**Scholars Academic Journal of Biosciences (SAJB)** **ISSN 2321-6883 (Online)**
**Sch. Acad. J. Biosci., 2017; 5(9):674-681** **ISSN 2347-9515 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI : 10.36347/sajb.2017.v05i09.016

# Tumors of the Spinal Cord: Histopathological and Radiological Correlation with Review of Literature

**Krishna Reddy CH[1], Bheemavathi A[2], Durga K[3].**
[1]Assistant Professor MNR Medical College, Hyderabad, India
[2]Consultant Pathologist, Maxcure Hospitals, Hyderabad, India
[3]Professor, Osmania medical college, Hyderabad, India

***Corresponding author**
*Krishna Reddy CH*

**Article History**
*Received: 04.09.2017*
*Accepted: 09.09.2017*
*Published:30.09.2017*

**Abstract:** Primary spinal cord tumors constitute 10% to 15% of all primary central nervous system tumors. Lesion outside the dura are termed extradural and lesions within dura are called intradural. Intradural lesions are divided into two categories, depending on whether they involve the substance of the spinal cord [intramedullary] or are outside the spinal cord but within the dura [extramedullary]. Tumors within the cord are referred to as intramedullary and account for 10-15% of spinal tumors. Intramedullary tumors consist mainly of astrocytomas and ependymomas making up to 70% of intramedullary tumors. Ependymomas are the most common glial tumor in adults, whereas astrocytomas are the most common intramedullary tumor in children. In our study, total no of spinal tumors encountered were 45 with males constituting 22 cases (49%) and females constituting 23 cases (51%) with almost equal preponderance. Most common symptom was Motor weakness followed by Pain, Sensory disturbance and Bladder dysfunction. Tumors presented in our study includes Schwannomas 15 cases (33%), Neurofibromas 10 cases (23%), Ependymoma 5 cases (11%), Meningiomas 5 cases (11%), Astrocytoma 4 cases (9%), Teratomas 2 cases (4%), and others. As any tumour can occur in spinal cord broad differentials are kept in mind before giving final diagnosis, as correct diagnosis helps in the treatment and prognosis.
**Keywords:** Astrocytoma, Intramedullary, Schwannoma, Spinal cord, Tumors

## INTRODUCTION

Primary spinal cord tumors constitute 10% to 15% of all primary central nervous system tumors [1]. Lesion outside the dura are termed extradural and lesions within dura are called intradural. Intradural lesions are divided into two categories, depending on whether they involve the substance of the spinal cord [intramedullary] or are outside the spinal cord but within the dura [extramedullary]. Tumors within the cord are referred to as intramedullary and account for 10-15% of spinal tumors [2,3]. Intramedullary tumors consist mainly of astrocytomas and ependymomas making up to 70% of intramedullary tumors. Other lesions include hemangioblastoma, paraganglioma and cystic lesions. Ependymomas are the most common glial tumor in adults, whereas astrocytomas are the most common intramedullary tumor in children [2,3]. Astrocytomas are common in the thoracic region whereas ependymomas are common in cervical region. Tumors between the cord and the dura are referred to as intradural extramedullary and account for 70-90 % of primary intradural spine tumors. Primary intradural extramedullary lesions include schwannomas, neurofibromas, dermoid cysts, neurenteric cysts and meningiomas. Of these schwannomas are common followed by meningiomas [4]. Schwannomas are common in cervical and lumbar region whereas meningiomas are common in the thoracic region. Extradural tumors account for less than 25% of primary spine neoplasms and mainly includes meningiomas and metastatic lesions. When these involve the spinal canal there may be displacement of the cord with narrowing of the subarachnoid space both ipsilateral and contralateral to the lesion. The present study was aimed to study all the tumors involving the spinal cord.

## MATERIAL AND METHODS

The study included all the resected specimens and biopsies of spinal cord tumors received at our department over a period of three year. Relevant clinical data and imaging details were also reviewed. Cases which were arising only from spinal cord were included, cases secondarily extending in to spinal cord from vertebrae and cases with insufficient data were excluded from the study. All the specimens were fixed in 10 % buffered formalin, embedded in paraffin. Sections of 5µ thickness were cut, stained with H & E and examined under microscope. Immunohistochemistry was done wherever necessary.

All the data was divided in to different categories and analysed.

## RESULTS

In our study, total no of spinal tumors encountered were 45 with males constituting 22 cases (49%) and females constituting 23 cases (51%) with almost equal preponderance. Tumors presented in our study includes Schwannomas 15 cases (33%), Neurofibromas 10 cases (23%), Ependymoma 5 cases (11%), Meningiomas 5 cases (11%), Astrocytoma 4 cases (9%), Teratomas 2 cases (4%), and others include one case each of Paraganglioma, PNET, Oligodendroglioma, Metastatic deposits (9%) (Table 1)(Figure 1).

**Table-1: List of tumors presented in our study with sex wise distribution.**

| Tumor Type | Males | Females | Total cases |
|---|---|---|---|
| Schwannoma | 9 | 6 | 15 [33%] |
| Neurofibroma | 6 | 4 | 10 [23%] |
| Ependymoma | 2 | 3 | 5 [11%] |
| Meningioma | 1 | 4 | 5 [11%] |
| Astrocytoma | 1 | 3 | 4 [9%] |
| Teratoma | 0 | 2 | 2 [4%] |
| Others | 3 | 1 | 4 [9%] |
| Total | 22 | 23 | 45 [100%] |

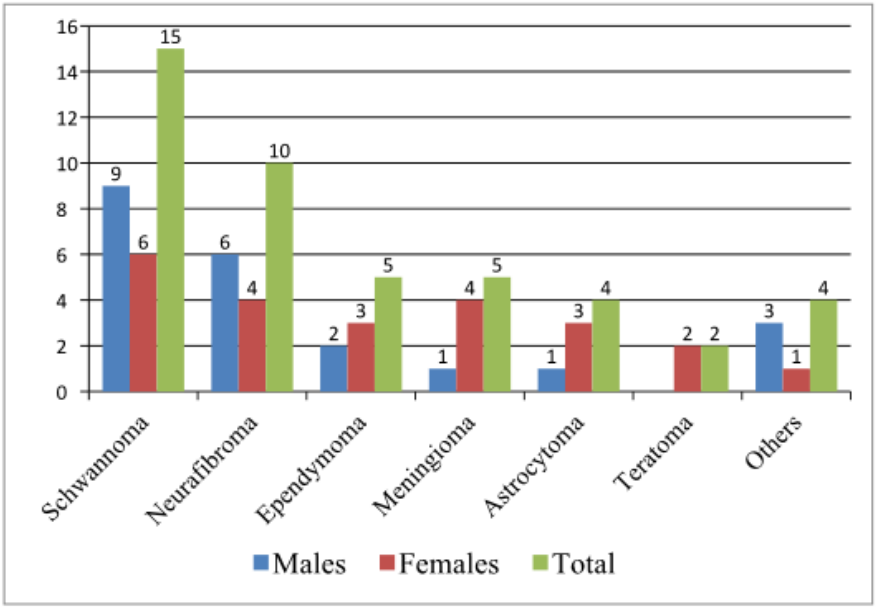


**Fig-1: Showing type and sex wise distribution of tumors.**

Most tumors are encountered in 20 – 30 years of age (Table 2).

**Table-2: Age wise distribution.**

| Age [Yrs] | No of cases |
|---|---|
| 0-10 | 5 |
| 11-20 | 5 |
| 21-30 | 14 |
| 31-40 | 9 |
| 41-50 | 8 |
| 51-60 | 1 |
| > 60 | 3 |

Based on location in relation to spinal cord they are divided into Intramedullary tumors constitute 8 cases (18%), Intradural extramedullary tumors constitute 25 cases (56%) and Extradural tumors constitute 12 cases (26%) (Table 3).

**Table-3: Location wise distribution of tumors.**

| Location | No of cases |
|---|---|
| Extradural | 12 [18%] |
| Intradural extramedullary | 25 [56%] |
| Intramedullary | 8 [26%] |

In relation to site, they constitute, Cervical region 7 cases (15%), Thoracic region 25 cases (56%), Lumbosacral region 13 cases (29%)（Table 4）.

**Table-4: Site wise distribution.**

| Site | No of cases |
|---|---|
| Cervical | 7 [15%] |
| Thoracic | 25 [56%] |
| Lumbo-sacral | 13 [29%] |

Most common symptom was Motor weakness followed by Pain, Sensory disturbance and Bladder dysfunction（Table 5）.

**Table-5: Clinical symptoms in Spinal tumors**

| Symptoms | No of Cases |
|---|---|
| Motor weakness | 20 [44%] |
| Pain | 13 [29%] |
| Sensory disturbances | 10 [22%] |
| Bladder dysfunction | 2 [5%] |

## DISCUSSION

Primary spinal cord tumors are one of the rarest categories of tumors, representing about 4–16 % of all tumors arising from the central nervous systems [1,2,]. Tumors of glial origin [e.g., astrocytomas, ependymomas] are usually intradural intramedullary in location, whereas nerve sheath tumors [e.g., neurofibromas and schwannomas] are typically intradural extramedullary lesions. Meningiomas can be either extradural or intradural extramedullary lesions. In our study total spinal tumors were 45 cases, with males constituting 22 cases (49%) and females constituting 23 cases (51%) with almost equal preponderance. According to the Engelhard *et al*. [3] the most common tumor types were meningioma (24.4%), ependymoma (23.7%), and schwannoma (21.2%). In our study most common tumor type include schwannomas (33%) which correlated with the study conducted by Hirano *et al*. [4], Garrido et [5] al and Ferreira *et al*. [6]. Pain, weakness, and sensory disturbances have been found to be the most frequent presenting symptoms and signs in adult and pediatric patients with intraspinal tumors [3,7]. In our study the most common symptom was motor weakness followed by pain.

Intramedullary tumors mainly include ependymomas followed by astrocytomas [2,8]. Astrocytomas are the most common intramedullary tumor in children. The mean age at presentation is 29 years. The most common site of involvement is the thoracic cord followed by the cervical cord. In our study they are second common intramedullary tumors which correlated with study done by Ferreira et al. We reported 4 cases of astrocytoma, 3 cases were found in the thoracic region and one case in cervical region, they are common in children with mean age of 22 yrs. They are mostly intramedullary but we reported one case of extramedullary intradural lesion. The tumor is hypo to isointense on T1W images, hyperintense on T2W, with variable contrast enhancement（Figure 2）.

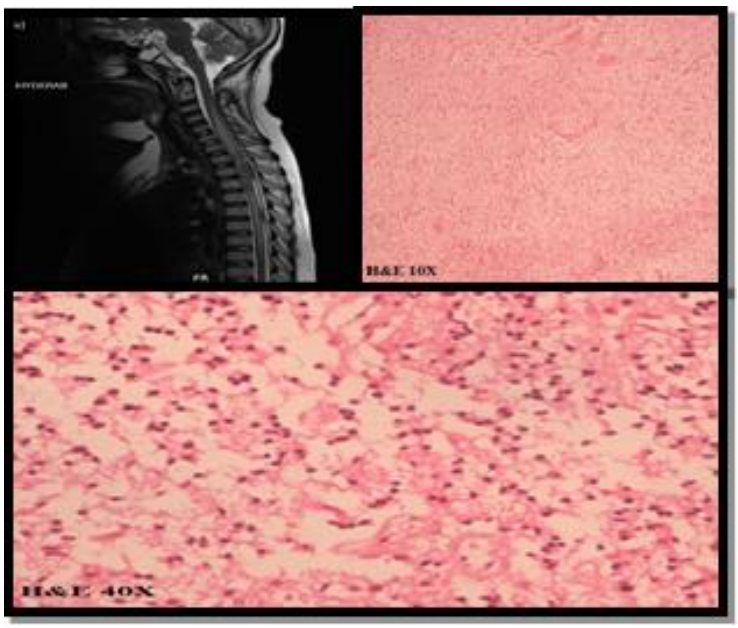


**Fig-2: Showing MRI and H&E section (10X & 40X) of Astrocytoma.**

Ependymomas are the most common glial tumor in adults, whereas astrocytomas are the most common intramedullary tumor in children [2,3,9]. Ependymomas tend to manifest in young adulthood,

with a mean age at presentation of 38.8 years and are more common in male patients. Cord ependymomas occur most commonly in the cervical region [2]. In our study ependymomas were common in adults with mean age of 32 yrs but were common in females in contrast to the study done by Hirano *et al*. [4] and common location in our study was thoracic region. Primary intradural extramedullary ependymomas of the spinal cord are rare. In our study 4 cases were intramedullary and one case was intradural extramedullary Similar lesion was also reported by Graca *et al*. [8] in thoracic region (Figure 3).

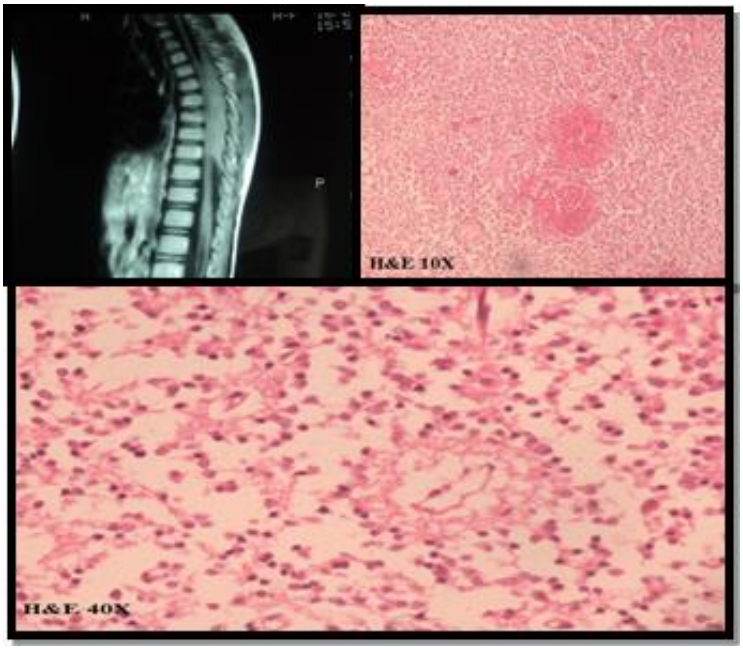


**Fig-3: MRI image and H&E section (10X & 40X) of Ependymoma with rosettes.**

The two most common ependymoma subtypes are cellular and myxopapillary ependymomas. Cellular ependymomas can arise anywhere but usually occur in the cervical cord, whereas myxopapillary ependymomas occur almost exclusively in the conus medullaris and filum terminale. Ependymomas by MRI appear as a focal enlargement of the cord and hyperintense on T2W and hypo or isointense to normal spinal cord on T1Wimages with heterogeneous contrast enhancement. We reported one case of myxopapillary ependymoma in female in filum turminale but similar case reported, occurring in both spinal cord and cerebral region.

Nerve sheath tumors constitute about 25 percent of tumors arising in the intradural extramedullary space. Benign spinal nerve sheath tumors [neurofibromas and schwannomas] often occur on dorsal nerve roots sporadically or in neurofibromatosis types 1 and 2 [10,11]. Neurofibromas rarely involve the spinal cord. According to some studied, spinal nerve sheath tumors, 83% neurofibromas were intradural, 10% extradural and 7% both intradural and extradural. In our study we reported 10 cases of neurofibromas, of these 60% were intradural and 40% were extradural. Cervical region constituted 10%, Thoracic region constituted 50% and lumbosacral region constituted 40%. Males were 60% and females were 40% with mean age of presentation of 36 yrs (Figure 4). Neurofibromata involving the roots of the spinal cord are usually singular, but occasionally may be multiple and may be part of NF1 [12]. Spontaneous pain and dysesthesias are the most common presenting symptoms. Patients with neurofibromatosis type I may have multiple spinal cord neurofibromas that often increase in number with age. On imaging, neurofibromas appear as rounded or fusiform tumors that are isointense on T1W images and hyperintense on T2W images [13]. In our study all cases were hypointense on T1 and hyperintense on T2.

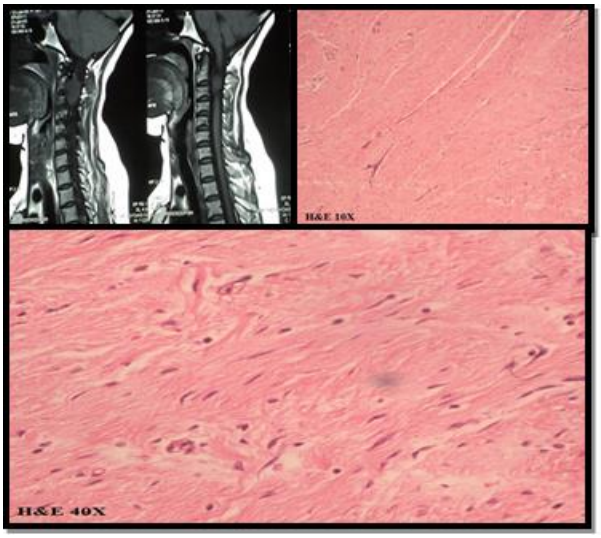


**Fig-4: MRI image and H&E section (10X & 40X) of neurofibroma.**

Spinal schwannomas account for about 25% of intradural spinal cord tumors in adults [14]. Most are solitary schwannomas, which can occur throughout the spinal canal. We reported 15 cases of schwannomas, of these 11 cases showed typical Antoni A and Antoni B areas whereas 4 cases were cellular schwannomas. There was no difference among males and females in many studies but our study reported predominance in males [60%] compare to females [40%] which was in concordance with the study of Hirano *et al*. [4] and Jeon *et al*. [14]. The mean age of presentation was 36 yrs. In our study 80% cases were found in thoracic and lumbar region with only 20% in cervical region. In the literature, 70 to 80% of spinal schwannomas are reported to be intradural in location, and those extending through the dural aperture as a dumbbell mass with both intradural and extradural components account for another 15%. In our study, intradural tumors were 67% and extradural tumors were 33%. Intramedullary schwannomas are extremely rare. Pain was the common complaint in our study [14]. On MRI, schwannomas appear as solid tumors in the dorsal sensory root region, with displacement of the spinal cord, conus medullaris, or filum terminale. They are isointense on T1W MRI and hyperintense on T2W images. Schwannomas are difficult to differentiate on MRI from other nerve sheath tumors, in our study 4 cases were diagnosed as neurofibromas on MRI but they turned to be schwannomas on histopathology. In our study all cases were hyperintense on T2W but 10 cases were isointense and 5 cases were hypointense on T1W imaging. When the diagnosis of schwannoma is early and operation is performed before the spinal cord compression, good results are achieved [15]. Hence there is a need for early and accurate diagnosis of these tumors（Figure 5）.

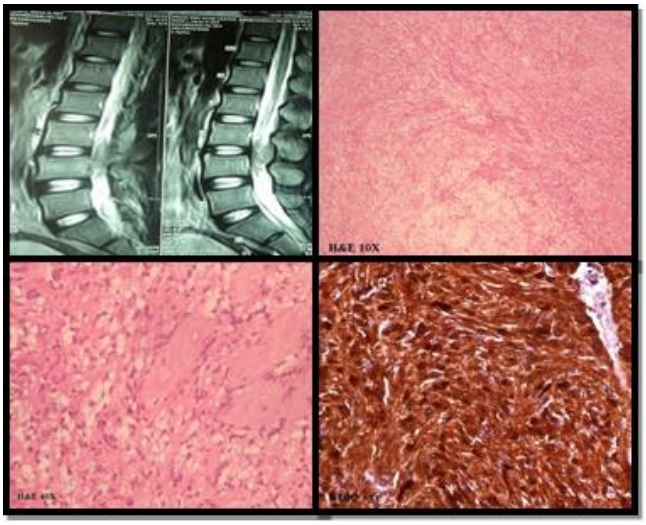


**Fig-5: MRI image and H&E section (10X & 40X) of schwannoma with S100 positivity.**

Meningiomas are benign tumours arising from arachnoid cells and mostly located in the intracranial compartment. Spinal meningiomas are rare and account about 1.2% of all meningiomas and 25% of all spinal cord tumours [16,17]. We reported 5 cases of spinal meningiomas of these 4 cases (80%) were in females and one case (20%) was in male with male to female ratio of 1:4 which correlated with other studies [Figure 6]. The most common location of spinal meningioma is the thoracic spine [4,10,13,16,17]. In our study also common site was thoracic with 80% cases which correlated with study done by Sandalcioglu et al, Setzer et al**.** In our study 80% cases were intradural extramedullary which correlated with other studies. We reported one case at extradural location which is a rare site for meningioma [4,13,16]. Mean age of presentation in our study was 31 yrs but most of the studies showed peak incidence at 5th to 7th decade. Most common symptoms were sensory and motor deficits.

On MRI, meningiomas appear as solid, well circumscribed lesions with an attachment to the dura. The tumor is iso- to hypointense on T1W and slightly hyperintense on T2W images. In our study all cases were hypointense on T1W but 3 cases were isointense and 2 cases were hyperintense on T2W, 2 cases were meningothelial （Grade Ⅰ), 2 case of transitional （Grade Ⅰ）and 1 case showed atypical features（Grade Ⅱ). Asymptomatic patients with spinal cord meningioma can be followed clinically with serial imaging studies. If treatment is indicated, surgery is the primary modality and can be curative with complete resection.

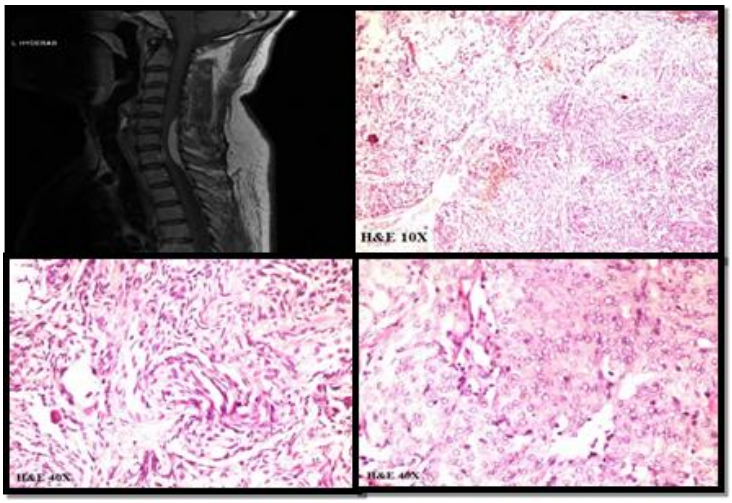


**Fig-6: MRI image and H&E section (10X & 40X) of Meningioma.**

Teratomas in the spine are extremely rare. In cord they constitute less than 0.5% of all intraspinal tumours [18,19]. Teratomas may be intra or extramedullary and usually fill the spinal canal. We reported two cases of spinal teratomas. Some studies showed predominance in males [18] whereas some showed equal predominance, but our study showed female dominance. Intraspinal teratomas can be located at the thoracic [18], conus medullaris [19], cervical or lumbar regions. In our study one case was found in cervical region and another case at lumbosacral region. Intramedullary teratomas were even rare, we reported one case at intramedullary location. The diagnosis of teratoma depends on the histopathological identification of the tissues representing the three germinal layers [ectoderm, mesoderm and endoderm]. However, the presence of just two layers does not rule out the diagnosis. Several theories of tumour pathogenesis have been raised in the literature [18,19]. MRI features cannot determine with certainty the differential diagnosis between teratoma and other intramedullary lesions, hence definitive diagnosis depends on the histological examination. Total excision is the primary treatment modality. Metastatic lesions to spinal cord are mostly extradural in location [20]. In our study we had one case of metastatic mucinous adenocarcinoma in thoracic region, extradural in location which was common site but other studies reported metastatic deposit from thyroid and bladder carcinoma intramedullary location which is rare. Primary spinal cord oligodendrogliomas constitutes 2% of the spinal cord tumors and 1.5% of the central nervous system oligodendrogliomas. Spinal cord is the anatomic location with the lowest predilection of oligodendrogliomas [21,22]. In our study we presented a case of oligidendroglioma in 26 yr old female at intramedullary location in thoraco lumbar region which was common site but occurrence at other locations like cervical, filum terminale were also reported. Our study had complaint of pain and younger age of presentation

which correlated with the literature. Our case showed typical grade II oligodendroglioma but anaplastic variants are also reported. At operation most oligodendrogliomas of the cord appear as infiltrating gelatinous tumors, though a minority have a firm consistency and apparently clear cut contours, which seem to be associated with a better prognosis.

## CONCLUSION

Spinal cord is part of central nervous system and all the tumors which occur in CNS can occur in spinal cord. Hence all the tumors should be kept in mind before giving final diagnosis. The appropriate diagnosis depends on clinical history, radiological features and pathological examination. Schwannomas were most common tumor type encountered in our study, followed by neurofibromas. Ependymomas and astrocytomas were common intramedullary tumors whereas schwannomas, neurofibromas and meningiomas were common intradural extramedullary tumors. As any tumour can occur in spinal cord broad differentials are kept in mind before giving final diagnosis, as correct diagnosis helps in the treatment and prognosis.

## REFERENCES


1. Kufe DW, Pollock RE, Weichselbaum RR, Bast RC, Gansler TS, Holland JF. Frei El, editors. Cancer Medicine. Canada, Hamilton: BC Decker Inc. 2003.
2. Koeller KK, Rosenblum RS, Morrison AL. Neoplasms of the spinal cord and filum terminale: radiologic-pathologic correlation. Radiographics. 2000 Nov;20(6):1721-49.
3. Engelhard HH, Villano JL, Porter KR, Stewart AK, Barua M, Barker FG, Newton HB. Clinical presentation, histology, and treatment in 430 patients with primary tumors of the spinal cord, spinal meninges, or cauda equina. Journal of Neurosurgery: Spine. 2010 Jul;13(1):67-77.
4. Hirano K, Imagama S, Sato K, Kato F, Yukawa Y, Yoshihara H, Kamiya M, Deguchi M, Kanemura T, Matsubara Y, Inoh H. Primary spinal cord tumors: review of 678 surgically treated patients in Japan. A multicenter study. European Spine Journal. 2012 Oct 1;21(10):2019-26.
5. Garrido PI, Laher-Mooncey S, Murphree NL, Jonker N, Levy LF, Makarawo S. Neoplasms involving the spinal cord in Zimbabweans: an analysis of 262 cases.
6. Ferreira NP, Chaves DL, Moraes AC, de Oliveira LM. Spinal tumors: apropos of 100 cases. Arquivos de neuro-psiquiatria. 1981 Mar;39(1):25-31.
7. Crawford JR, Zaninovic A, Santi M, Rushing EJ, Olsen CH, Keating RF, Vezina G, Kadom N, Packer RJ. Primary spinal cord tumors of childhood: effects of clinical presentation, radiographic features, and pathology on survival. Journal of neuro-oncology. 2009 Nov 1;95(2):259.
8. Graca J, Gültaşli N, D'Haene N, Brotchi J, Salmon I, Balériaux D. Cystic extramedullary ependymoma. American journal of neuroradiology. 2006 Apr 1;27(4):818-21.
9. Wilson RE, Oleszek JL, Clayton GH. Pediatric spinal cord tumors and masses. The journal of spinal cord medicine. 2007 Jan 1;30(sup1):S15-20.
10. Ali M, Khan Z, Sharafat S, Khan KM, Rehman S. Experience with 349 cases of intradural spinal tumors at Lady Reading Hospital Peshawar. PJS-Pakistan Journal of Surgery. 2010:221-5.
11. Murovic JA, Cho SC, Park J. Surgical strategies for managing foraminal nerve sheath tumors: the emerging role of CyberKnife ablation. European Spine Journal. 2010 Feb 1;19(2):242-56.
12. Rasmussen TB, Kernohan JW, Adson AW. Pathologic classification, with surgical consideration, of intraspinal tumors. Annals of surgery. 1940 Apr;111(4):513.
13. Chamberlain MC, Tredway TL. Adult primary intradural spinal cord tumors: a review. Current neurology and neuroscience reports. 2011 Jun 1;11(3):320-8.
14. Jeon JH, Hwang HS, Jeong JH, Park SH, Moon JG, Kim CH. Spinal schwannoma; analysis of 40 cases. Journal of Korean Neurosurgical Society. 2008 Mar 1;43(3):135-8.
15. Subaciūte J. Early diagnosis of spinal cord schwannoma: the significance of the pain syndrome. Medicina (Kaunas, Lithuania). 2002;38(11):1086-8.
16. Sandalcioglu IE, Hunold A, Müller O, Bassiouni H, Stolke D, Asgari S. Spinal meningiomas: critical review of 131 surgically treated patients. European Spine Journal. 2008 Aug 1;17(8):1035-41.
17. Setzer M, Vatter H, Marquardt G, Seifert V, Vrionis FD. Management of spinal meningiomas: surgical results and a review of the literature.
18. Borlot F, Soares MS, Espíndola AÁ, Reed UC, Matushita H, Teixeira MJ. Intramedullary spinal teratoma: a rare condition with a good outcome. Arquivos de neuro-psiquiatria. 2009 Sep;67(3A):733-5.
19. Hamabuchi MA, Hasegawa RY, Murase TE. Teratoma of the spinal cord. A case report with CT scans. Bone & Joint Journal. 1989 May 1;71(3):390-2.
20. Honma y, kawakita k, nagao s. Intramedullary Spinal Cord and Brain Metastases from Thyroid Carcinoma Detected 11 Years after Initial

Diagnosis—Case Report—. Neurologia medico-chirurgica. 1996;36(8):593-7.

21. Yucel E, Ekinci Ö, Gürkanlar D, Koçak H, Aciduman A. Primary spinal cord oligodendroglioma. Case illustration. Neurocirugia. 2006 Dec 31;17(6):542-3.
22. Nathoo AR, Halliday NP. Spinal cord oligodendroglioma. Postgrad Med J. 1967 Dec;43(506):789-91.