

Myxopapillary ependymoma as a cause of back pain in a young male – A case report

Trung P Ngo¹, BSc, DC
 John Dufton^{1,2}, DC, MSc, MD
 Paula J Stern¹, BSc, DC, FCCS(C)
 Omar Islam², MD, FRCPC, DABR

Objective: *Primary spinal cord tumours are rare causes of low back pain but can be a significant cause of morbidity if undiagnosed and untreated. The following is a case of a young male patient presenting with low back pain and radicular symptoms caused by myxopapillary ependymoma.*

Clinical Features: *A nineteen year old male presented to an orthopaedic surgeon with a long history of back pain. He was initially diagnosed with soft tissue injuries and discharged. He began to experience erectile and bowel dysfunction two years later and was re-referred to the orthopaedic surgeon by his family physician but was lost to follow-up. The patient did not present to the surgeon until two years after his symptom profile changed. At that point, MRI examinations revealed a large myxopapillary ependymoma extending from T12 to L4 that was confirmed by a pathologist.*

Intervention and Outcome: *The tumour was surgically resected with subsequent adjuvant radiotherapy. After one year, the patient required continued catheterization and had poor anal tone. His back and leg complaints were almost normal. Follow-up MRI examinations revealed no disease progression or new spinal lesions at 4 years after the initial diagnosis.*

Conclusion: *The clinical presentation of primary spinal cord tumours is non-specific and can easily be missed. In cases of chronic back pain, signs and*

Objectif : *Les tumeurs primaires de la moelle épinière sont des causes rares de lombalgies, mais peuvent être une importante cause de morbidité si elles sont pas diagnostiquées ni traitées. Le cas suivant concerne un jeune homme qui présente une lombalgie et des symptômes radiculaires causés par un épendymome myxopapillaire.*

Caractéristiques cliniques : *Un homme de dix-neuf ans s'est présenté chez un chirurgien orthopédique avec de longs antécédents de douleur dorsale. Il lui a d'abord diagnostiqué des blessures des tissus mous et il a été libéré par la suite. Deux ans après, il a commencé à souffrir de dysfonctionnement érectile et intestinal et a été renvoyé vers le chirurgien orthopédique par le médecin de famille mais a été perdu au suivi. Le patient s'est présenté au chirurgien deux ans après que son profil de symptôme a changé. À ce moment-là, les examens par IRM ont révélés un épendymome myxopapillaire se prolongeant de T12 à L4, confirmé par un pathologiste.*

Intervention et résultat : *La tumeur a été prélevée chirurgicalement à l'aide d'une radiothérapie adjuvante subséquente. Après un an, le patient avait besoin d'un cathétérisme continu et présentait un tonus anal faible. Il ne se plaignait presque plus de douleurs au dos et aux jambes. Les examens de suivi par IRM n'ont pas révélé de progression de la maladie ou de nouvelles lésions de la moelle épinière, quatre ans après le diagnostic initial.*

Conclusion : *La présentation clinique de tumeurs primaires de la moelle épinière n'est pas spécifique et peut facilement ne pas être repérée. Dans les cas de*

¹Canadian Memorial Chiropractic College, Toronto, Ontario, Canada

² Department of Diagnostic Radiology, Kingston General Hospital, Queen's University, Kingston, Ontario, Canada

Corresponding author: Trung P Ngo, Graduate Student, Clinical Sciences, Canadian Memorial Chiropractic College, 6100 Leslie Street, Toronto, Ontario M2H 3J1. Phone: (416) 482-2340 ext. 208. Email: trungptngo@gmail.com

Conflict of interest: none

©JCCA 2013

symptoms should be regularly monitored for changes indicative of progressive neurological compromise such as sensory, motor and bowel/bladder dysfunction. If there is deterioration of clinical signs and symptoms, a spinal tumour should be considered in the list of differential diagnoses. Delayed diagnosis and treatment of these rare causes of back pain could lead to poor outcomes; therefore, a referral to a surgeon should be done immediately with proper follow up to ensure continuity of care.

KEY WORDS: Spinal cord tumour, ependymoma, back pain, case reports

douleurs dorsales chroniques, les signes et symptômes devraient être contrôlés régulièrement dans le cas de changements indiquant des troubles neurologiques progressifs tels que les dysfonctionnements sensoriels, moteurs, intestinaux et érectiles. Dans le cas de dégradation des signes et symptômes cliniques, une tumeur de la moelle épinière devrait être considérée parmi les diagnostics différentiels. Un diagnostic tardif et un traitement de ces causes rares de douleur dorsale peut mener à de pauvres résultats; et un renvoi chez un chirurgien devrait par conséquent être effectué dans les plus brefs délais avec un suivi approprié pour assurer la continuité des soins.

MOTS CLÉS : Tumeurs de la moelle épinière, épendymome, dorsalgie, études de cas

Introduction

Low back pain caused by neoplasia accounts for less than 1% of cases of back pain¹ but can be a significant cause of morbidity if undiagnosed and untreated. Primary spinal cord tumours represent approximately 15% of all neoplasms of the central nervous system (CNS).^{2,3} These tumours pose a diagnostic challenge for practitioners since the symptoms are often non-specific with the most common initial symptom being pain.³

Spinal cord tumours are classified according to their anatomical location and are separated into three broad categories: intramedullary, intradural extramedullary, and extradural.² Intramedullary tumours are predominantly gliomas (astrocytomas and ependymomas), extramedullary tumours are most commonly peripheral nerve sheath tumours or meningiomas, and extradural lesions are usually metastatic. The most common primary spinal cord tumour in adults is ependymoma.^{4,5}

Spinal cord tumours can significantly compress and displace the spinal cord, nerve roots or surrounding structures, impacting the neurologic status.³ Therefore, early recognition of the signs and symptoms of primary spinal cord tumours allows for early treatment, potentially minimizing neurologic morbidity and improving outcome. The following is a case of a young male patient presenting with low back pain and radicular symptoms caused by myxopapillary ependymoma.

Case History

A 19 year old male student presented to an orthopaedic surgeon with complaints of a 3 year history of back pain. He was referred to the surgeon by the family physician. He attributed the onset to being unexpectedly struck in the kidney area, after which he had ongoing recurring back pain. He was physically active but noted increasing pain in his lower back with some vague radicular discomfort into his proximal thighs more towards the end of the day. Systems review was unremarkable. His lumbar spine range of motion was slightly restricted in forward flexion with a mild list to the right. Straight leg raising was to 90° bilaterally with no evidence of nerve root tension signs. Sacroiliac and hip joint dysfunction tests were negative. On neurological examination of the lower limbs, deep tendon reflexes were 3+ but symmetric, his muscle strength was graded 5/5 and he had normal sensation. Palpation revealed L4/5 and L5/S1 local tenderness.

Conventional radiographs and a CT scan showed a spina bifida occulta of S1 but no pathology that would account for his symptomatology. The patient was diagnosed with soft tissue injuries and prescribed NSAIDs and analgesics to be taken on an as needed basis and discharged with no further follow-up scheduled.

Approximately one year later he visited his family physician because he needed a form to be filled out for a motor vehicle accident. When he was asked about his

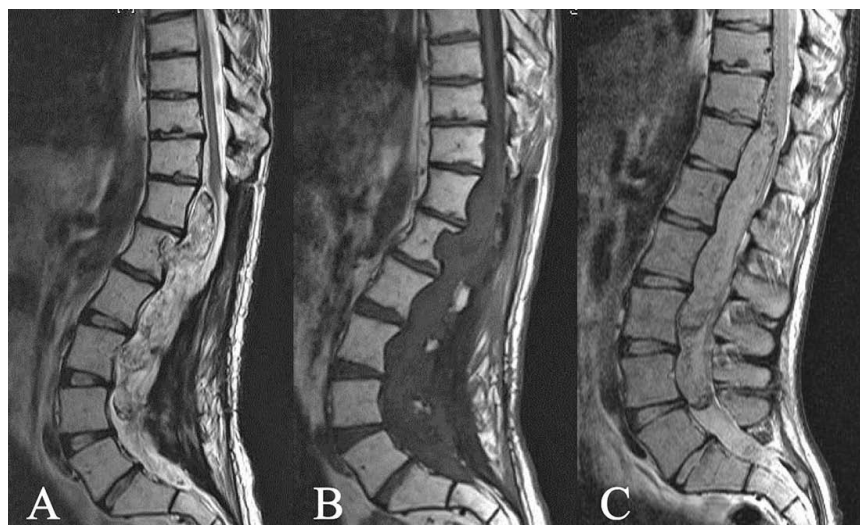


Figure 1: Magnetic Resonance Imaging Mid-Sagittal Lumbar Spine (A) Hyperintense intradural spinal canal mass noted on T2-weighted MRI; (B) The mass was hypointense on T1-weighted MRI and showed avid enhancement (C). Both images demonstrate that the upper extent of the mass is inseparable from the distal spinal cord with a large exophytic component. The spinal canal is expanded with posterior vertebral body scalloping extending from T12 to L4. Intra- and extra-lesional flow voids represent hypervascularity at the upper aspect of the mass surrounding the distal spinal cord.

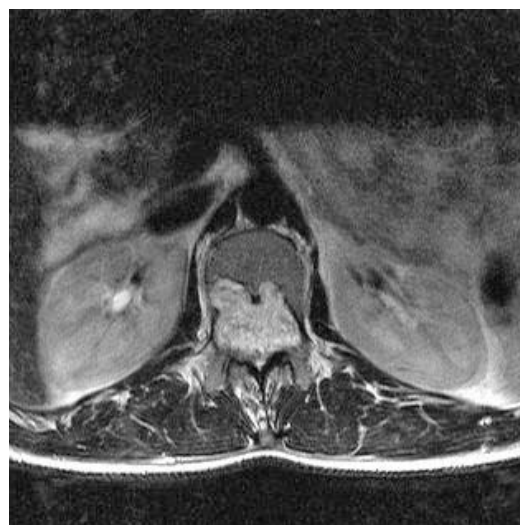


Figure 2: Axial Magnetic Resonance Image Lumbar Spine T2-weighted MRI showing the erosion of the L1 vertebral body

low back pain, he stated that it was episodic but was not troubling him that much overall since he was managing with home exercises, and Diclofenac (Voltaren).

Another year later, at the age of 21, the patient returned to his family physician with similar episodic low back pain, but at this time reported intermittent erectile dysfunction and subjectively stated that he needed to bear down harder in order to have a bowel movement. At this point, he was re-referred to the orthopaedic surgeon; however, he did not follow up with the surgical consultation.

Another two years had elapsed before the patient returned to his family physician for reasons unrelated to his back pain (he had recently broken-up with his partner and was going through a custody battle and experiencing depression). At this point, he was 23 years old. When asked about his low back and the surgical consultation, he stated that he was never contacted by the orthopedic clinic and did not think much about it. He reported that he continued

to experience the erectile and bowel dysfunction so was again referred to the orthopaedic surgeon.

Four months later, the patient was seen by the orthopaedic surgeon. At this consultation, he presented with back pain radiating into his legs with weakness and numbness in his legs and bowel, bladder and erectile dysfunction. He ambulated independently with a limp. His deep tendon reflexes were absent bilaterally whereas they were 3+ four years earlier. The remainder of the physical examination was unremarkable. An MRI examination revealed a large heterogenous, T1 hypointense, T2 hyperintense, enhancing spinal canal mass that invaded multiple neural foramina bilaterally and eroded the posterior one third of the L1 vertebral body. There was posterior vertebral body scalloping extending from T12 to L4 with expansion of the spinal canal (Figures 1 and 2). Pathologic examination confirmed a neuroepithelial neoplasm, favouring myxopapillary ependymoma.

An orthopaedic surgeon and neurosurgeon performed a conjoint procedure that involved T12 to S2 laminectomies, excision of the intradural spinal tumour and a spinal duraplasty. Adjuvant radiotherapy was recommended since the tumour could not be completely resected. Initially post surgery, he had significant bowel, bladder and erectile dysfunction. After one year, he still needed catheterization and had poor anal tone but his back and leg complaints were almost totally alleviated. Multiple bi-yearly follow-up MRI examinations revealed marrow changes secondary to radiation therapy. At 4 years after his initial diagnosis, the most recent post-operative MRI examinations revealed no disease progression or new spinal lesions.

Discussion

Spinal ependymal tumours are glial tumours derived from ependymal cells in the spinal cord and represent 40-60% of primary spinal cord tumours.⁶⁻⁸ The myxopapillary subtype of ependymomas (MPE) occurs mostly in the thoracolumbar region and is the most common form of ependymoma in the lumbar spine^{4,8-10}, accounting for 13% of all spinal ependymomas and 90% of tumours in the conus medullaris⁴⁻¹⁰. It is considered a benign tumour since it is usually encapsulated and anatomically isolated from direct access to lymphatic or other routes of dissemination.⁷ The key pathological characteristics of MPE are mucinous degeneration within the vascular connective tissue cores of papillary tumours.¹¹

MPE usually occurs in the adult population in the third and fourth decades of life and affect males more frequently than females.^{4,12,13} Clinically, the most common finding is lumbar, sacral or radicular pain^{5,14} which is often worse in the recumbent position and, thus, at night⁷. To a lesser extent, sensory changes, motor deficits, bladder abnormalities and impotence/ejaculation dysfunctions also occur.^{5,14} The average duration of symptoms preceding diagnosis ranges from 13 months to 8.3 years^{5,12,15} since they are slow growing. Differential diagnoses for MPE include other primary spinal cord tumours and metastases, or possibly disc herniation depending on the symptoms.⁷

Diagnosis of MPE is best accomplished with magnetic resonance imaging (MRI). MRI findings typically include an intradural mass that tends to be hypointense or isointense with the spinal cord on T1 weighted images,

hyperintense on T2 weighted images and will have intense homogenous enhancement after the administration of intravenous contrast material.^{4-7,11,13} On average, the lesions affect 2 to 4 vertebral body levels.^{4-6,11,13} Confirmation of the diagnosis cannot be made until the excised tumour tissues are examined pathologically.

Treatment for MPE mainly involves surgical excision of the tumour. If the capsule ruptures or the tumour is not confined to the filum terminale, the mass could infiltrate and adhere to the cauda equina and/or conus medullaris or disseminate via the cerebral spinal fluid.^{5,11} Therefore, adjuvant radiotherapy is recommended when en bloc excision (removal of the entire tumour as one piece) cannot be accomplished.^{11,12} However, the efficacy of radiation therapy has not been established and can result in adverse effects such as radiation myelopathy^{5,12} and residual dysuria¹¹. The 5-year survival rate of spinal ependymomas ranges from 57-100%^{12,14,15} and 10-33% of patients will experience local invasion of the tumour or recurrence^{8,14}. Metastasis is rare in MPE but there have been several reported cases.¹⁶⁻²⁹ A good prognosis is correlated with both a greater extent of resection⁵ and a high preoperative functional status². Longer symptom duration prior to treatment is correlated with poorer functional outcomes³⁰; therefore, early detection of the tumour is also associated with a better prognosis^{11,30}.

In the chiropractic literature, three cases of spinal ependymomas have been published in two articles.^{31,32} One patient presented with chronic (six months duration) back pain whereas the other two presented with acute exacerbations of back pain. The clinical presentations were variable but each had an unusual characteristic: one had atrophy and weakness of the left hand; the second had bowel and bladder dysfunction; and the third experienced a disproportionate amount of pain relative to the mechanism of his injury. Ependymomas in the cervical spine (in the first case) and lumbar spine (in the latter two cases) were revealed via MRI and all cases had good outcomes after surgical excision of the tumour. The subtypes of ependymomas were not discussed in these three cases.

In the present case, it is unlikely that the initial presentation of chronic low back pain was associated with the ependymoma. It could be argued that since MPE is a slow growing tumour, it could have begun to develop at that time. However, he did not report any red flags, the physical examination did not reveal any gross abnormalities and

the conventional radiographs and CT scan were negative. Although his reflexes were 3+ in the lower extremities initially, this is not necessarily indicative of disease.³³ If the original low back pain was indeed of a soft tissue nature as opined by the orthopaedic surgeon, then this case reinforces the necessity to regularly re-evaluate patients for changes in signs and symptoms.

The patient's symptom profile changed between the ages of 20 and 21 when he reported erectile and bowel dysfunction to the family physician. No physical examination was performed during that visit so it cannot be determined when signs of neurological deterioration would have appeared. He was referred back to the orthopaedic surgeon but unfortunately, no appointment occurred. Treatment may have been further delayed had he not returned to the family physician for unrelated reasons 2 years later. At the time of the second surgical consultation, the patient had weakness in his legs, bowel and bladder dysfunction, erectile dysfunction and night pain. Physical findings further revealed a change from 3+ reflexes bilaterally to absent reflexes.

This delayed diagnosis perhaps contributed to his slow healing and residual bladder and anal tone dysfunction after treatment. This highlights the importance of follow-up by the primary contact provider to ensure proper management of the case. If the patient had seen the specialist closer to the time of initial symptom change, surgical resection could have been performed sooner, perhaps resulting in a better clinical outcome.

Conclusion

Myxopapillary ependymomas are the most common primary tumours in the region of the conus medullaris and filum terminale. The clinical presentation of these tumours is non-specific with lumbar and radicular pain being the most common symptoms. In cases of chronic back pain, signs and symptoms should be regularly monitored for changes indicative of progressive neurological compromise such as sensory, motor and bowel/bladder dysfunction. If there is deterioration of clinical signs and symptoms, a spinal tumour should be considered in the list of differential diagnoses. Delayed diagnosis and treatment of these rare causes of back pain could lead to poor outcomes; therefore, a referral to a surgeon should be done immediately with proper follow up to ensure continuity of care.

References

1. Jarvik JG, Deyo RA. Diagnostic evaluation of low back pain with emphasis on imaging. *Annals Internal Medicine*. 2002; 137:586-97.
2. Grimm S, Chamberlain MC. Adult primary spinal cord tumours. *Expert Review of Neurotherapeutics*. 2009; 9:1487-95.
3. Arnautovic K, Arnautovic A. Extramedullary intradural spinal tumours: A review of modern diagnostic and treatment options and a report of a series. *Bosnian J Basic Medical Sciences*. 2009; 9(Supplement):S40-S45.
4. Choi JY, Chang KH, Yu IK, et al. Intracranial and spinal ependymomas: Review of MR images in 61 patients. *Korean J Radiol*. 2002; 3:219-228.
5. Sakai Y, Matsuyama Y, Katayama Y, et al. Spinal myxopapillary ependymoma: Neurological deterioration in patients treated with surgery. *Spine*. 2009; 34:1619-1624.
6. 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. *Am J Radiol*. 1995; 165:1263-67.
7. Bagley CA, Wilson S, Kothbauer KF, Bookland MJ, Epstein F, Jallo GI. Long term outcomes following surgical resection of myxopapillary ependymomas. *Neurosurg Rev*. 2009; 32:321-334.
8. Bavbek M, Altinors MN, Caner HH, Bilezikci B, Agildere M. Lumbar myxopapillary ependymoma mimicking neurofibroma. *Spinal Cord*. 2001; 39:449-452.
9. Sonneland PR, Scheithauer BW, Onofrio BM. Myxopapillary ependymoma: A clinicopathologic and immunohistochemical study of 77 cases. *Cancer*. 1985; 56:883-93.
10. Celli P, Cervoni L, Cantore G. Ependymoma of the filum terminale: Treatment and prognostic factors in a series of 2 cases. *Acta Neurochir*. 1993; 124: 99-103.
11. Nakamura M, Ishii K, Watanabe K, et al. Long-term surgical outcomes for myxopapillary ependymomas of the cauda equine. *Spine*. 2009; 34:E756-760.
12. Volpp PB, Han K, Kagan AR, Tome M. Outcomes in treatment for intradural spinal cord ependymomas. *Int J Radiation Oncology Biol Phys*. 2007; 69:1199-1204.
13. Sun B, Wang C, Wang J, Liu A. MRI features of intramedullary spinal cord ependymomas. *J Neuroimaging*. 2003; 13:346-351.
14. Hanbali F, Fourney DR, Marmor E, et al. Spinal cord ependymoma: Radical surgical resection and outcome. *Neurosurgery*. 2002; 51:1162-74.
15. Asazuma T, Toyama Y, Suzuki N, et al. Ependymomas of the spinal cord and cauda equine: An analysis of 26 cases and a review of the literature. *Spinal Cord*. 1999; 37:753-59.
16. Friedman DP, Hollander MD. Neuroradiology case of the day. *Radiographics*. 1998; 18:794-98.

17. Patterson RH Jr, Campbell WG Jr, Parsons H. Ependymoma of the cauda equina with multiple visceral metastases. *J Neurosurg.* 1961; 18:145–150.
18. Agapitos E, Kavantzias N, Karaitianos J, Davaris P. Subcutaneous sacrococcygeal myxopapillary ependymoma: a case report. *Archives d’anatomie et de cytologie pathologiques.* 1995; 43:157–159.
19. Al Moutaery K, Aabed MY, Ojeda VJ. Cerebral and spinal cord myxopapillary ependymomas: a case report. *Pathology.* 1996; 28:373–376.
20. Helwig EB, Stern JB. Subcutaneous sacrococcygeal myxopapillary ependymoma: a clinicopathologic study of 32 cases. *Am J Clin Pathol.* 1994; 81:156–161.
21. Ilhan I, Berberoglu S, Kutluay L, Maden HA. Subcutaneous sacrococcygeal myxopapillary ependymoma. *Medical and Pediatric Oncology.* 1998; 30:81–84.
22. Kline MJ, Kays DW, Rojiani AM. Extradural myxopapillary ependymoma: report of two cases and review of the literature. *Pediatric Pathology and Laboratory Medicine.* 1996; 6:813–822.
23. Kramer GW, Rutten E, Sloof J. Subcutaneous sacrococcygeal ependymoma with inguinal lymph node metastasis. *J Neurosurg.* 1988; 68:474–477.
24. Pulitzer DR, Martin PC, Collins PC et al. Subcutaneous sacrococcygeal (“myxopapillary”) ependymal rests. *Am J Surgical Pathology.* 1988; 12:672–677.
25. Woesler B, Moskopp D, Kuchelmeister K et al. Intracranial metastasis of a spinal myxopapillary ependymoma. A case report. *Neurosurgery Review.* 1998; 21:62–65.
26. Graf M, Blaeker H, Otto HF. Extranural metastasizing ependymoma of the spinal cord. *Pathology Oncology Research.* 1999; 5:56–60.
27. Mavroudis C, Townsend JJ, Wilson CB. A metastasizing ependymoma of the cauda equine: case report. *J Neurosurg.* 1977; 47:771–775.
28. Rubinstein LJ, Logan WJ. Extranural metastases in ependymoma of the cauda equina. *J Neurol Neurosurg Psychiatry.* 1970; 33:763–770.
29. Rickert CH, Kedziora O, Gullotta F. Ependymoma of the cauda equine. *Acta Neurochir.* 1999; 141:781–2.
30. Quigley DG, Farooqi N, Pigott TJD, et al. Outcome predictors in the management of spinal cord ependymoma. *Eur Spine J.* 2007; 16:399–404.
31. O’Shaughnessy J, Bussieres A. Subtle clinical signs of a spinal cord ependymoma at the cervicothoracic level in an adult: A case report. *JCCA.* 2006; 50:244–48.
32. Lensgraf AG, Young KJ. Ependymoma of the spinal cord presenting in a chiropractic practice: 2 case studies. *J Manipulative Physiol Ther.* 2006; 29:676–81.
33. Bickley LS, Szilagy PG. *Bates’ Guide to Physical Examination and History Taking.* 8th ed. Philadelphia: Lippincott Williams & Wilkins; 2003