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Spinal nerve sheath tumors: Analysis of 20 cases with review of literature

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Abstract

Spinal cord tumors are rare among the tumors of central nervous system (CNS). They constitute 4 - 16 % of all the CNS tumors and of these spinal nerve sheath tumors constitute significant group, situated mostly in the intradural location. The study was undertaken to report the spinal nerve sheath tumors received at the tertiary care centre over a period of 3 years with review of literature. Schwannomas were common nerve sheath tumors with 60% cases followed by Neurofibromas with 40% cases, mostly located intradurally. Both the lesions showed male predominance. These tumors have to be distinguished from one another as the treatment and prognosis varies.

Keywords: Intradural; Nerve sheath; Neurofibroma; Schwannoma; Spinal cord

Introduction

Spinal cord tumors comprises 4 - 16 % of all the Central Nervous System (CNS) tumors [1-4]. Spinal nerve sheath tumors constitute 25 % of all spinal intradural extramedullary tumors. Most of these tumors include schwannomas and neurofibromas. These tumors are usually located in an intradural-extramedullary location and intradural growth along the nerve into the spinal canal also occurs, but rare cases of intramedullary location are also reported [5].

Neurofibromas are well-demarcated intraneural or diffusely infiltrative extraneural tumor consisting of a mixture of cell types, including Schwann cells, perineurial-like cells, and fibroblasts. Neurofibroma presents most commonly as a cutaneous nodule occasionally involve spinal roots, but are almost unknown on cranial nerves. They may be solitary or may be associated with neurofibromatosis [6]. They are benign but malignant transformation is not uncommon hence regular follow up is mandatory.

Schwannomas are benign nerve sheath tumor that are typically encapsulated and composed entirely of well-differentiated schwann cells, also called as Neurilemoma or Neurinoma. They represent 25% of intradural spinal cord tumors in adults [1]. They arise from the lining of the nerve cells of the spine. Nerves of the spine are lined by the protective sheath called myelin which transmits nerve impulse throughout the body. When tumors grows, it compresses the spinal nerves and causes symptoms such as tingling sensation, numbness, weakness and pain in the limbs.

Material and Methods

Present study was done over a period of 3 yrs from June 2010 to may 2013 at our institute. Specimens received to the pathology department were subjected to the routine processing and paraffin embedded sections were stained with H & E and examined. Immunohistochemistry (IHC) was done wherever necessary. MRI findings and clinical data were also taken into consideration. Post operative follow up was available.

Results

A total 20 cases of spinal nerve sheath tumors were studied, of these 12 cases (60%) were Schwannomas and 8 cases (40%) were Neurofibromas. males were 13 cases (65%) and females were 7 cases (35%) with male : female ratio of 1.8: 1. They are common in 3rd to 4th decade with mean age of presentation was 36 yrs. Most common presenting symptom was Pain (55%) followed by Sensory disturbances (25%). Based on location, 3 cases were in Cervical (15%), 8 cases in Thoracic (40%) and 9 cases were in Lumbosacral region (45%). Of the four cases diagnosed as neurofibromas on MRI were turned to be schwannomas on histopathology.

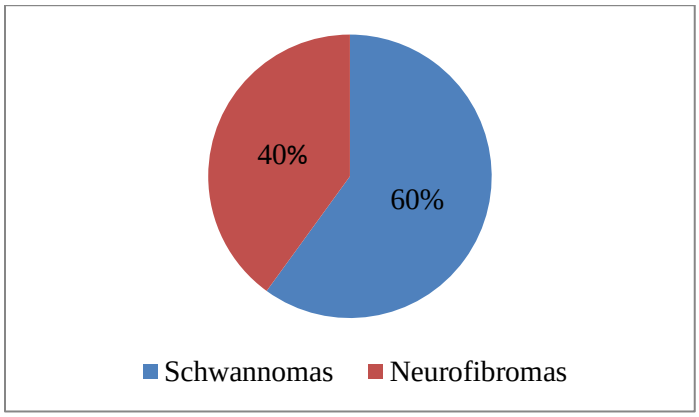

Figure 1: Total distribution of cases

Table 1: Sex distribution of tumors

Tumors	Males	Females	Total
Schwannomas	8(40%)	4(20%)	12(60%)
Neurofibromas	5(25%)	3(15%)	8(40%)
Total	13(65%)	7(35%)	20(100%)

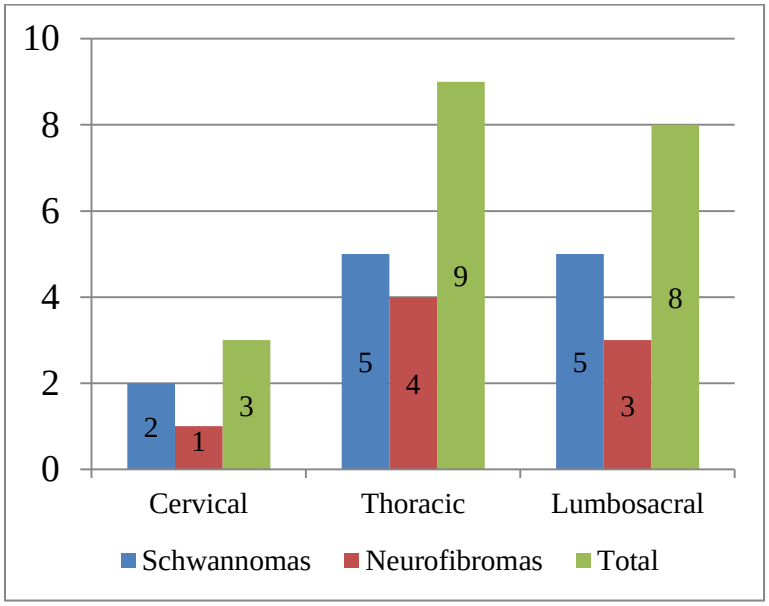

Figure 2: Site distribution of tumors

Table 2: Site distribution of Tumors

Location	Schwannomas		Neurofibromas	
	Intradural Extramedullary	Extra dural	Intradural Extramedullary	Extra dural
Cervical	1	1	1	0
Thoracic	4	1	2	2
Lumbosacral	3	2	2	1

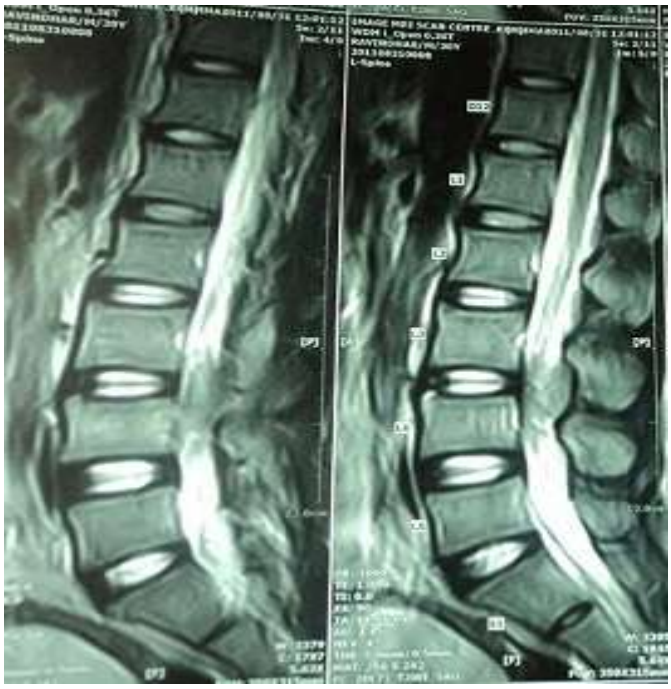

Figure 3: MRI showing hyperintense lesion in lumbar region (Schwannoma)

Table 3: Clinical symptoms of lesions

Symptom	Cases	Percentage
Pain	11	55 %
Sensory Disturbances	5	25 %
Weakness	3	15 %
Autonomous dysfunction	1	5 %

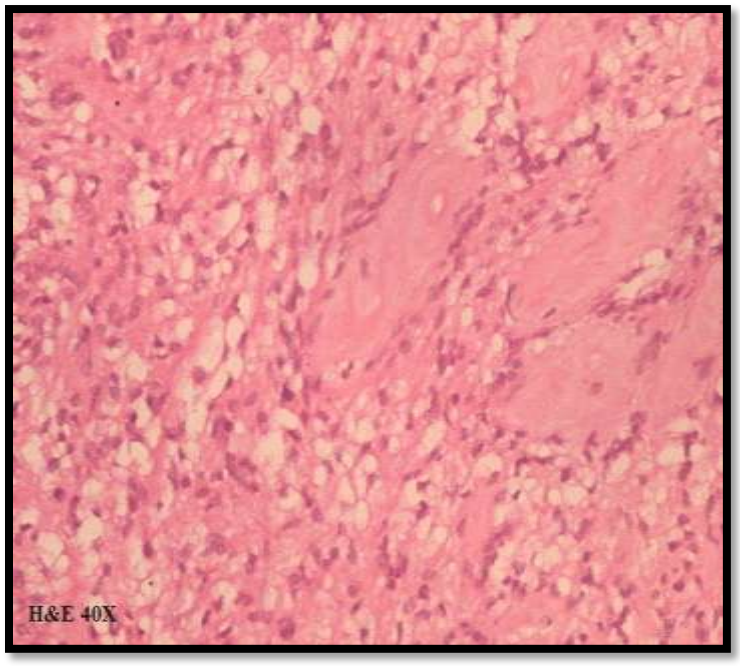

Figure 4: H&E Schwannoma with hypercellular and hypocellular areas with verocay bodies

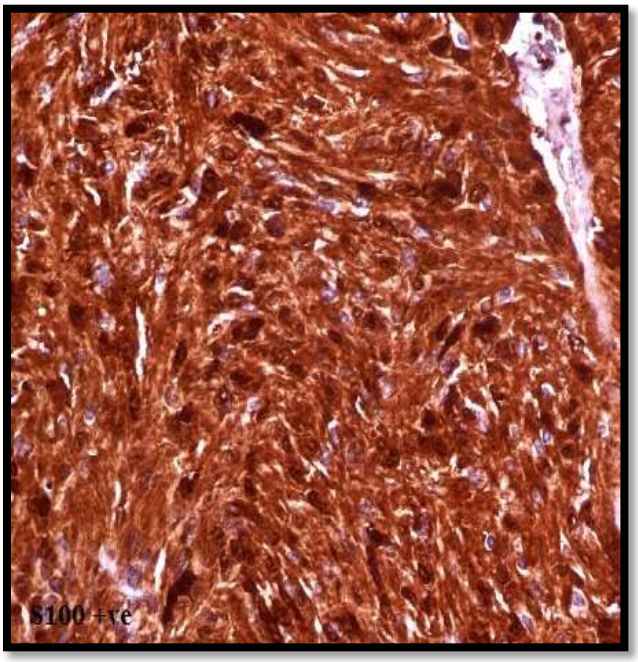

Figure 5: Schwannoma showing positivity for S100 marker

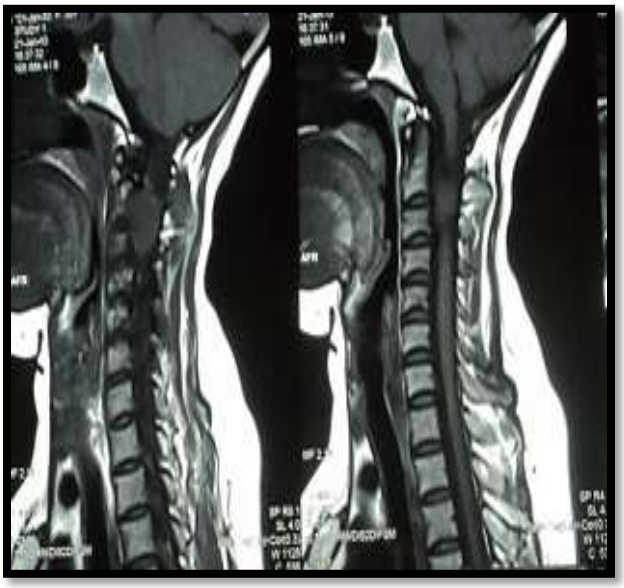

Figure 6: MRI showing isointense lesion in cervical region (Neurofibroma)

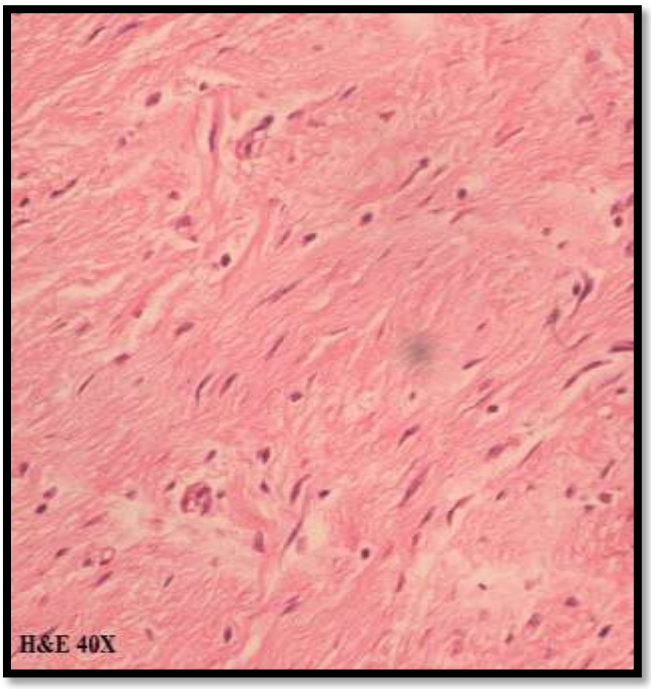

Figure 7: H&E 40X Neurofibroma with wavy nuclei and intersecting collagen bundles

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 [1-3]. 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 [1].

Nerve sheath tumors constitute about 25 percent of tumors arising in the intradural extramedullary space[4]. Benign spinal nerve sheath tumors [neurofibromas and schwannomas] often occur on dorsal nerve roots sporadically or in neurofibromatosis types 1 and 2 [5-8]. Neurofibromas rarely involve the spinal cord. According to *Levy et al* who studied 66 patients with spinal nerve sheath tumors, 83% neurofibromas were intradural, 10% extradural and 7% both intradural and extradural. In our study we reported 8 cases of neurofibromas, of these 63% were intradural and 37% were extradural. Cervical region constituted 12%, Thoracic region constituted 50% and Lumbosacral region constituted 38%. Males were

63% and females were 37% with mean age of presentation of 36 yrs.

Neurofibromata involving the roots of the spinal cord are usually solitary, but occasionally may be multiple and may be part of NF1 which often increases with age [9,10]. Spontaneous pain and dysesthesias are the most common presenting symptoms. On imaging, neurofibromas appear as rounded or fusiform tumors that are isointense on T1W images and hyperintense on T2W images [10]. In our study all cases were hypointense on T1 and hyperintense on T2. All cases were solitary but one case seen as a part of neurofibromatosis type 1. On follow up one case showed recurrence.

Spinal schwannomas account for about 25% of intradural spinal cord tumors in adults [11-13]. Most are solitary schwannomas, which can occur throughout the spinal canal. We reported 12 cases of schwannomas, of these 8 cases showed typical Antoni A and Antoni B areas whereas 4 cases were cellular schwannomas with low proliferative index. There was no difference among males and females in many studies but our study reported predominance in males [67%] compare to females [33%] which was in concordance with the study of *Hirano et al* and *Jeon et al* The mean age of presentation was 36 yrs.

In our study 83% cases were found in thoracic and lumbar region with only 17% 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% [12]. In our study, intradural tumors were 67% and extradural tumors were 33%. Intramedullary schwannomas are extremely rare [12-14]. *Zeki et al* reported one case of intramedullary schwannoma, our study has no intramedullary schwannoma. Pain was the common complaint in our study [15].

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 [10]. In our study all cases were hyperintense on T2W but 8 cases were isointense and 4 cases were hypointense on T1W imaging. 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. 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.

Conclusion

Spinal nerve sheath tumors are common tumors involving the spinal cord. Schwannomas are common tumors followed by Neurofibromas. They have male predominance and present mostly with pain and sensory disturbances. MRI with histopathological confirmation is the diagnostic modality. Accurate diagnosis is essential as the treatment and prognosis often varies with the lesion. Total surgical excision is necessary to prevent recurrence and for good prognosis.

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