¹	59, M	Local	STR		12 mos	Disseminated	Chemo	RT	
Khalatbari et al., 2013 ³³	16, M	Disseminated	STR	RT	No recurrence at 5-yr follow-up				
Fujiwara et al., 2018 ³⁴	26, F	Local	GTR		2 yrs	Local	GTR	RT	
					4 yrs	Disseminated	RT		
					6 yrs	Local	GTR	TMZ	
Fonseca et al., 2019 ³⁵	21, M	Local	STR	RT	21 yrs	Disseminated	STR	RT	

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TABLE 1. Patient and treatment information for cases of disseminated MPE at initial presentation or as recurrent disease

Authors & Year	Age, Sex	Initial Presentation				Time to Recurrence	Local/Disseminated	Recurrence	
		Local/Disseminated	1° Treatment	2° Treatment	Local/Disseminated			1° Treatment	2° Treatment
Awaya et al., 2021 ⁵	22, M	Disseminated	GTR of dominant lesion		No recurrence at 12 mos				
Zhu et al., 2019 ³⁶	23, M	Disseminated	STR	RT	No recurrence at 18 mos				
Lee et al., 2019 ²²	13, M	Disseminated	STR	RT & Avastin	Stable residual disease at 4 yrs	None			
	16, F	Disseminated	GTR		4 yrs	Disseminated	Resection NFS		RT
	31, M	Disseminated	STR	Chemo RT	9 mos	Local	STR		
	31, F	Disseminated	Resection NFS	RT	14 yrs	Disseminated	GTR		Chemo
	40, F	Disseminated	STR	Proton therapy	None at 2.5 yrs				
	45, M	Disseminated	STR	RT	12 yrs	Disseminated	GTR		
Fassett et al., 2005 ³⁷	13, F	Disseminated	GTR	RT	15 yrs	Disseminated	GTR		RT
	14, M	Disseminated	GTR	RT	No recurrence at 5 yrs				
	14, M	Disseminated	STR	RT	No recurrence at 3.5 yrs				
	8, M	Disseminated	STR	RT	Stable residual, local disease at 4.5 yrs	None			
Macedo et al., 2011 ³⁸	33, M	Disseminated	GTR of cerebral lesion	Biopsy of lumbar lesion & RT	Stable disease at 2 mos from completion of RT	None			
Deniel et al., 2019 ³⁹	30, F	Local	GTR		3 recurrences over 10 yrs	Local	GTR each time		
					11 yrs	Local	RT		
					13 yrs	Disseminated	TMZ		Patient died within a few weeks of initiating TMZ
Rege et al., 2016 ⁴⁰	23, M	Local	STR	RT	8 mos after RT	Disseminated	STR		RT
Abdu et al., 2021 ²³	47, F	Local	GTR	RT	28 mos	Disseminated	GTR 44 mos after initial GTR		
Plans et al., 2006 ⁴¹	23, M	Local	STR	RT	48 mos	Disseminated	Biopsy		
					3 yrs	Disseminated	Watchful waiting for 4 yrs		Biopsy & RT 7 yrs after

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TABLE 1. Patient and treatment information for cases of disseminated MPE at initial presentation or as recurrent disease

Authors & Year	Age, Sex	Initial Presentation				Time to Recurrence	Recurrence		
		Local/Disseminated	1° Treatment	2° Treatment	Local/Disseminated		1° Treatment	2° Treatment	Local/Disseminated
Al-Halabi et al., 2010 ⁴²	14, M	Disseminated	STR	RT	4 mos	Disseminated	STR	RT	
	14, M	Local	GTR		2 mos	Disseminated	RT		
	18, M	Local	GTR		36 mos	Disseminated	RT		
	18, M	Disseminated	GTR	RT	No recurrence at 78 mos				
Bandopadhyay et al., 2016 ⁴³	17, F	Disseminated	GTR		NA	Disseminated	None		
	8, M	Local	STR	RT		Disseminated	None		
	21, F	Disseminated	STR			Disseminated	STR	RT	
	8, M	Disseminated	GTR			Disseminated	STR	Chemo, RT	
	15, F	Disseminated	STR			Disseminated	STR	Chemo, RT	
	19, M	Disseminated	STR	RT		Disseminated (asymptomatic)	None		
12, M	Local	GTR			Disseminated (asymptomatic)	None			
10, F	Disseminated	GTR			Local	Resection	NFS		
14, F	Disseminated	STR			None				
17, F	Disseminated	STR			None				
11, M	Disseminated	GTR			Local (asymptomatic)				
Pencovich et al., 2014 ⁴⁴	36, M	Disseminated	STR		None at 9 mos				
35, M	Disseminated	STR			19 mos	Local			
28, F	Disseminated	STR			None at 1.5 yrs				
37, F	Disseminated	GTR	RT		None at 6 yrs				
18, M	Disseminated	STR			None at 4.5 yrs				
14, M	Disseminated	STR	RT		None 2 yrs				
12, M	Disseminated	GTR	RT		None at 3 yrs				
15, M	Disseminated	STR			None at 7 yrs				
17, M	Disseminated	STR			None at 7.5 yrs				
Higgins et al., 2005 ⁴⁵	36, F	Disseminated	Biopsy	RT	6 mos after RT	Disseminated	Dexamethasone		

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TABLE 1. Patient and treatment information for cases of disseminated MPE at initial presentation or as recurrent disease

Authors & Year	Age, Sex	Initial Presentation			Time to Recurrence	Recurrence		
		Local/Disseminated	1° Treatment	2° Treatment		Local/Disseminated	1° Treatment	2° Treatment
Kraetzig et al., 2018 ¹⁰	21, M	Local	STR		4 mos	Disseminated	Resection NFS	Chemo, RT
					12 mos	Local	Resection NFS	
	27, M	Local	GTR		2 mos	Disseminated (asymptomatic)	None	
	9, M	Local	GTR	RT	20 yrs	Disseminated	Resection NFS	Chemo, RT
	30, M	Local	STR	RT	20 mos	Disseminated (asymptomatic)	None	
	43, M	Disseminated	GTR		None at 36 mos			
	20, F	Disseminated	GTR		None at 24 mos			
	18, F	Disseminated	GTR		None at 6 mos			
	32, M	Local	GTR		3 mos	Disseminated (asymptomatic)	None	
	15, M	Local	GTR	RT	30 yrs	Disseminated	Resection NFS	
48, M	Disseminated	STR	RT	42 mos	Disseminated (asymptomatic)	None		

Chemo = chemotherapy; LM = leptomeningeal; NA = not applicable; NFS = not further specified; NTR = near total resection; TMZ = temozolomide.

TABLE 2. Recommendations for POVL prevention for spine surgery

Preoperative
During the informed consent, inform the patient of risks of prolonged* prone positioning, including POVL, and substantial blood loss† Debrief members of the surgical team regarding the importance of monitoring labs and pressure points and adequately compensating for any fluid/blood losses
Intraoperative Monitoring and Management
Blood pressure MAP maintained between 75 and 90 mm Hg
Blood/fluid compensation Hemoglobin, hematocrit, and sodium levels consistently monitored throughout the duration of the procedures Combination of blood products, crystalloids, and colloids administered to offset losses
Vasopressors Phenylephrine administered to maintain MAP in desired range
Positioning Jackson table and Wilson frame utilized to avoid direct pressure over the eyes and face with continued monitoring for pressure points during the case Maintained head above the level of the heart to reduce risk of increased intracranial and intraocular pressure
Staging of procedures Tumor resections performed on operative day 1 and fusion with instrumentation is carried out on the subsequent day
Postoperative Evaluation and Management
Visual acuity examined during the postoperative evaluation

MAP = mean arterial pressure.

* Prolonged procedures are defined as spine procedures >4 hours.²⁸

† Substantial blood loss is defined as blood loss >800 mL.²⁸

The ultimate concern for these patients is that at some point they may become symptomatic and surgery will not be an option if the tumor spread is too diffuse. Symptomatic recurrence can impact quality of life, and for patients who are symptomatic, surgery appears to offer the greatest benefit.^{10,12,24}

Technical Considerations for Achieving GTR with Disseminated MPE at Recurrence

GTR appears to offer the highest likelihood of progression-free survival.^{14,24,25} The surgical goal is to resect each MPE lesion en bloc without disruption of the capsule. However, the natural tendency for these lesions to be adherent to neurological structures makes en bloc or even piecemeal GTR challenging; therefore, it is often not achieved. In our review of the literature, we identified 50 patients with disseminated disease at initial diagnosis, with 18 (36%) of them receiving GTR of any of their lesions whereas 30 (60%) had subtotal tumor resection. Among the 40 patients with disseminated lesions at the time of recurrence, 5 (12.5%) received GTR of at least one tumor, and STR was achieved in 14 (35%) of the patients. Moreover, among patients with disseminated disease, microscopic spread is assumed to have occurred and GTR is not achieved beyond the primary tumor targeted for resection.

In our case, preoperative contrasted MRI highlighted five discrete intradural tumor foci at T7, T10, L2, L3, and S1–3. The initial surgical exposure extended from T10 to the sacrum. We performed wide laminectomies at each affected level to maximize the likelihood of safe GTR of lesions adherent to the spinal cord and/or nerve roots.²⁶ We began with the S1–3 lesion, which was resected along with its encapsulating dura while preserving the exiting nerve roots and temporarily occluding the caudal thecal sac to avoid

cerebrospinal fluid leakage, although no fluid was noted until the L2 and L3 lesions were resected. We then moved rostrally in a step-wise manner, performing additional wide laminectomies and opening the dura along the midline and resecting the lesions at the corresponding levels individually. We then extended our initial exposure to T7, allowing resection of the lesion at this level.

We used intraoperative neurophysiological monitoring because it has proven to be an effective tool to guide intraoperative surgical and medical adjustments to reduce the likelihood of neurological injury. Identification of the tumor-nerve/spinal cord interface is paramount, particularly when involving encapsulated lesions such as MPE.²⁷ Neuromonitoring capabilities were essential for dissection of the lesions at the S1–2 level, L2, and L3 because these lesions were adherent to nerve roots. We were able to identify the tumor-nerve interface surrounding the S1–2 lesion. We were unable to identify a plane circumferentially around the lesions at L2 and L3, and they were removed in a piecemeal fashion with periodic nerve stimulation for tumor-nerve plane confirmation. Furthermore, neuro-monitoring enabled us to identify a lesion within the nerve sheath of the S1 nerve root after this root failed to respond to stimulation.

Considerations for Prolonged Prone Positioning

Based on our literature review, this may be the longest reported prone position case completed without complication. Our patient was in the prone position for 14.5 hours on operative day 1. On the subsequent day, she was placed prone for an additional 10.5 hours, for a total of 25 hours prone in less than a 34-hour period. Various complications are associated with prolonged prone positioning with or without intraoperative hypotension, including perioperative vision loss (POVL).^{19,20} Specific thresholds for vital signs, lab values, and

positioning are difficult to ascertain. However, there are practice advisories with recommendations for reducing the risks of POVL during spine surgery (see Table 2 for a summary of considerations to minimize POVL).²⁸

We used a Jackson table and Mayfield head-holder for our procedure to avoid any direct contact with the face or eyes, and neuro-monitoring was used for identifying neurological injury secondary to the procedure itself or prolonged positioning. All potential pressure points were padded appropriately, radial arterial pressure was monitored via catheter, and a left subclavian triple lumen vascular access was placed.

During the case, avoidance of hypotension, fluid maintenance, and offset of blood loss were strictly observed during rotation of two anesthesiologists and seven certified registered nurse anesthetists. Despite the potential for exacerbation of surgical blood loss, the patient's head was maintained higher than the level of the heart throughout the procedure to avoid elevation of intracranial and intraocular pressures. The patient's mean arterial pressure was maintained between 75 and 90 mm Hg with phenylephrine throughout the procedure on operative days 1 and 2. The following data were recorded for operative day 1: 1.85 L of estimated blood loss (EBL), urine output of 11.325 L, hemoglobin (N = 10) of 8.3 to 11.6, hematocrit (N = 16) of 21 to 33.2, and sodium (N = 11) between 143 and 150. Blood and fluid losses were corrected with 1.5 L of packed red blood cells (RBCs), 120 mL of autologous RBCs, 500 mL of fresh frozen plasma (FFP), 6 L of lactated Ringer's (LR) solution, 5 L of 0.45% NaCl, 6.6 L of Plasmalyte, and 250 mL of albumin.

On operative day 2 there was an EBL of 1.5 L and a urine output of 8.9 L. Monitoring values included hemoglobin (N = 6) of 9.4 to 12, hematocrit (N = 13) of 22 to 33.7, and sodium (N = 9) of 140 to 149. Blood and fluid losses from operative day 2 were compensated with 1.05 L of packed RBCs, 450 mL of autologous RBCs, 784 mL of FFP, 243 mL of platelets, 6.9 L of LR solution, 1.55 L of 0.45% NaCl, 6.9 L of Plasmalyte, and 250 mL of albumin.

Limitations

Limitations to the generalizability of lessons of this case may preclude extrapolation to other cases of disseminated MPE. In the absence of randomized, controlled trials and evidence-based guidelines, clinical decision-making in the setting of disseminated MPE remains a matter for discussion between patients and physicians. Because each patient brings their own risks into surgery and because of variability in surgical skills, one should not generalize that resection of disseminated spinal cord tumors followed by radiation outweighs the risks of other management strategies.

Furthermore, our experience of uncomplicated prolonged prone positioning may not be generalizable because our patient may not be representative of the larger population of patients with disseminated MPE or those being considered for procedures necessitating prolonged prone positioning. It is important to acknowledge that our patient was in her teens and had a BMI of 22.1. Both factors likely played a protective role against complications with prolonged prone positioning and may not be present in other patients presenting with disseminated MPE. Thus, our experience should be considered cautiously when planning the positioning of other patients with different risk factor profiles.

Conclusions

MPE is a rare, WHO grade II neoplasm with the potential to recur as disseminated disease after primary surgical intervention.

Here we demonstrate that in an appropriately selected patient with disseminated lesions, resection can be a safe and effective treatment modality to achieve prolonged asymptomatic progression-free survival. Additionally, we report on potentially the longest duration of prone positioning for spine surgery without complication. Further studies are needed to establish evidenced-based treatment guidelines for focal and disseminated MPE.

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Author Contributions

Conception and design: Samadani, Johnson, Cramer. Acquisition of data: all authors. Analysis and interpretation of data: Samadani, Johnson. Drafting the article: Samadani, Johnson, Cramer. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Samadani. Statistical analysis: Johnson. Administrative/technical/material support: Johnson, Cramer. Study supervision: Cramer.

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