*Acta Orthop. Belg.*, 2008, **74**, 282-288

*CASE REPORT*

# Intradural tumours of the lumbar spine presenting with low back pain : Report of two cases and review of the literature

Pavlos KATONIS, George KONTAKIS, Dritan PASKU, Michael TZERMIADIANOS, George TZANAKAKIS, Alexander HADJIPAVLOU

*From the University Medical School of Crete, Heraklion, Greece*

**Two cases of spinal cord tumours (one schwannoma and one ependymoma) of the lumbar spine are reported. The treatment with radical excision and posterolateral fusion, along with adjuvant radiation therapy in the case with ependymoma was successful, with follow-up of six and seven years respectively.**
**A literature review is presented, and a possible presentation with low back pain is analysed.**

**Keywords** : intradural ; tumours ; low back pain.

## INTRODUCTION

Intradural tumours of the spine are usually benign and carry a good prognosis, if they are diagnosed and removed early. Early symptoms of these tumours are non-specific and their progression can be subtle. The duration of symptoms usually ranges from 3 to 4 years prior to the diagnosis. Low back pain may be the only apparent symptom during this time (*19*).

Schwannomas are the most common intradural tumours with an incidence of 29% (*26*). They are usually benign, but can be locally aggressive and cause catastrophic neurological compromise. They arise from the sheath of the spinal nerve roots and are occasionally seen in the cauda equina (*3*). Intramedullary ependymomas are the most common spinal cord tumours in adults, representing 34.5% of all central nervous system tumours and approximately 60% of all intramedullary tumours respectively (*20,25*). They arise from the ependymal cells in the central nervous system and the cauda equina.

These tumours have the unusual ability to disseminate via the cerebrospinal fluid. Haematogenous dissemination is far less common, seen in about 1% of cases (*1,6*).

We report two cases of low back pain, caused by schwannoma and ependymoma of the lumbar spine, treated with radical excision and posterolateral fusion, along with adjuvant radiation therapy in the patient with ependymoma.

■ Pavlos Katonis, Assistant Professor of Orthopaedics.
■ George Kontakis, Assistant Professor of Orthopaedics.
■ Dritan Pasku, Resident in Orthopaedics.
■ Michael Tzermiadianos, Orthopaedic Surgeon.
■ Alexander Hadjipavlou, Professor of Orthopaedics.
*Orthopaedic Department University of Crete, Heraklion, Greece.*
■ George Tzanakakis, Associate Professor of Histology.
*Medical School University of Crete, Heraklion, Greece.*
Correspondence : Pavlos Katonis, 277 Knossou Av, 71409 Heraklion, Greece. E-mail : katonis@hol.gr




*No benefits or funds were received in support of this study*

## CASE REPORT

### Case 1

A 56-year old woman presented in January 2000 with a history of low back pain of three months duration. Previous treatment by means of NSAIDs and physiotherapy was ineffective. The pain was dull and aching, deteriorating over time and interfered with her normal daily activities. It was not related to physical activities and was worse at night, awaking the patient. During the last five months she gradually developed numbness and pain in her left lower extremity.

Plain radiographs and routine laboratory investigation were normal. Bone scan did not reveal areas of increased uptake and CT-scan showed only degenerative changes in the facet joints. MRI examination of the lumbar spine revealed a well-demarcated intradural mass (1.5 cm × 2 cm), at the level of the L3 vertebra, which compressed and displaced the roots of the cauda equina. No bone erosion was noted. The lesion was hypointense relative to the normal cord on T1-weighted MRI images and showed some heterogeneity and hyperintensity on T2-weighted MRI images. Heterogeneous enhancement was noted after IV contrast administration (fig 1). Schwannoma was the probable diagnosis, but the differential diagnosis also included ependymoma, meningioma and glioma.

During the investigation period (about one month after her admission and while the decision to carry out surgical excision of the tumour was taken) the patient developed left sciatica with numbness over the medial aspect of the left leg and the big toe. Progressive deterioration was noted over a few days, to the point that the patient was unable to walk without using a walking aid. Neurological examination revealed left leg weakness (iliopsoas 2+, quadriceps 3+) and hypoaesthesia. The left patellar reflex was diminished. There was no associated neurological deficit in the right leg.

During surgery the dura was widely exposed after wide laminectomy of L2 and L3 vertebrae, through a posterior midline incision. Once the dura was incised, a whitish-yellow, elastic nodule compressing and laterally displacing the nerve roots

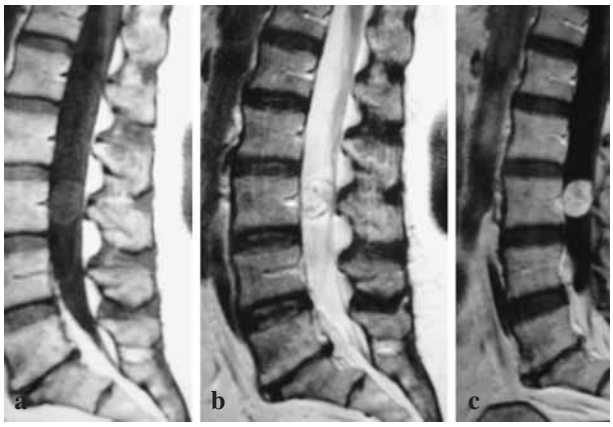


*Fig. 1.* — Sagittal T1-weighted image showing a tumour hypointense to normal cord (a). On T2-weigheted image the tumour shows some heterogeneity and hyperintensity (b). It enhanced intensely but heterogeneously after contrast administration (c).

was evident. One of the nerve roots was adherent to the tumour, and was sacrificed to achieve complete resection. After tumour removal, a posterolateral fusion from L1 to L4 was performed to stabilise the spine, using laminar hooks of the Varigrip spinal system (Advanced Spine, Irvine, CA) and autologous iliac bone mixed with coralline hydroxyapatite (fig 2). At histological examination the lesion exhibited compact fascicular tissue, consisting of elongated spindle cells disposed in fascicles. The nuclei were long, sometimes club-shaped, with occasional palisading. The diagnosis of schwannoma was confirmed (fig 3).

Pain resolved immediately after surgery. Quadriceps weakness mandated the use of a thoracolumbosacral orthosis along with a knee locking orthosis because of persisting quadriceps weakness during the immediate postoperative period. Gradual improvement of muscle strength was evident over the next three months, and there was no evidence of recurrence. Seven years after surgery, the patient had normal daily activities, with no neurological deficit or pain.

### Case 2

A 25-year old male presented in October 2001 with a history of intermittent dull back pain of

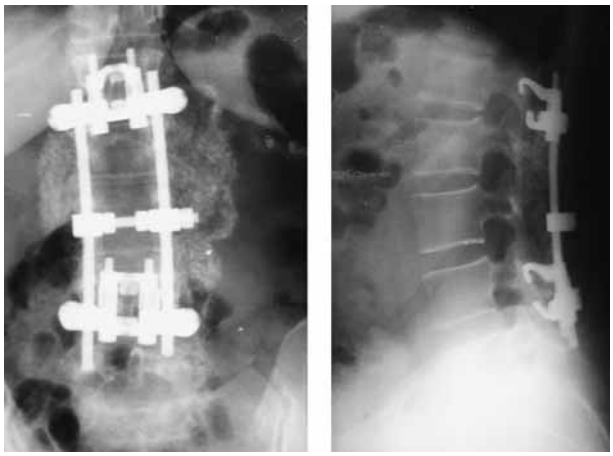

*Fig. 2.* — Postoperative plain radiographs showing wide laminectomy and posterolateral fusion with laminar hooks on L1 and L4.

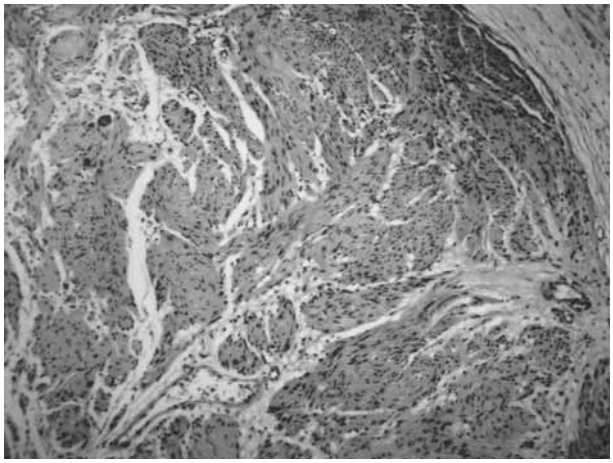

*Fig. 3.* — Elongated spindle cells arranged in fascicles, without cytologic atypia.

1.5 year duration. The pain was initially localised to the L1-L2 level and interfered with his sleep. A few months later the patient developed numbness and dysesthaesia initially over the distal part of the lower extremities, which gradually progressed proximally. One year after the onset of symptoms, she experienced severe, constant burning back and leg pain. Pain was not associated with any muscle weakness or bowel and bladder dysfunction.

MRI revealed a cylindrical, elongated mass (1.2 cm × 4.5 cm), isointense relative to the normal

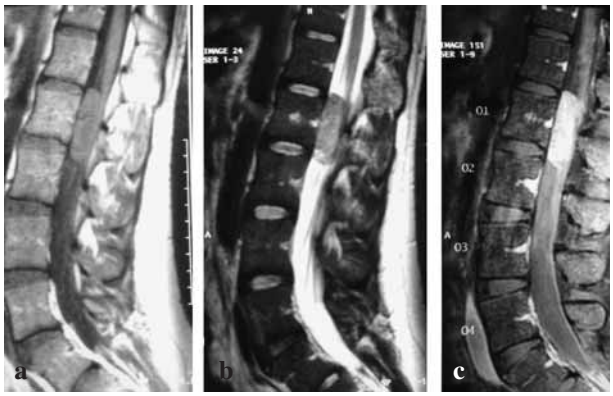


*Fig. 4.* — Sagittal T1-weighted MR image showing a cylindrical, elongated mass (1.2 cm × 4.5 cm), isointense relative to the normal cord (a). On T2-weighted image, the tumour is slightly hyperintense (b). As seen on both T1 and T2 images it is located just below the conus medullaris at the spinal level of the L1 vertebra. After contrast administration it shows a fairly intense enhancement with well-defined borders (c).

cord on MRI images. On MRI, the tumour was slightly hyperintense. It was located just below the conus medullaris at the level of the L1 vertebra. After contrast administration it showed a fairly intense enhancement with well-defined borders (fig 4). Differential diagnosis included ependymoma, astrocytoma and schwannoma.

At surgery, the dura was widely exposed after wide laminectomy from L1 to L2, through a posterior midline incision. After the dura was incised, a smooth reddish gray glistening tumour was visible, which was clearly demarcated from the surrounding spinal cord. The tumour appeared to be an ependymoma rather than an astrocytoma, because it displayed the characteristic feature of a vascular tumour, with blood vessels crossing its surface. Once the tumour was identified, the small feeding vessel and fibrous adhesions between the tumour and the spinal cord were cauterised and the tumour was debulked. After excision of the tumour, the spine was stabilised by a posterolateral fusion from T12 to L3, using a combination of hooks and screws of the Varigrip-Varifix spinal system (Advanced Spine, Irvin, CA) (fig 5). Histological examination revealed a cellular neoplasm, consisting of ovoid or round cells with eosinophilic cytoplasm and uniform, moderately hyperchromatic nuclei with finely dispersed chromatin and clear

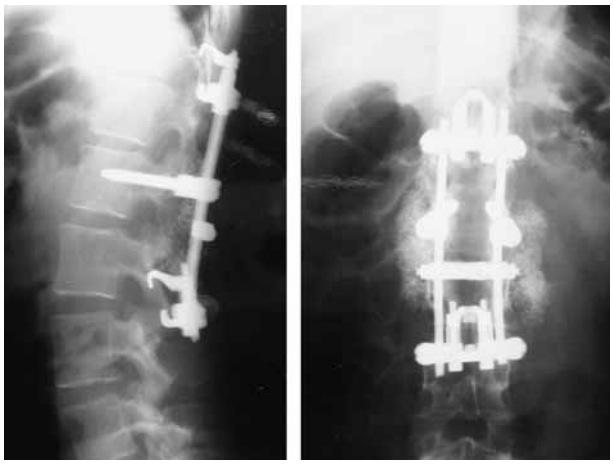

*Fig. 5.* — Post-operative plain radiographs showing wide laminectomy and posterolateral fusion T12-L3 by means of hook and screws combination.

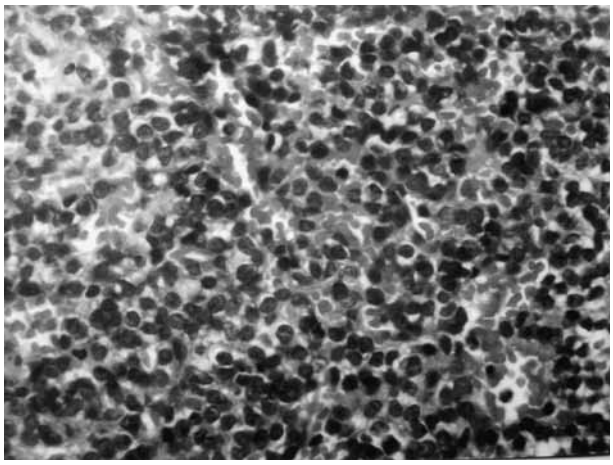

*Fig. 6.* — Histological section of the tumour showing an area of dense cellularity with ovoid to round cells with moderately hyperchromatic nuclei and clear nuclear borders. There is dense vascularity with a perivascular pseudorosette.

nuclear borders. The tumour was hypervascular with a small number of perivascular pseudorosettes. The diagnosis of an ependymoma was made (fig 6).

The patient ambulated with a thoracolumbosacral brace four days after surgery. Fifteen days after surgery, adjuvant radiotherapy was given at the dose of 40 Gy in 2 Gy daily fractions. During the follow-up period of 6 years, the patient was free of symptoms and repeated MRI examinations did not demonstrate any evidence of tumour recurrence.

## DISCUSSION

Schwannomas are benign tumours, which arise as an eccentric growth from the cells of the peripheral or cranial nerves (excepting the optic and olfactory nerves) and spinal roots sheath. They have also been described to occur at various other sites such as skin, oral cavity, and lacrymal glands. Schwannomas are usually solitary but can also be found at multiple sites along the same nerve sheath. They usually occur intradurally, extradurally and occasionally intra and extradurally. The location of the tumour can be anywhere along the neuroaxis but it has a major predilection for the lumbosacral area (40%), which is followed by the thoracic and the cervical spine area (*3,14*).

The main diagnostic problem in our cases was the clinical presentation as non- specific low back pain. This symptom may be related with a silent tumour, and thus warrants further investigation including laboratory screening along with the imaging of the spine. The presence of solitary schwannoma deserves complete MRI examination of the neuroaxis due to its multiple origins, even in the absence of neurofibromatosis (*3*).

The treatment of schwannoma is controversial (*11*). Some authors recommend complete excision of this benign but locally aggressive tumour, and sacrifice of the nerve root, because they consider that the nerve root is non- functional and the neurological status of the patient will not be compromised. Additionally inadequate removal of the tumour leads to an increased risk of recurrence, which is more difficult and dangerous to treat at a later stage (*21*). Kotura *et al* (*13*) supported preservation of the nerve root, even if the tumour is not resected completely because of the risk of neurological deficit. Kim *et al* (*12*) reported that the involved nerve roots are non-functional at the time of surgery and the risk of further neurological deficit after sacrificing these nerve roots small. We prefer complete excision of the tumour, even if this entails the sacrifice of the adherent nerve root.

The other problem one faces in the management of these tumours is the post excision instability of the spine. Review of the literature revealed that surgical resection of the cauda equina tumours produced an unstable spine in half of the cases. In our first case, we performed spinal fusion because of the wide laminectomy and partial removal of facets, which was done in this patient with preexisting degenerative spinal stenosis. In the second case, the wide laminectomy concerned the transitional thoracolumbar area and consequently additional fixation was considered essential (*8*).

Ependymoma is the most common tumour among the adult intramedullary spinal cord tumours (IMSCT). The next most common IMSCT tumour in adults is astrocytoma (24%) (*6,19*). This uncommon and usually nonmalignant tumour arises from the ependymal cells in the central nervous system and cauda equina. Ependymoma has the unusual ability to disseminate via the cerebrospinal fluid. Haematogenous dissemination is far less common : it is seen in about 1% of cases. All patients with the diagnosis of ependymoma should have a thorough investigation of their entire central nervous system, including the cauda equina region, as the tumour has a tendency to disseminate both in cranial and caudal direction via the cerebrospinal fluid (*19*).

Imaging studies in the diagnostic workup of an IMSCT may be difficult to interpret because of the slow rate of growth of the tumour. Widening of the spinal canal or the interpedicular distance is a valuable warning sign. The reactive marginal osteosclerosis can be evaluated more accurately on the CT scans. MRI has revolutionised the detection and the diagnosis of intramedullary spinal cord tumours. With its multiplanar capabilities and high contrast resolution, it allows the identification and characterisation of the lesion in a non-invasive fashion. IV contrast enhancement allows further delineation of the mass, differentiating the tumours from oedema and cysts. The exact histological diagnosis of the tumours remains elusive on the basis of MRI signal intensity and the enhancement alone, although the characteristic imaging features are suggestive of the diagnosis (*19*).

In the future, newer and more sophisticated pulse sequences, such as magnetic resonance spectroscopy (MRS) may have a vital role in the diagnosis of these tumours and particularly in excluding other diagnoses such as demyelinisation, inflammation, or infarction. Other sequences of importance are the fast fluid-attenuated inversion recovery (FLAIR), diffusion-weighted imaging (DWI), diffusion tensor imaging (DTI), short – inversion – time inversion recovery (STIR), and the magnetisation transfer – gradient echo (MT-GE) (*17*).

The role of surgery in the management of intramedullary tumours has evolved significantly in the recent years. Once employed for the diagnosis alone, surgery has become the most effective form of treatment for these well-circumscribed benign tumours (*2,4,14,18*). Since the majority of these intramedullary spinal cord neoplasms are low-grade lesions, microsurgical removal alone results in their long-term control and cure while preserving the neurological functions (*2,4,18*). Ependymomas although not encapsulated, are non-infiltrating lesions and typically display a distinct plane. Therefore a gross-total removal of these tumour results in optimal disease control (*14*).

Surgery, in combination with post-operative radiation therapy for the treatment of ependymomas has provided up to 50-100% ten-year survival rates, even with significant local failure (*5,16*). The most unusual feature of radiation therapy in the case of intramedullary spinal tumours is that the target tissue and the normal tissue are exposed to the same risk of radiation damage. The review of the data revealed that a radiation dose of 45 Gy in 22-25 fractions does not result in myelopathy. It is concluded that the spinal cord can be irradiated safely in doses less than 55 Gy.

Ependymomas are of low-grade nature and amenable to radical resection ; they can be simply observed. The evidence which supports the post-operative radiation after subtotal resection is difficult to be evaluated as it is based on a small sample size, limited follow-up with inadequate or no matched controls treated without radiation therapy. Complete resection alone in the case of spinal cord ependymoma can achieve excellent local control

and survival. Patients should have complete resection of the tumour if technically possible. Postoperative radiotherapy is not recommended after complete resection. For incomplete resection, postoperative local radiotherapy is recommended and it can also achieve excellent local control and survival (*9,15,24*). Local radiotherapy with 50-60 Gy is effective and safe. Salvage radiotherapy improves quality of life for patients with leptomeningeal local infiltration. Doses less than 40 Gy are clearly too low, but local failures have been reported with doses as high as 55 Gy which also significantly increase the risk of myelopathy with the conventional fractioning (*7,22,23*). We thus consider that the evidence is not strong enough to recommend post-operative radiation therapy in these types of ependymomas.

Research in this field, also suggests that the strongest predictor of the post-operative functional outcome in cases of intramedullary tumours, is the preoperative functional capability of the individual patient (*2,4,14,18*). Surgical morbidity is also higher in patients with more significant preoperative deficits (*10,14*).

In conclusion, the spine surgeon should always keep in mind that in the differential diagnosis of low back pain, intradural spine tumours are included and that despite complete resection, these benign intramedullary tumours present a continued risk of recurrence. Long term clinical and radiographic follow-up is thus warranted in these patients. An early post-operative MRI scan, at 6-8 weeks postoperatively can not only establish the completeness of the resection but can also serve as a baseline for future comparisons. Serial gadolinium enhanced MRI scans should be obtained on an annual basis as the radiographic recurrence usually precedes clinical symptoms. We also suggest that a wide laminectomy in the thoracolumbar or lumbar area may cause iatrogenic instability and a spinal fusion should be undertaken.

## REFERENCES


1. **Cassidy JR, Ducker TB, Dienes EA.** Intradural tumours. In : Frymoyer JW. *The Adult Spine Principles and Practice.* Lippincott- Raven, Philadelphia, 1997, pp 1015-1029.
2. **Cooper PR.** Outcome after operative treatment of intramedullary spinal cord tumours in adults : intermediate and long-term results in 51 patients. *Neurosurgery* 1989 ; 25 : 855-859.
3. **Daras M, Koppel B, Heise C, Mazzeo MJ, Poon TP, Duffy KR.** Multiple spinal intradural schwannomas in the absence of Von Recklinghausen's disease. *Spine* 1993 ; 18 : 2556-2559.
4. **Epstein FJ, Farmer JP, Free D.** Adult intramedullary spinal cord ependymomas : the results of surgery in 38 patients. *J Neurosurg* 1993 ; 79 : 204-209.
5. **Garrido E, Stein BM.** Microsurgical removal of intramedullary spinal cord tumours. *Surg Neurol* 1977 ; 7 : 215-219.
6. **Graf M, Blacker H, Otto H.** Extraneural metastasing ependymoma of the spinal cord. *Pathol Oncol Res* 1999 ; 5 : 56-60.
7. **Halperin EC.** Concerning the inferior portion of the spinal radiotherapy field for malignancies that disseminate via the cerebrospinal fluid. *Int J Radiat Oncol Biol Phys* 1993 ; 26 : 357-362.
8. **Iida Y, Kataoka O, Sho T *et al.*** Postoperative lumbar spine instability occurring or progressing secondary to laminectomy. *Spine* 1990 ; 15 : 1186-1189.
9. **Isaacson SR.** Radiation therapy and the management of intramedullary spinal cord tumours. *J Neuro-Oncol* 2000 ; 47 : 231-238.
10. **Kara-Terki R, Gaudin P, Brun F, Juvin R**, **Pasquier B**, **Phelip X.** Bone metastases from a cerebral and sacral ependymoma. Report of a case. *Joint Bone Spine* 2000 ; 67 : 471-474.
11. **Kayaga K, Abe E, Sato K *et al.*** Giant cauda equina Schwannoma. A case report. *Spine* 2000 ; 2 : 268-272.
12. **Kim P, Ebersold MJ, Onofrio BM, Quast LM.** Surgery of spinal nerve schwannoma. Risk of neurological deficit after resection of involved root. *J Neurosurg* 1989 ; 71 : 810-814.
13. **Kotoura Y Shikata J Yamamoto T.** Radiation therapy for giant intrasacral schwannoma. *Spine* 1991 ; 16 : 239-242.
14. **Lewis TT, Kingsley PTE.** Magnetic resonance imaging of multiple spinal neurofibromata-neurofibromatosis. *Neuroradiology* 1987 ; 29 : 562-564.
15. **Lin YH, Huang CI, Wong TT *et al.*** Treatment of the spinal cord ependymomas by surgery with or without radiotherapy. *J Neuro-Oncol* 2005 ; 71 : 205-210.
16. **Linstadt DE, Wara WM, Leibel SA, Gutin PH**, **Wilson CB**, **Sheline GE.** Postoperative radiotherapy of primary spinal cord tumours. *Int J Radiat Oncol Biol Phys* 1989 ; 16 : 1397-1403.
17. **Lowe GM.** Magnetic resonance imaging of intramedullary spinal cord tumours. *J Neuro-Oncol* 2000 ; 47 : 195-210.
18. **McCormick PC, Stein BM.** Intramedullary tumors in adults. *Neurosurg Clin North Am* 1990 ; 1 : 609-630.
19. **Miller DC.** Surgical pathology of intramedullary spinal cord neoplasm's. *J Neuro-Oncol* 2000 ; 47 : 189-194.

**20. Mork SJ, Loken AC.** Ependymoma : a follow-up study of 101 cases. *Cancer* 1977 ; 40 : 907-915.
**21. Santi MD, Mitsunaga MM, Lockett JL.** Total sacrectomy for a giant sacral schwannoma. A case report. *Clin Orthop* 1993 ; 294 : 285-289.
**22. Scott M.** Infiltrating ependymomas of the cauda equina. Treatment by conservative surgery plus radiotherapy. *J Neurosurg* 1974 ; 41 : 446-448.
**23. Schultheiss TE, Stephens LC.** The pathogenesis of radiation myelopathy : widening the circle. *Int J Radiat Oncol Biol Phys* 1992 ; 23 : 1089-1091.
**24. Schultheiss TE, Kun LE, Ang KK, Stephens LC.** Radiation response of the central nervous system. *Int J Radiat Oncol Biol Phys* 1995 ; 31 : 1093-1112.
**25. Schwartz TH, McKormick PC.** Intramedullary ependymomas : clinical presentation, surgical treatment strategies and prognosis. *J Neuro-Oncol* 2000 ; 47 : 211-218.
**26. Sloof JL, Kernohan JW, MacCarty CS.** *Primary Intramedullary Tumours of the Spinal Cord and Filum terminale*. WB Saunders, Philadelphia, 1964, pp 31-61.