Primary Spinal Schwannoma: A Single Center Study of 37 Consecutively Operated Cases

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Abstract:

Background: Primary spinal Schwannoma (PSS) remains uncommon nerve sheath tumor of the spine with male preponderance and presenting in fourth to fifth decades. They arise from the Schwann cells in sensory root as PSS or part of Neurofibromatosis 2.

Methods: This is a retrospective study of histopathologically confirmed cases of PSS treated between 2010 to 2021. They were operated and age, sex, clinic-radiological findings, surgical technique used and outcome studied. Laminectomy or laminoplasty or combined anterior-posterior approach was used with either an intracapsular or extracapsular tumor removal. Follow-up was at 15 days, 2 months, 6 months and yearly intervals.

Results: There were 37 cases operated with age ranging from 16 to 81 years and majority in the 21–40-year group (40%) followed by 41 to 60 years (32%). There were 22 males and 15 females. Localized pain in the back or cervical region was the commonest finding (75%) followed by myelopathy in 38%, radiculopathy in 35% and bladder disturbances in 13%. They were most common in the lumbar region (35%) followed by thoracic (30%). With relation to the level and sex, the cervical level showed equal sex distribution while the rest had male preponderance

Conclusions: PSS although can frequently present to the spine surgeon and the diagnosis is clinched with help of an MRI. Surgery remains the main modality of treatment either by laminectomy, laminoplasty or minimal invasive spine surgery. The nerve root of origin must be preserved in all that can be aided with help of intraoperative neurophysiological monitoring.

Key words: Laminectomy, Laminoplasty, Myelopathy, Radiculopathy, Schwannoma, Spine, Tumor

Introduction

Spinal tumors are rare with meningiomas more frequent than schwannoma in intradural location. Primary Spinal Schwannoma (PSS), defined as those not associated with other syndromes, remains the most common nerve sheath tumor of the spine with studies showing male preponderance.[1,2] They most commonly present in the fourth and fifth decades of life and in the American population commoner in whites than African or Asians.[3] These tumors arise from Schwann cells in sensory root as PSS or part of Neurofibromatosis 2/ Schwannomatosis. Symptoms are due to either radiculopathy or myelopathy. They are slow growing and diagnosis is done with help of clinical findings supplemented with radiological investigations. Surgery remains the primary modality of treatment and radiotherapy remains the only option for inoperable tumors.[4,5] This article discusses a single center retrospective study of cases operated and outlines the clinic-radiological, operative findings/technique, post-surgical follow-up and discussion regarding recent trends in its management.

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Methods

This is a retrospective study of histopathologically confirmed cases of PSS treated in this center from 2010 to 2021. A total number of 37 cases were operated and age, sex, clinic-radiological findings, surgical technique used with outcome studied. Follow-up was between one to ten years. All cases associated with neurofibromatosis 2, tumors histopathologically negative for Schwannoma, meningioma and other lesions were excluded from the study.

Diagnosis: All cases with features of either radiculopathy, myelopathy, backache, spinal deformity, bowel or bladder disturbances and other localizing symptoms were subjected to contrast enhanced Magnetic resonance Imaging (MRI) of the spine. Once findings were suggestive of intradural tumors then localization was done with preoperative radiological markers on the skin and plain skiagram taken to identify the spinal level. Preoperative nerve conduction studies were performed in some of the cases.

Surgical technique: all cases underwent preoperative hematological and biochemical laboratory tests to rule out medical illnesses. Under general anesthesia the spinal level was identified (except for the cervical and lower lumbar levels) according to the skin marking and either laminectomy (for 2 spinal level) or laminoplasty for more than 3 level was performed. High speed drill was used to cut the lamina for laminoplasty and titanium plates used to fix them at end. After opening the dura and identifying the tumor it was gently teased out, separated from either spinal cord or cauda equina and removed under operating microscope. Either an intracapsular (Small tumor) or extracapsular (larger tumor) approach was used depending on the size of the tumor. The nerve root was deliberately preserved and in those where it was severed the ends were sutured with 10-0 polypropylene sutures. Dura was closed water tight with or without use of sealant and wound closed in multiple anatomical layers. They were all extubated and made ambulatory on the second day. Sutures were removed on the tenth day and discharged home. Follow-up with regards to clinical findings was done at 15 days, 2 months, 6 months and yearly intervals. MRI was done at 6 months.

Results

There were 37 cases that were operated and included as per the inclusion criteria in the study period. The age ranged from 16 to 81 years with the majority in the 21–40-year group (40%) followed by the 41 to 60 years group (32%) (Table 1). There were 22 males and 15 females. Regarding the relation of age with sex, females were most common in the 41 to 60-year group while the males were common in all the other groups (Table 1). Regarding the clinical symptoms, localized pain in the back or cervical region was the most common finding (75%) followed by myelopathy in 38%. Radiculopathy in 35%, bladder disturbances in 13%, dysphagia/odynophagia was present in one case and clinically palpable lump in 2 cases were the other common findings. With regards to the vertebral level of the tumors, they were most common in the lumbar region (35%) followed by thoracic in 30% (Figure 1-3). With relation to the level and sex, the cervical level showed equal sex distribution while the rest had predominantly male preponderance (Figure 1).

Regarding the method of surgery, laminectomy was done in 31 cases and laminoplasty in 6 cases. Anterior-posterior approach was done in 4 cases. There were two cases each of lumbar and cervical extradural extra spinal schwannoma extending into the anterior compartment. They were removed by either anterior extraperitoneal transabdominal approach or anterior cervical paratracheal approach. Two cases of dumbbell tumors in the upper cervical were removed by additional far lateral approach. Complete excision was done in 34 cases (92%) of cases. One cervical anterior dumbbell tumor and two thoraco-lumbo-sacral tumors was excised incompletely and sent for radiotherapy. Postoperatively there was good motor improvement in 33 cases and improvement of bowel and bladder disturbances in 34 cases. There was no case with infection but there were two with localized cerebrospinal fluid collection which was managed with repeated aspiration, use of acetazolamide for 7 days and lumbar corset with prone positioning for one week. Three cases had residual tumor due to subtotal resection because of the excessive adhesions to nerve roots. The follow up period ranged from 1-7 years and was possible in only75% of cases.

Symptoms A	Number (n=37)	%
Back or cervical pain	28	75
Myelopathy	14	38
Radiculopathy	13	35
Bladder distrubances	8	13
Dysphagia/odynophagia	1	3
Localized lump	2	5

Age B	Number (n=37)	%
<20	1	3
21-40	15	40
41-60	12	32
61-80	8	21
>80	1	3

Table 1: Table with (A) showing the clinical symptoms at diagnosis and (B) showing the age group distribution with number.

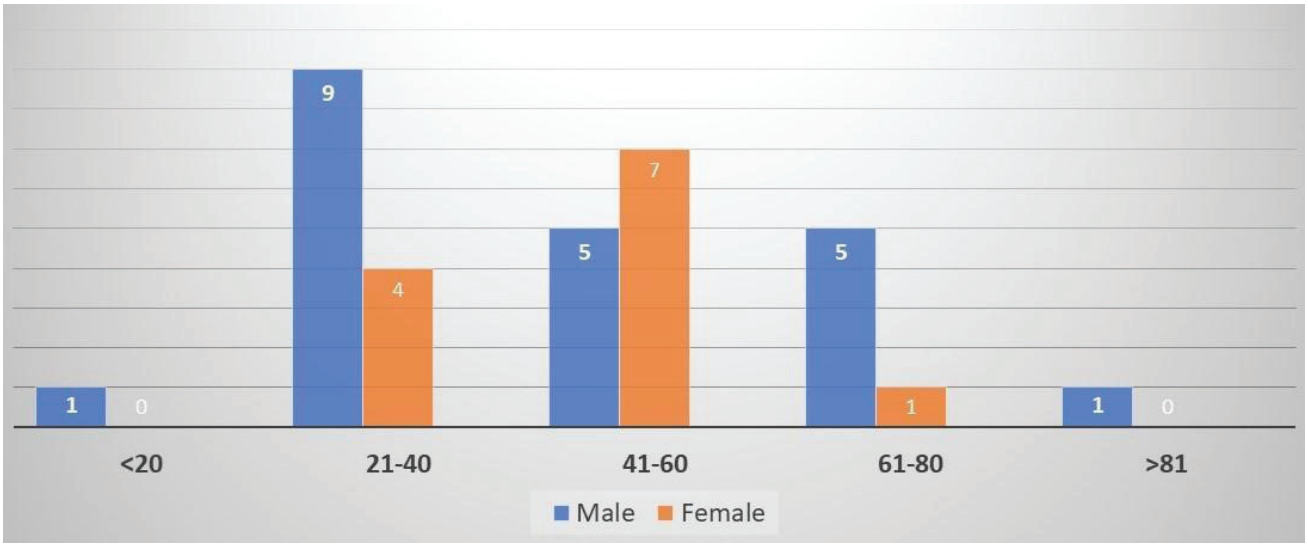

Table 2: Figure showing the relation of sex with age group of presentation.

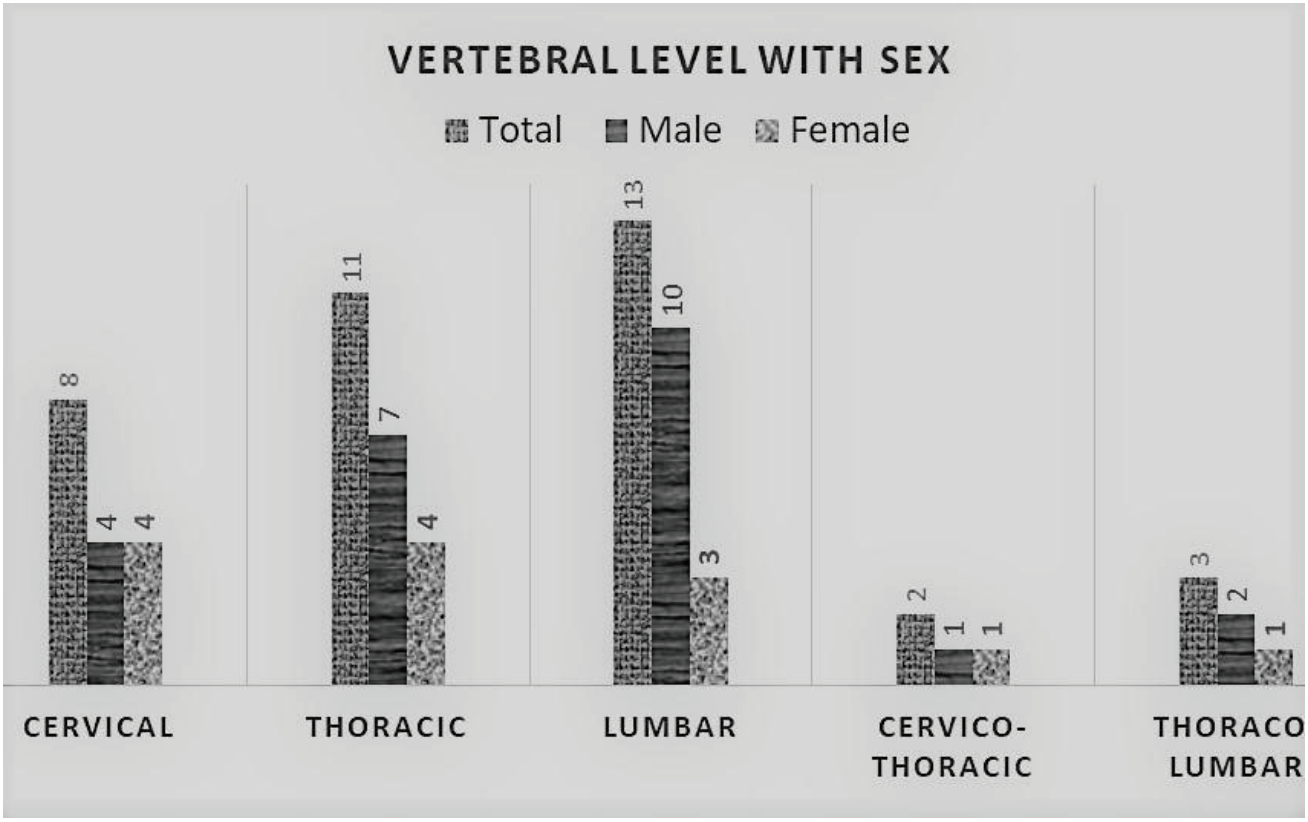

Figure 1: Figure showing the different vertebral levels of the tumors. It also shows the subgroup in relation to the level with sex of the patient.

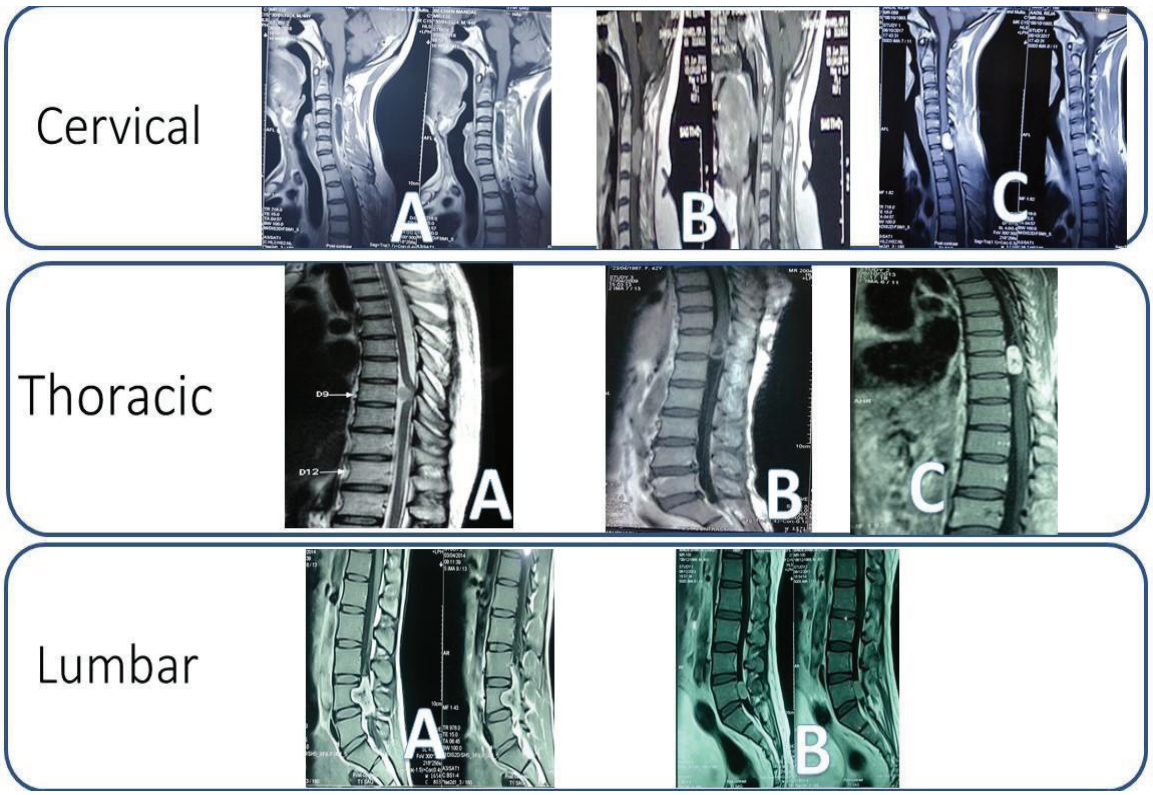

Figure 2: Figure showing examples of different levels of schwannoma. Cervical C2-7 level (A), C2-3 (B) and C6-7 (C). Thoracic D 9(A), D12 (B) and D7 (C). Lumbar 4-5 (A) and L5 (B).

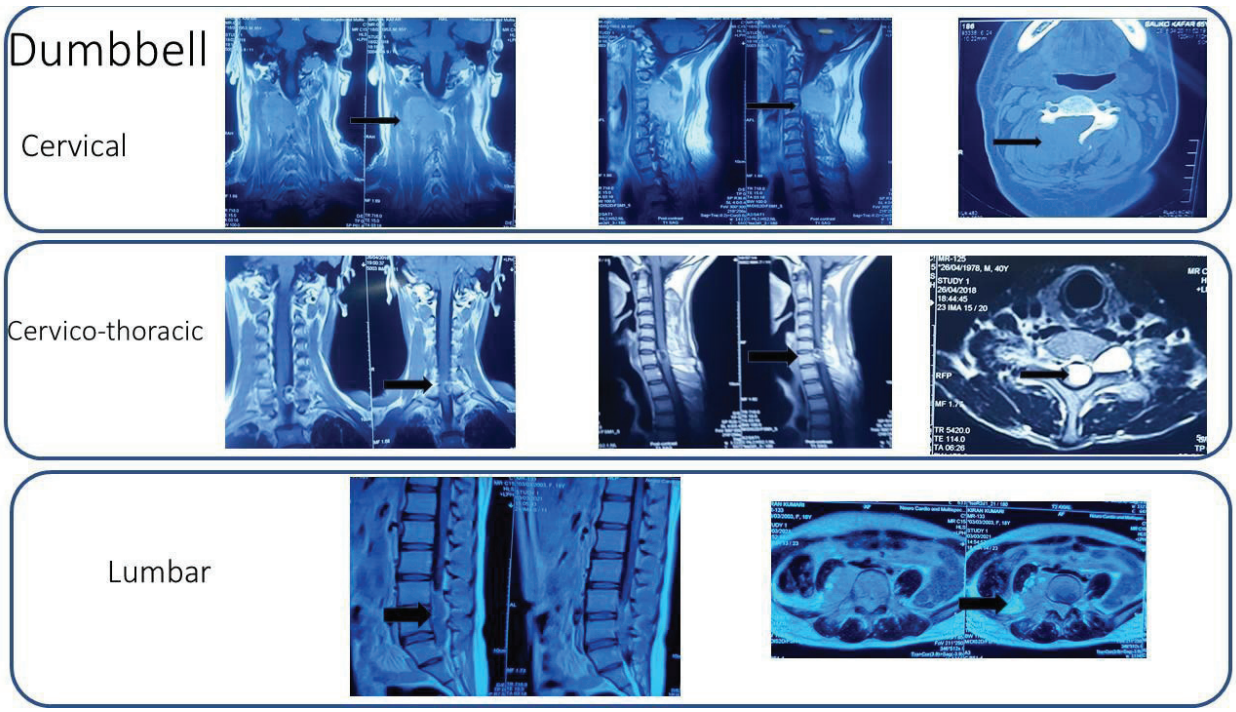

Figure 3: Figure showing different level of dumbbell tumors in the cervical, thoracic and lumbar regions, shown by black arrows.

Discussion

PSS arise from the Schwann cells of a sensory nerve root and are the most common nerve sheath tumor of spine presenting in the fourth to sixth decades of life. They are WHO grade I tumors that are usually sporadic but can also be part of neurofibromatosis 2 syndrome.[4,5]In this series they were most common in the 21 - 40 age group which is a decade lower than that found in other studies. In a comparison of age with sex, males were leading in all subgroups except for the fourth to sixth decade which was dominated by females. The males were predominantly in this series which is as per many other studies. Most studies have found either an equivocal sex distribution or slight male preponderance.[6,7] Pathologically they can be Antoni A (solid hypercellular/fascicular) or B (myxoid hypocellular/vacuolated) predominant type of tumors.[8] Various classifications are available in literature based on different characteristics but none of them encompass all the features of spinal schwannomas.[9-12]

***Clinical symptoms*:** [12-16]Common clinical symptoms are pain, followed by radiculopathy and in later stages myelopathy. Paraparesis or paraplegia and bowel or bladder incontinence is noted only in the later stages. Pure motor symptoms are very rare. The symptoms are either due to local compression, irritation or disruption of the conduction pathway. They are most common in the lumbosacral region and although definitive cause is unknown the long path of cauda equina roots seems to predispose them for PSS. Cystic variants can cause earlier symptoms than their solid counterpart. In our series the most common presentation was pain which suggest the possible sensory origin of the tumor. Due to ignorance, poverty, lack of awareness and diagnostic facilities they tend to present late in developing countries. Around 13% had bladder involvement due to the chronicity of the tumor. Large dumbbell tumors in this series also presented as posterior cervical lump. One was a very unusual tumor in the cervical region that was extending anteriorly and although it was limited to the prevertebral fascia, due to vascularity it could not be removed even through the anterior cervical approach.

***Diagnosis*:**[16-19] Diagnosis is done with an MRI that shows the vertebral level, number, size, vascularity, relation to surrounding structures, the status of spinal cord and in chronic cases extradural spread. It will also show the dumbbell nature if any and similar findings in the extra spinal space. MRI shows fluid signal intensity with mild hyperintensity on T2WI and equal or low intense on T1WI. There may be uniformly strong enhancement or heterogenous or ring type lesion especially in cystic variants. Although few studies have correlated heterogenous MRI findings with pathological Antoni A or B types others refute these findings. CT can show the relation of the vertebral foramen and adjacent bone deformity in dumbbell or extradural tumors. Angiography may also be needed in some cases where the vertebral artery may be encompassed.[1]. Interestingly in the cervical region there was equal sex distribution whereas in the rest it was dominated by the male sex. Those tumors with extensive postero-lateral myofascial extension and anterior bone erosion termed are called giant invasive spinal schwannoma.[9] In our series the lumbar region was the most common followed by thoracic and in males were more common in all levels except for the cervical level where it was equal. The majority were intradural and only four were extradural.

Surgery: Surgery in one or two settings, anterior or posterior combined remains the mainstay of treatment with total excision leading to permanent cure. Localization can be done with radiological markers and plain skiagram or with help of spinal neuro-navigation. Classical laminectomy for smaller segments and laminoplasty for longer level are the surgical techniques of choice. Depending on the lesion the other options are far lateral, suboccipital, trans-peduncular, costo-transversectomy and combined with transthoracic or retroperitoneal approaches.[1] For tumors that involve more than 25% of the vertebra may need additional spinal instrumentation.[20-21] Recent trends in minimal invasive spine surgery have allowed for removal with help of smaller incisions. The technique is of course limited to expert as the closure of dura, bleeding, large size and dense adhesiolysis can be challenging.

The tumor can be gently separated and delivered out followed by either an intracapsular first or extracapsular excision with or without debulking. In our series except for 2 cases all were removed totally. Smaller tumors were removed with extracapsular and larger with intracapsular approach. Gentle traction on one end of the capsule and delivery of the tumor out helps in clearer view of surrounding structures and safer resection. The nerve root usually has another root attached to its capsule and it is pertinent that it be save. The feeding vessels exit parallel to the nerve root and must be controlled early to reduce the incidence of intraoperative bleeding. Unlike previous studies which showed no deficit with parent nerve cutting newer studies have shown that some nerves may have motor components that can lead to motor deficits postoperatively. The past view that the tumor leads to stunning of the parent nerve that leads to the surrounding nerves taking their function may no longer be valid.[1, 22]

The present view is that the parent nerve be saved in all cases possible. We have saved the nerve root in the majority and in those where it was cut the ends were approximated with 10-0 polypropylene. The method advised by us is to cut as near as possible so that the ends do not cause traction which can lead to difficulty in suturing, healing, recovery and postoperative clinical improvement. Although total excision is the rule it

may not be possible in those cases that are extensively large and their chronicity leading to dense adhesions to surrounding structures. It may also not be possible in cases with extradural infiltration, intra vertebral scalloping, inflammation, involving major vascular structures and in those that recur primarily or after radiotherapy. In such cases maximal excision with preservation of neurological function is to be desired. The monoclonal antibody level of Ki-67 more than 3 has been associated with higher recurrence rates in some studies.[24-27]

Neurophysiological monitoring: many centres now advice the routine use of neurophysiologic monitoring. This can be done preoperatively to assess the sensory or motor involvement or intraoperatively to avoid severing the nerve and causing deficits. Only half of the cases in this series had preoperative monitoring and none were monitored during surgery. Except for three cervical cases all those studied had normal studies. Motor evoked potential, Sensory evoked potential or Electromyography can be used in combination depending on the spinal location of the tumor with significant motor root function preservation has been reported with their use.[1] It is thus advisable that all cases undergo intraoperative neurophysiological monitoring or preserve the nerve at all costs as in this series.

Radiotherapy: Radiotherapy or close observation remains the only option in subtotal removal or in recurrence cases.[28] In our experience scalloping of the posterior edges of adjacent vertebra, extensive extradural infiltration, dumbbell tumors adherent to neuro-vascular structures are some of the factors that leads to incomplete excision. Single-session robotic radiosurgery and three-dimensional conformal radiotherapy (3D-CRT) has been reported as a safe alternative for tumor control in recurrent cases.[29] Stereotactic body radiotherapy also has shown to be effective with radiographic control of growth of 100% at a median follow up of 18-43 months.[30] In the two incompletely operated cases in this series they were advised radiotherapy at another centre. Only one completed treatment with no improvement in the clinical symptoms.

The limitations of this study are the small number of cases, the absence of follow-up for all the cases and the lack of intraoperative use of neurophysiology. The availability of the technique in the center at present will overcome the lack of neurophysiological monitoring. Regular follow up is difficult in this country with one possible reason being that good clinical improvement after surgery amounts to cure and hence patients do not bother to return to the hospital.

Conclusion

PSS although can frequently present to the spine surgeon. Based on clinical findings the diagnosis is clinched with help of an MRI. Surgery remains the main modality of treatment either by laminectomy, laminoplasty or minimal invasive spine surgery. The nerve root of origin must be preserved in all cases which can be aided with help of intraoperative neurophysiological monitoring.

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