Paediatric calcified intramedullary schwannoma at conus: A common tumor in a vicarious location

Aniruddha Tekkatte Jagannatha, Krishna Chaitanya Joshi, Shilpa Rao, Umesh Srikantha, Ravi Gopal Varma, Anita Mahadevan

Department of Neurosurgery, M S Ramaiah Institute of Neurosciences, M S Ramaiah Medical College, 1Department of Neuropathology, National Institute of Mental Health and Neurosciences, Bengaluru, Karnataka, India

Address for correspondence: Dr. Aniruddha Tekkatte Jagannatha, Department of Neurosurgery, M S Ramaiah Institute of Neurosciences, M S Ramaiah Medical College, New BEL Road-54, Bengaluru - 560 054, Karnataka, India. E-mail: anirudhhmc@yahoo.co.in

ABSTRACT

Spinal schwannomas are commonly intradural extramedullary in location. As Schwann cells are not common in the central nervous system, intramedullary schwannomas are a rare entity. In adults, an estimated sixty cases have been reported in English literature. They are rarer in children (less than ten cases), and preoperative diagnosis becomes a prerogative in achieving total excision. Cervical cord is a common location and less commonly they occur in the conus. We report a rare case of calcified conus intramedullary schwannoma in a child without neurofibromatosis, who presented with conus-cauda syndrome of 1-year duration. Literature has been reviewed regarding its origin, pathophysiology, radiological features, and surgical management. This child underwent laminotomy and subtotal resection of the lesion. Histopathologically, tumor had typical features of schwannoma and was positive for S-100 immunoperoxidase. We believe that schwannoma needs to be considered in the preoperative differential diagnosis of a conus tumor in children as complete excision is possible in these benign tumors, thus affecting a cure.

Key words: Calcification, conus lesion, differential diagnosis, foot drop, intramedullary schwannoma

Introduction

Spinal intramedullary schwannomas are well described in adults and have been reported as early as in 1932.[1] Twelve percent of these occur in patients with neurofibromatosis, and over sixty cases have been reported outside neurofibromatosis.[2,3] These are rarer in children and are slow growing. We report a rare case of pediatric intramedullary calcified xanthomatous schwannoma managed successfully at our center.

Case Report

An 11-year-old male child presented with conus-cauda syndrome of 1-year duration. He presented with weakness in both lower limbs and difficulty in walking with the involvement of bladder and bowel. On examination, he had proximal muscle weakness, bilateral foot drop with sacral sensory loss. His ankle reflexes were sluggish and plantars were mute. He...
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had no neurocutaneous markers. His magnetic resonance imaging [Figure 1] of the spine revealed intramedullary tumor opposite D11–D12 vertebra, hyperintense on T1-weighted images with well-defined areas of hypointensity. There was a syrinx noted higher above, at D8–D9 level. On T2-weighted images, calcification was seen (hypo), and on contrast images, the lesion was partially coalescing and intensely enhancing. A provisional diagnosis of ependymoma, astrocytoma, or a calcified tuberculoma was made.

He underwent D10–D12 laminotomy and near total excision of the tumor. The tumor was relatively avascular grayish-white, firm with well-defined plane of cleavage. It had calcification superiorly, and xanthochromic thick mucinous material was seen with in the tumor. As dense adhesions were noted at the superior pole, complete excision was not possible. Interestingly, histopathological examination [Figure 2] of the tumor specimen revealed round hyalinized mass with entrapped nerve roots, with a few anterior horn cell containing lipofuscin. Large foamy histiocytes, reticulin-rich Schwannian fibrillary stroma, and calcification were noted. No giant cells or granulomas were seen and stains for acid-fast bacilli and fungal hyphae were negative. Immunohistochemistry was positive for S-100 (spindle cells) and CD-68 (foamy histiocytes). A diagnosis of xanthomatous schwannoma was made. Patient’s postoperative course was uneventful. He was recovering well at 6 months follow-up.

**Discussion**

Intramedullary schwannomas are rare and constitute a 1.1% of all spinal schwannomas.\(^2\) Usually, it is seen in males and has been described in all ages from birth to 78 years.\(^2,4,5\) They are slow-growing tumors and commonly cervical in location (61%). The frequency drops as we move down to thoracic (29%) and lumbar spinal cord (10%).\(^1,4\)

Many theories have been proposed regarding the origin of intramedullary schwannoma. It is proposed that the Schwann cells may have originated from the small nerves on the blood vessels (vasa nervorum), along the vagus nerve fibers, or along the posterior spinal nerve roots. Other possibilities include an embryonic cell rest origin or from the mesodermal pia mater cells or from disordered migration of neural crest cells during closure of spinal cord. They also may have originated in the cells at dorsal root entry zone migrating inward into the parenchyma.\(^4,6\)

Outside neurofibromatosis, only eight pediatric cases have been reported in the PubMed published English literature, including an extensive thoracolumbar congenital intramedullary schwannoma.\(^4,5,7-9\) As it is rare, preoperative diagnosis is often negative, and most of the times, it is a histopathological surprise. In an intramedullary tumor, intense contrast enhancement, well-defined plane, absence of syrinx, cystic degeneration, and presence of an extramedullary component with enhancement along a nerve root suggest schwannoma.\(^1,13\) Mukerji et al. reported on a pediatric intraspinal (intra-extramedullary) cervical dumbbell schwannoma operated in two stages where the extramedullary component was excised in the first surgery and then the intramedullary component. Intraoperative frozen section is of good help in diagnosing it during surgery. Typically, they have well-defined plane of cleavage and total excision is possible.\(^9\)
Eljebbouri et al. reported good outcome in a 10-year-old male child with dorsal intramedullary schwannoma after complete excision. However, sometimes, dense adhesions may prevent achieving a total excision, like in the present case. Our patient’s lesion is rare as it was a calcified conus-epiconus tumor with xanthomatous and cystic changes and an associated syrinx. This may represent long-standing degenerative changes within the tumor and a similar case has been reported by Shenoy and Raja. In cases of spinal schwannomas, calcification is rare and is reported in fewer than ten cases. We believe that schwannoma needs to be considered in the differential diagnosis of a calcified conus lesion in children as total excision is possible.

**Conclusion**

This report of a rare calcified intramedullary schwannoma highlights the diversity of intramedullary tumors which possibly occur at conus-epiconus location in a child. A high degree of suspicion is needed to consider this in the differential diagnosis as total excision is curative and possible.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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