

MYXOPAPILLARY EPENDYMOMAS: Review of Literature

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INTRODUCTION:

Myxopapillary ependymoma (MPE) is a distinct type of spinal cord ependymoma that has a predilection for the lumbosacral region.¹ 13% of all spinal ependymomas are comprised of these tumours, and these tumours account for 90% of all tumours in the conus medullaris.^{2,3} Myxopapillary ependymoma is a benign slow-growing neoplasm according to the world health organization (WHO) classification. It has been classified as Grade². Myxopapillary ependymomas comprise 1% to 5% of all spinal neoplasms. The age of onset varies between 30 and 50 years, and the mean age of presentation is 35 years. Myxopapillary ependymoma is commonly located in the lumbosacral spine segment, commonly in the conus medullaris and cauda equine regions. Other locations include the cerebral ventricles and the brain⁴; however, they are also present rarely in uncommon locations without any primary lesion in the spinal cord, such as subcutaneous or deep soft tissue.⁵ As it is encapsulated and a slow-growing tumour, surgical excision without capsule rupture is generally curative.⁶ Surgical resection is the primary mode of treatment, and the goal is to achieve complete resection while minimizing postoperative neurological deficits. The extent of the surgical resection is variable and complex because of the anatomical relationship of these tumours to the surrounding nerve roots and conus medullaris. The strategic location of MPE in the conus-cauda area and the fact that these lesions present late with attachment to eloquent neural structures and extensive local disease are

limiting factors in the completeness of surgical resection.^{7,8}

Though Myxopapillary ependymomas are an uncommon primary neoplasm, it has been seen to have higher recurrence rates.⁹ Tumours with anaplastic features are sporadic, and they usually have a high recurrence rate with poor clinical outcomes and tend to metastasize.¹⁰

Myxopapillary ependymomas can occur at ectopic sites, such as the sacrum and in the presacral tissue where ependymal cells may be found. These ectopically located tumours have a worse prognosis and a higher rate of extraneural metastasis. Sacrococcygeal,^{11,12,13} intracranial,¹⁴ and soft tissue myxopapillary ependymomas have been described, as well as extra neural metastases in the lungs, pleura, liver, and thoracic and abdominal lymph nodes.¹⁵ Case series on extradural ependymomas showed a higher local recurrence rate (60% for presacral and 25% for tumours on the dorsal aspect of sacrum) compared with classical MPEs, with a mortality rate of up to 50% in one case of local recurrence¹⁶ and a 100% 5-year mortality if metastases occur. Giant sacral MPEs may require resection, lumbopelvic reconstruction, and fusion, followed by radiotherapy.¹² Intradural lumbosacral ependymomas can spread throughout the central nervous system but rarely metastasize beyond it. In contrast, extradural ependymomas

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seldom disseminate within the CNS but pose a significant risk for systemic metastases.¹⁷

CLINICAL PRESENTATION:

Myxopapillary ependymomas do not show specific clinical presentation and offer a similar presentation as that of other intradural tumours located in the region of the filum terminale.¹ The presenting symptoms are related to the size of the tumour, the site of presentation, and the local extent of the tumour. A sluggish and slow clinical course is a characteristic of myxopapillary ependymomas. The majority of the patients have a nonspecific presentation. Thus, patients often experience symptoms for months to years before diagnosis.¹⁸ Patients usually complain of radicular and back pain worsening during the night and in the lounging position.¹ Myxopapillary ependymomas may patients show motor, sensory, urinary, and gait abnormalities.¹ They may also present with enlargement of neural foramina, scoliosis, and scalloping of the vertebral body. Sudden worsening of the symptoms may occur secondary to haemorrhage with leg weakness and sphincter disturbances. However,¹ Saadah M et al. conducted a study in which some patients presented atypical presentations like haemorrhage, intracranial hypertension, low CSF sugar, hydrocephalus, acute autonomic crisis, and SIADH.¹⁹

Diagnosis of Myxopapillary Ependymomas:

Radiology:

Magnetic resonance imaging (MRI) is the best imaging modality for diagnosing myxopapillary ependymoma because of its superior soft-tissue contrast. Other imaging modalities include plain radiographs and computed tomography (CT) scans. Plain radiographs and CT scans reveal scalloping of the vertebral bodies secondary to the expansion of the spinal canal caused due to sizeable myxopapillary ependymoma.

Magnetic resonance imaging identifies the extent of the tumour and its relationship to intraspinal structures.^{20,21} On magnetic resonance imaging. The lesion is isointense to hypointense on T1 and hyperintense on T2 (figure 1a). Sometimes the tumour may show T1 hyperintensity owing to its mucin content. Superficial siderosis is nonspecific but reflects its vascular nature. On postcontrast imaging, robust enhancement is present.²² Hemorrhage and cyst formation are common features that contribute to signal inhomogeneity.²¹ This may vary from hypointense on T2

with heterogeneous diffusion restriction and lack of post-contrast enhancement, as was seen in the study by Albadr F et al.²³

On imaging, tumours do contain certain features that help suggest the diagnosis.

These features include;

- a. An intradural extramedullary thoracolumbar mass extending several vertebral levels in the lumbar and sacral canal.
- b. Mass hypointense to isointense on T1-weighted images.
- c. It is hyperintense on T2-weighted images.
- d. Cystic rostral or caudal degeneration in 50% of cases.

Differential Diagnosis of MPEs on Radiology:¹⁴

The differential diagnosis for small myxopapillary ependymomas on radiology include; Schwannoma, Sub ependymoma, Astrocytoma, Hemangioblastoma and Ganglioblastoma.

The differential diagnosis for large myxopapillary ependymomas on radiology include; Aneurysmal bone cyst, Chordoma, Plasmacytoma, Metastasis and Giant cell tumour.

On MRI, Schwannomas are isointense to hypointense on T1-weighted images and are heterogeneous with the hyperintense signal on T2-weighted images.²⁵ Sub ependymomas are hypointense on T1 weighted images, whereas they are hyperintense on T2 weighted images, but they lack distinctive imaging features from other neoplasms of glial origin. Astrocytomas usually are eccentrically located with infiltrative edges and show hypo to iso intensity on T1 images and hyperintensity on T2 images on MRI. Hemangioblastomas on MRI are isointense as other tumours; however, with gadolinium contrast, they show an enhancing tumour nodule which is homogenous, whereas, on T2, they show peritumoral oedema along with syrinx mostly. Gangliogliomas on MRI are often heterogeneously hyper to hypointense on T1 images, which is distinct from the other gliomas, whereas, on T2 images, they are hyperintense with absent peritumoral oedema. Metastatic lesions on MRI show two types of intensity patterns on T1. Either they are homogeneously enhancing, or they show a ring-like pattern owing to central necrosis in some cases where, as they are almost always hyperintense on T2. Chordomas on MRI show hyperintensity due to

haemorrhage, myxoid and mucinous areas. At the same time, they are hypo (due to fibrous tissue and hemosiderin) to hyperintense (due to high water content) on T2. Plasmacytomas are hypointense on T1 and hyperintense on T2, with the bony trabeculae showing hypointensity on T2. Giant cell tumour On MRI imaging shows hyperintensity, whereas they show low to intermediate intensity on T2.

HISTOPATHOLOGY OF MPEs:

"Squash Cytology Features For Quick Diagnosis In MPEs:"

On crush cytology, myxopapillary ependymoma displays high cellularity and a papillary pattern with fibrovascular cores. Some clusters of cells are arranged in pseudorosettes with a central myxoid to a hyaline area surrounded by a radial layer of cuboidal to columnar cells. Hyaline globules, fibrillary cytoplasmic structures, and myxoid material may also be seen. The nuclei of these cells are round to oval with speckled chromatin. Intranuclear inclusions, nuclear grooves, cytologic atypia, or mitotic activity are usually absent (figure 1b). According to a study by Bradly DP et al., the cytological features included round to spindle cells arranged in clusters, dispersed singly along with focal papillary and adenoid cystic-like areas. The cells had indistinct cytoplasmic borders with delicate cytoplasm, oval to plump nuclei with finely dispersed chromatin. In addition, the stroma is comprised of thick metachromatic material.²⁶

Fine Needle Aspiration Cytology of MPEs:

On guided FNAC of Sacro lumbar lesion, Kulesza P et al. found that cytological features are similar to those seen in squash smear with a biphasic tumour with both epithelial and stromal elements. The epithelial element comprises loosely cohesive clusters and papillae with fine chromatin and cytoplasm with indistinct cytoplasmic borders; the stroma consists of thick fibrillary hyaline globules.^{27,28}

Macroscopic Features:

Myxopapillary ependymomas are sausage-shaped and often encapsulated. They are soft in consistency and reddish to purplish.²⁹ Sometimes, mucinous degeneration and haemorrhage may be seen in these tumours.³⁰

Microscopic Features:

A papillary architecture histologically characterizes myxopapillary ependymomas blood vessels surrounded by

cuboidal to spindled glial cells in the papillae's centre. The stroma is myxoid and often Alcian blue-positive.³¹ Vascular hyalinization and other degenerative changes are often prominent (figure 1c & 1d). Some tumours may show a more compact fascicular pattern with occasional perivascular pseudorosettes with mucin-rich microcysts.

In some cases, the tumour may exhibit rounded to spiculated collagen balloons, and mitotic figures are usually scarce. Endothelial proliferation and necrosis are absent. In isolated case reports, some authors have described anaplastic and giant cell variants of myxopapillary ependymomas. As per the WHO, the tumour is Grade 2. Sometimes in children, it may behave aggressively and shows poor clinical outcome in partially resected tumours.

Differential Diagnosis of MPEs on Histopathology:

The differential diagnosis of filum terminale and small conus myxopapillary ependymomas include Meningiomas, Schwannoma, Metastatic carcinoma and Paraganglioma.

Fibrous Meningioma and Schwannoma: The fibrillary variant of myxopapillary ependymomas may be confused with fibrous meningioma; however, stainable mucin in myxopapillary ependymoma helps it to differentiate from meningiomas and schwannomas which rarely produce mucin. The epithelial and papillary variants of myxopapillary meningioma may mimic carcinoma or meningioma. Still, the myxopapillary ependymomas lack malignant cytology and are focally fibrillar, and on IHC, GFAP stain helps to differentiate it from above.

Paragangliomas can mimic myxopapillary ependymomas. However, the presence of mucin and absence of secretory granules, along with chromogranin-A negativity and GFAP positivity, helps to confirm the diagnosis.

The differential diagnosis of a sizeable myxopapillary ependymoma responsible for sacral destruction includes Aneurysmal bone cyst, Chondrosarcoma, Chordoma, Giant cell tumour and Metastatic carcinoma.

However, from the above lesions, imaging and lack of cytological atypia, focally fibrillar background, on IHC lower Ki67 and GFAP positivity help establish the diagnosis.

Immunohistochemistry:

An immunohistochemical study is frequently required to distinguish myxopapillary ependymoma from other

neoplasms. The tumour cells are often positive for S-100, GFAP, CD99, and COX-2. However, they are harmful to epithelial membrane antigen (EMA), but some may demonstrate punctate cytoplasmic staining. They may show focal positivity for CK^{5,7,8,18,20} and expression of CK AE1/AE3, which may create difficulty in distinguishing it from metastatic carcinoma; however, these should not be interpreted as markers for metastasis without GFAP negativity in the tumour, they can also show positivity for NSE in cases of neuroglial differentiation.³²

Molecular profile:

Myxopapillary ependymomas arise from the ependymal glial cells of the filum terminale. No risk factors for the development of this tumour have been described. Coccyeal medullary vestige or extradural remnants of filum terminale are thought to give rise to extradural myxopapillary ependymomas²⁰. These tumours usually harbour loss-of-function/NF² mutations or deletions.³³ A study conducted by Zeng H et al. found that these tumours had higher expression of HOX genes and lncRNA HOTAIR³⁴ In another study by Omerhodzic¹ et al., the HOXB¹³ gene, a molecular signature of these tumours, was more specific and equally expressed in pediatric and adult tumours.²⁴ Expression of the HOXB13 gene was found to be limited to the nucleus by Stephen JH et al.³⁵ In Omerhodzic I et al. study; it was also found that these tumours had low MIB-1, index, and expression of Epidermal growth factor receptor (EGFR) protein was associated with poor clinical outcomes in intracranial MPes.²⁴

TREATMENT AND MANAGEMENT:

There are no definitive treatment guidelines owing to the rarity of myxopapillary ependymomas. The consensus is that surgical excision is the preferred initial treatment modality.³⁶ Early detection and treatment of patients affected by myxopapillary ependymomas is crucial to achieving optimal patient outcomes. Complete surgical resection is the best treatment modality for myxopapillary ependymoma, which is associated with a full resolution of the tumour for most patients.^{37,38} Early surgery should be performed to guarantee better postoperative outcomes.³⁹ Complete resection of the tumour may be hazardous if the tumour involves the conus medullaris or is interlaced with the surrounding nerve roots of the cauda equina. Surgeons

should be cautious since nerve roots may penetrate the tumour directly.⁴⁰

Adjuvant radiotherapy is recommended in the case of subtotal resection; however, chemotherapy is used in young children, who are more prone to side effects from radiation therapy.³⁹ Although chemotherapy has been suggested as a potential treatment to prevent a recurrence of these tumours. Its efficacy has not been established.⁴¹ Radical surgery for myxopapillary ependymoma has been associated with a favourable outcome in children and adults, as with other spinal cord ependymomas.⁴² Long-term survival is better in the case of gross total resection of myxopapillary ependymoma rather than subtotal resection.^{1,40}

ADJUVANT RADIOTHERAPY:

Owing to the benign and slow-growing nature of the tumour, the therapeutic benefits of radiotherapy for myxopapillary ependymomas are still debated. In one of the studies, after total or subtotal resection, there was no difference in irradiated versus nonirradiated patients.⁴⁰ Some of the studies have reported the benefit of postoperative radiotherapy for patients who had subtotal resection of myxopapillary ependymoma due to high-risk of long-term recurrence in these patients^{43,44,45} Other studies demonstrated that radiotherapy is significantly more beneficial in the case of sub totally resected myxopapillary ependymomas smaller than 6cm in diameter compared to larger tumours (58% versus 92% 5- and 10- year progression-free survival). It has been found that high-risk cases (STR or questionable GTR) should be considered for postoperative RT because of the chances of recurrence considering the proposed benefits of adjuvant treatment.⁴⁶ Some studies have also reported metastasis of these tumours to the spinal cord (9%) and brain (6%) in children, after partial resection and absence neoadjuvant radiation therapy.⁴⁹

PROGNOSIS:

To predict the natural history, the likelihood of recurrence, or metastases, neither the histopathology nor the MIB-1 index appears helpful. Patients should be followed for monitoring tumour recurrence; even if a gross total resection was accomplished, postoperative serial magnetic resonance imaging is advised. Recurrence rates of 10% to 19% have been reported in patients who underwent gross

total resection.^{1,40}

Tumour recurrence and dissemination may occur despite the good results of surgery. Clinical history of less than one year, the extent of resection, atypical histology, and lesions extending into the substance of the spinal cord is associated with Local recurrence and metastases.^{45,48,49} Myxopapillary ependymomas are central nervous system tumours with long-term survival rates and a tendency for local recurrence. However, cerebrospinal fluid dissemination and even systemic metastases may be seen.⁴⁹ Myxopapillary ependymomas may occasionally show aggressive behaviour. Local progression is common and systemic metastases more commonly involving lungs have also been described. For tumours with primary extraspinal location, systemic dissemination is more common.¹ Patient with myxopapillary ependymoma has excellent 5-year overall survival rates varying from 90% to 100%. Patients of myxopapillary ependymoma who underwent gross total resection have a good prognosis, with a mean survival time of 19 years.¹ Although rarely encountered in children, myxopapillary ependymomas have an aggressive clinical course with an increased risk of recurrence, and dissemination is seen in children although it is rare in children. The reason for this more aggressive course in children is unclear.

COMPLICATIONS:

En-bloc removal of encapsulated myxopapillary ependymomas of the filum terminale has lower dissemination rates (10%) than myxopapillary ependymomas removed piecemeal (19%).¹ Rupture of the capsule of myxopapillary ependymoma during surgery may result in cerebrospinal seeding and dissemination. However, other complications like neurological deficits, pulmonary embolism, wound infections, cerebrospinal fluid leakage, cysts, paraplegia, kyphosis and scoliosis may also occur.^{49,50} Dissemination and treatment failure may occur in 30% of the cases.^{49,51} Extension of myxopapillary ependymoma into the substance of the spinal cord, a clinical history of less than one year, the extent of resection, and atypical histology of the tumour are associated with increased risk of recurrence and metastases.⁴⁹

CONCLUSION:

Although myxopapillary ependymomas are classified as

benign tumours, dissemination and local recurrence pose a significant challenge in their clinical treatment. Therefore, a close follow-up of the patients with myxopapillary ependymomas is mandatory to detect local recurrence or spread of the tumour.

CONFLICT OF INTERESTS:

The authors declare that they have no conflict of interest regarding the publication of the paper.

REFERENCES:

1. Sonneland PR, Scheithauer BW, Onofrio BM. Myxopapillary ependymoma. A clinicopathologic and immunocytochemical study of 77 cases. *Cancer*. 1985; 56(4):883-93.
2. Allen JC, Siffert J, Hukin J. Clinical manifestations of childhood ependymoma: a multitude of syndromes. *Pediatr Neurosurg*. 1998; 28(1):49-55.
3. Celli P, Cervoni L, Cantore G. Ependymoma of the filum terminale: treatment and prognostic factors in a series of 28 cases. *Acta Neurochir(Wien)*. 1993;124(2-4):99-103.
4. Feldman WB, Clark AJ, Safaee M, Ames CP, Parsa AT. Tumour control after surgery for spinal myxopapillary ependymomas: distinct outcomes in adults versus children: a systematic review. *J Neurosurg spine*. 2013; 19(4):471-6.
5. Haiyin Zheng, Katherina Baranova, Jun Song, Lei Yan, Saumik Biswas, Subrata Chakrabarti, Qi Zhang. Overexpression of long Noncoding RNA HOTAIR Is a unique epigenetic characteristic of myxopapillary ependymoma. *Journal of Neuropathology & Experimental Neurology*, Vol 79, Issue 11, 2020, pp1193-1202.
6. Kanno H, Kanetsuna Y, Shinonaga M. Anaplastic myxopapillary ependymoma: A case report and review of literature. *World Journal of Clinical Oncology*. 2021;12(11):1072.
7. Anderson MS. Myxopapillary ependymomas presenting in the soft tissue over the sacrococcygeal region. *Cancer*. 1996; 19(4): 585-90.
8. Quraishi NA, Wolinsky JP, Bydon A, Witham T, Gokaslan ZL. Giant destructive myxopapillary ependymomas of the sacrum. *J Neurosurg Spine*.

- 2010;12(2):154-9.
9. Alessandro Pesce, Mauro Palmieri, Daniele Armocida, Alessandro Frati, Massimo Miscusi, Antonino Raco. Spinal Myxopapillary Ependymoma: Experience and Comprehensive Literature Review Concerning the Clinical Course of 1602 patients. *World Neurosurg*. 2019;129:245-253
10. Lorenzo Gitto, Serenella Serinelli, Kristyn Galbraith, Michael Williams, Kanish Mirchia, Michael A Galgano, Satish Krishnamurthy, Gustavo de la Roza, Mariano S Viapiano, Jamie M Walker, George Jour, Jonathan Serrano, BS, Michael DeLorenzo, BS, Matija Snuderl, Timothy E Richardson, Anaplastic Transformation in Myxopapillary Ependymoma: A Report of 2 Cases and Review of the Literature, *Journal of Neuropathology & Experimental Neurology*, Volume 79, Issue 10, 2020, Pages 10441053.
11. Hayashi T, Haba R, Kushida Y, Katsuki N, Bando K, Shibuya S, Matsunaga T. Cytopathologic characteristics and differential diagnostic considerations of osteolytic myxopapillary ependymoma. *Diagn Cytopathol*. 2014;42(9): 778-83.
12. Wang M, Wang H, Zhan R, Wan S. Myxopapillary ependymoma in the third ventricle area and sacral canal: dropped or retrograde metastasis? *Neurol Med Chir(Tokyo)*. 2013;53(4): 237-41.
13. Graf M, Blacker H, Otto HF. Extraneural metastasizing ependymoma of the spinal cord. *Pathol Oncol Res*. 1999;5(1): 56-60.
14. Shors SM, Jones TA, Jhaveri, Huckman MS. Best cases from the AFIP: myxopapillary ependymoma of the sacrum. *Radiographics*. 2006;26(Suppl 1): S111-6.
15. Fassett DR, Schmidt MH. Lumbosacral ependymomas: a review of the management of intradural and extradural tumours. *Neurosurg Focus*. 2003;15(5): E13.
16. De Verdellhan, O Haegelen, C Carsin-Nicol, B Riffaud, L Amlashi, S.F.A Brassier, G Carsin, M Morandi, X. (2005). MR imaging features of spinal schwannomas and meningiomas. 32(1), 42-49.
17. Waldron JS, Cha S. Radiographic features of intramedullary spinal cord tumors. *Neurosurgery clinics*. 2006;17(1):13-9.
18. Chin DM, Donaldson SS, Dahl GV, Wilson JD, Huhn SL, Fisher PG. Management of children with metastatic spinal myxopapillary ependymoma using craniospinal irradiation. *Med Pediatr Oncol*. 2000; 35(4):443-5.
19. Sa'adah M, Al Shunnar K, Saadah L, Shogan A, Inshasi J, Afifi H. Atypical presentations of conus medullaris and filum terminale myxopapillary ependymomas. *Journal of Clinical Neuroscience*. 2004;11(3):268-72.
20. Koeller KK, Rosenblum RS, Morrison AL. Neoplasms of the spinal cord and filum terminale: radiologic-pathologic correlation. *Radiographics*. 2000;20(6):1721-49.
21. Wippold FJ, Smirniotopoulos JG, Moran CJ, Suojanen JN, Vollmer DG. MR imaging of myxopapillary ependymoma: findings and value to determine extent of tumour and its relation to intraspinal structures. *AJR Am J Roentgenol* 1995; 165(5):1263-7.
22. Koeller KK, Shih RY. Intradural extramedullary spinal neoplasms: radiologic-pathologic correlation. *Radiographics*. 2019; 39(2):468-90.
23. Albadr F, Albahkali SM, Alahmadi MS, Alsakkaf HM, Al-Habib A, Alkhalidi H, Basalamah AA. Atypical Imaging of Hemorrhagic Lumbosacral Myxopapillary Ependymoma with Histopathological Correlation: A Case Report. *The American Journal of Case Reports*. 2020;21:e925449-1.
24. Omerhodžić I, Pojskić M, Arnautović KI. Myxopapillary Ependymomas. In *Spinal Cord Tumors 2019* (pp. 273-300). Springer, Cham.
25. Patnaik S, Jyotsnarani Y, Uppin SG, Susarla R. Imaging features of primary tumors of the spine: A pictorial essay. *The Indian journal of radiology & imaging*. 2016; 26(2):279
26. Bradley DP, Reddy VB, Cochran E, Gattuso P. Comparison of cytological features of myxopapillary ependymomas on crush preparations. *Diagnostic cytopathology*. 2009; 37(8):607-12.
27. Kulesza P, Tihan T, Ali SZ. Myxopapillary ependymoma: cytomorphic characteristics and

- differential diagnosis. *Diagnostic cytopathology*. 2002; 26(4):247-50.
28. Manasa PL, Uppin MS, Sundaram C. Analysis of squash smear cytology of ependymomas. *Acta Cytol*.2012;56(2):183-8.
 29. Chan HS, Becker LE, Hoffman HJ, Humphreys RP, Hendrick EB, Fitz CR, Chuang SH. Myxopapillary ependymoma of the filum terminale and cauda equine in childhood: report of 7 cases and review of the literature. *Neurosurgery*.1984;14(2):204-10.
 30. Van Hoe W, Mebis W, Bali A, Gielen J, Dekeyser S. Myxopapillary ependymoma of the sacrum. *Acta Neurol Belg*.2020; 120(1): 227-229.
 31. Specht CS, Smith TW, DeGirolami U, Price JM. YUY light and electron microscopic study. *Cancer*.1986 15;58(2): 310-7.
 32. Lamzabi I, Arvanitis LD, Reddy VB, Bitterman P, Gattuso P, Immunophenotype of myxopapillary ependymomas. *Applied*.
 33. Mack SC, Taylor MD. Put away your microscopes: the ependymoma molecular era has begun. *Current opinion in oncology*. 2017; 29(6):443.
 34. Zheng H, Baranova K, Song J, Yan L, Biswas S, Chakrabarti S, Zhang Q. Overexpression of long noncoding RNA HOTAIR is a unique epigenetic characteristic of myxopapillary ependymoma. *Journal of Neuropathology & Experimental Neurology*. 2020; 79(11):1193-202.
 35. Stephen JH, Sievert AJ, Madsen PJ, Judkins AR, Resnick AC, Storm PB, Rushing EJ, Santi M. Spinal cord ependymomas and myxopapillary ependymomas in the first 2 decades of life: a clinicopathological and immunohistochemical characterization of 19 cases. *Journal of Neurosurgery: Pediatrics*. 2012 1;9(6):646-53.
 36. Bates JE, Choi G, Milano MT. Myxopapillary ependymoma: a SEER analysis of epidemiology and outcomes. *J Neurooncol*.2016;129(2):251-8.
 37. Klekamp J. Spinal ependymomas. Part 1: Intramedullary ependymomas. *Neurosurg Focus*. 2015; 39(2):E6.
 38. Tobin MK, Geraghty JR, Engelhard HH, Linninger AA, Mehta AI. Intramedullary spinal cord tumours: a review of current and future treatment strategies. *Neurosurg Focus*. 2015; 39(2):E14.
 39. Ferrante L, Mastronardi L, Celli P, Lunardi P, Acqui M, Fortuna A. Intramedullary spinal cord ependymoma: a study of 45 cases with long-term follow-up. *Acta Neurochir (Wien)*.1992;119(1-4):74-9.
 40. Bagley CA, Wilson S, Kothbauer KF, Bookland MJ, Epstein F, Jallo GI. Long term outcomes following surgical resection of myxopapillary ependymomas. *Neurosurg Rev*.2009; 32(3):321-34;discussion 334.
 41. Fegerl G, Marosi C. Stabilization of metastatic myxopapillary ependymoma with sorafenib. *Rare Tumours*.2012 26;4(3):e42.
 42. Schweitzer JS, Batzdorf U. Ependymoma of the cauda equine region: diagnosis, treatment, and outcome in 15 patients. *Neurosurgery*.1992; 30(2):202-7.
 43. Akyurek S, Chang EL, Yu TK, Little D, Allen PK, McCutcheon I, Mahajan A, Maor MH, Woo SY. Spinal myxopapillary ependymoma outcomes in patients with surgery and radiotherapy at M.D. Anderson Cancer Center. *J Neurooncol*. 2006; 80(2):177-83.
 44. Gilhuis HJ, Kappelle AC, Beute G, Wesseling P, Grotenhuis A, Boerman RH. Radiotherapy for partially resected spinal ependymomas: a retrospective study of 60 cases. *Oncol Rep*. 2003;10(6):2079-82.
 45. Schild SE, Wong W, Nisi K. In regard to the radiotherapy of myxopapillary ependymomas. *Int J Radiat Oncol Biol Phys*.2002;53(3):787.
 46. Scarpelli DB, Turina CB, Kelly PD, Khudanyan A, Jaboin JJ, McClelland III S. National trends in management of adult myxopapillary ependymomas. *Journal of Clinical Neuroscience*. 2020 ;73:162-7.
 47. Chang UK, Choe WJ, Chung CK, Kim HJ. Surgical outcome and prognostic factors of spinal intramedullary ependymomas in adults. *J NeuroOncol* 2002;57: 133-139.
 48. Wahab SH, Simpson JR, Michalski JM, Mansur DB. Long term outcome with post-operative radiation therapy for spinal canal ependymoma. *J Neurooncol*.2007; 83(1):85-9.

49. Rezai AR, Woo HH, Lee M, Cohen H, Zagzag D, Epstein FJ. Disseminated ependymomas of the central nervous system, J Neurosurg. 1996; 85(4):618-24.
50. Akyurek S, Chang EL, Yu TK, Little D, Allen PK, McCutcheon I et al. Spinal myxopapillary ependymoma outcome in patients treated with surgery and radiotherapy at M.D. Anderson Cancer Center. J NeuroOncol 2006;80:177-183.
51. Weber DC, Wang Y, Miller R, Villa S, Zaucha R, Pica A, Poortmans P, Anacak Y, Ozyigit G, Baumert B, Haller G, Preusser M, Li J. Long term outcome of patients with spinal myxopapillary ependymoma: treatment results from the MD Anderson Cancer Center and institutions from the Rare Cancer Network. Neuro Oncol. 2015;17(4):588-95.