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Extradural spinal schwannoma in a 5 year old non NF child: A rare entity

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Abstract: Pediatric spinal schwannomas constitute 2.5-4% of all pediatric spinal tumors. They often occur in the setting of neurofibromatosis type 2 (NF-2) and are almost always intradural (with/without dumbbell extension). Rarely are they confined to extradural space alone. Till date only one case of totally extradural lumbosacral schwannoma in a non NF child has been reported. We here report the second case of totally extradural spinal schwannoma in a non NF child. A five year old female child presented with progressively increasing paraperesis and bladder symptoms, with no clinical features of Neurofibromatosis type I or II. Imaging revealed a dorsolumbar extradural lesion from DV11 to LV1. Patient underwent laminoplasty with total excision of tumour. Intraoperatively, the tumor was confined to extradural space alone. Histopathology confirmed Schwannoma. This case highlights that possibility of schwannomas should be considered as a differential diagnosis for intraspinal extradural tumors even in children without NF.

Key words: Extradural Schwannoma, Neurofibromatosis – 2, Pediatric Spinal Schwannoma

Introduction

Spinal tumors constitute 1-10% of all pediatric central nervous system tumors. (7, 9) Schwannomas comprise about 30% of primary intraspinal neoplasms. (5) Pediatric spinal schwannomas/neurofibromas constitute 2.5-4% of all pediatric spinal tumors (6), and only 0.7% of all schwannomas occur in children. Pediatric spinal schwannomas often occur in the setting of neurofibromatosis type 2(NF-2). (8) They usually present as intradural mass lesions (with/without dumb-bell shaped extensions) and are rarely confined to the extradural space alone. (3) Till date only one case of giant totally extradural lumbosacral schwannoma in a non NF child has been reported. (4) We are reporting a rare case of 5 year old non NF female child of totally extradural schwanomma. This is probably the second such reported case.

Case History

A five year old female child presented to us with subacute onset and progressively increasing weakness in bilateral lower limbs and frequent falls of 3 months duration. She

had developed dribbling of urine for the last 7days. On examination she had spastic paraparesis (MRC grade 4/5) with 50% loss of all modalities of sensation in Left L5, S1, S2 dermatomes.Deep tendon jerks were brisk in bilateral lower limbs and plantars were extensor bilaterally. There were no clinical features suggestive of Neurofibromatosis type I or II.

X-ray Dorsolumbar Spine showed widened DV12- LV1 inter vertebral foramina and scalloping of DV12 vertebral body (Figure 1). Contrast MRI showed an extradural lesion extending from DV11 to LV1; it was isointense to hypointense on T1 weighted images and heterogenously enhancing on post contrast imaging (Figures 2A & 2B). The lesion was seen to expand the spinal canal with extension of the tumor through the left

DV12-LV1 neural foramen with associated widening of foramen and left paravertebral extension. The spinal cord was severely compressed by the tumor and displaced anteriorly towards right.

Patient underwent laminoplasty DV10 – LV1 with total excision of tumor. Intra operatively the lesion wass totally extradural, soft, moderately vascular, yellowish pink in colour and extending from D11-L1 with left paravertebral extension through DV12-LV1 foramen. The dura was opened and it was confirmed that there was no intradural component (Figure 3). Histopathological examination showed monomorphic spindle cells arranged in parallel arrays in a variably collagenous stroma with verrocay bodies. Final opinion was compatible with schwannoma. Post Operatively patient improved neurologically with power of MRC 5/5. Post Op MRI showed complete resection of tumor (Figure 4). Urinary symptoms also improved. Patient is on regular follow up for past 3 months and has no neurological symptoms suggestive of recurrence.

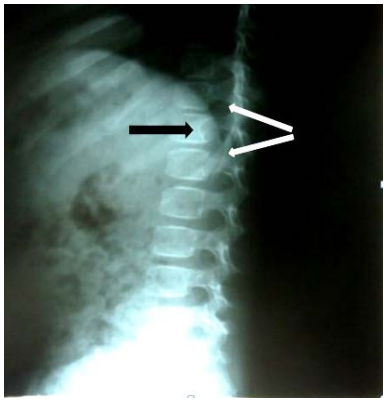

Figure 1 - X Ray Dorsolumbar spine showing widened intervertebral foramen (white arrows) and scalloped vertebral body (black arrow)

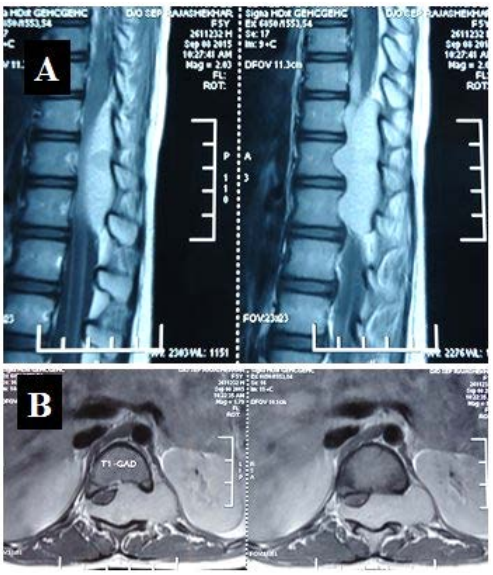

Figure 2 - Pre Op Contrast MRI. Sagittal (A) and axial (B) images showing the extradural tumor with transforaminal extension

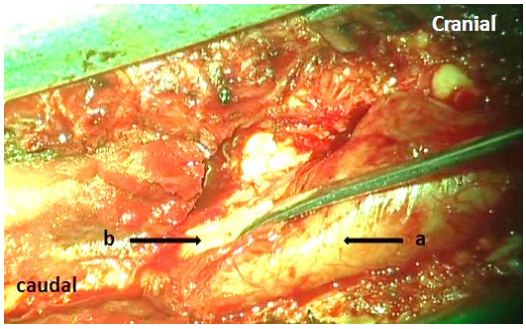

Figure 3 - Introperative image showing totally extradural tumor (a) and compressed spinal cord (b)

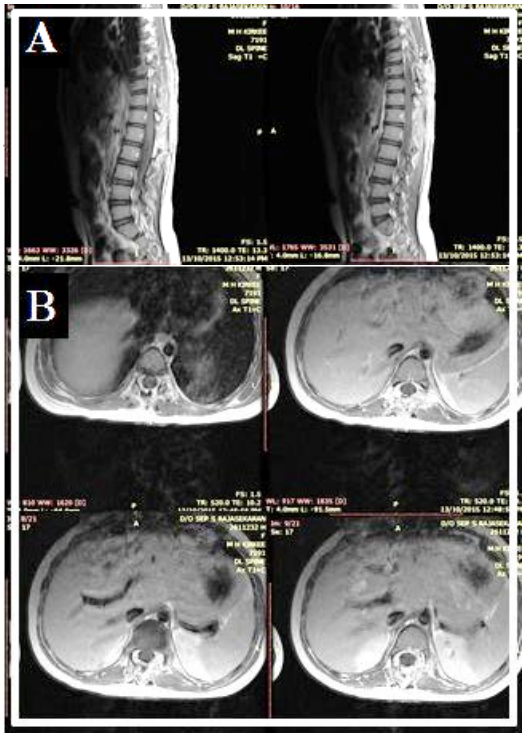

Figure 4 - Post Op contrast MRI showing no residual lesion (A- Sagittal, B - Axial)

Discussion

Schwannomas are more commonly found in the intradural, extramedullary space. Extra dural schwannomas have been reported in adults. (1) In pediatric age group, schwanomas are very rare and when they occur they are intradural and generally in the setting of NF-2. Common extradural spinal tumors in paediatric population are sarcoma, neuroblastoma, teratoma, ganglioneuroma and lymphoma. (3) There is no significant prevalence difference between males and females. However, some studies indicate a higher incidence of spinal schwannomas in males. (2) Extradural schwannomas in pediatric age group have rarely been reported and they are generally see in NF children. To our knowledge, till date only one case of giant extradural lumbosacral schwannoma in a non NF child has been reported by Kataria et al. (4) Their patient was an eight year male who presented with backache. Clinically there was no neurological deficit. Radiological evaluation revealed a large extradural mass from L4 to S2 level with paravertebral extension. The patient underwent L3 to S2 laminectomy with complete excision of the spinal part of the lesion and partial excision of the paravertebral part, with good clinical result. Our patient is a five year old female child, tumor was extradural only and was not associated with Neurofibromatosis. We believe this is the second such reported case. Postoperative results are consistent with literature, which suggests that nerve sheath tumors of the spine are often single, benign lesions that are straightforward to remove and are associated with a good postoperative outcome. Patients with intra spinal extradural schwannoma require early intervention in order to prevent irreversible neurological deficits especially when they start exhibiting neurological signs and symptoms. (3, 6, 8)

Conclusion

Schwanomas are rare in pediatric age group and when they are there they occur in NF children. Even in the NF children they are rarely, totally confined to extradural space. Ours is probably the second such reported case. Though histologically benign they can produce profound neurological deficits. Microsurgical excision for spinal schwannomas usually results in good postoperative functional outcomes. This case highlights that possibility of schwannomas should be considered as a differential diagnosis for extradural intraspinal tumors even in children without NF.

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