Giant Lumbosacral Neurofibroma in a Child

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Abstract

Neurofibromas account for 16-30% of all spinal tumours and 13.7% of spinal tumors of nerve sheath origin. Occurrence of giant spinal neurofibroma in pediatric age group is rare. Complete excision with good neurological recovery is a rule rather than exception. Neurofibroma in lower lumbar and sacral regions can enlarge significantly and can further extend into the paravertebral and presacral space. We report a case of giant extradural lumbosacral neurofibroma in an 11-year-old female child without neurofibromatosis which was treated successfully with surgical intervention without persistent neurological deficit.

Keywords: Extradural mass, giant neurofibroma, spinal tumor

Case Report:

An 11-year-old female child presented with complaints of low backache for 3 years. Neurological examination of the patient revealed swelling and tenderness at L5 and sacral spinous processes. Plain X-ray of the lumbosacral spine (Figure 1) revealed increased interpedicular distance at L5 level and scalloping of the posterior surface of L5 and S1,2 vertebral body.

Fig.-1(a and b): Plain radiographs of the lumbosacral spine showing increased interpedicular distance at L5 level and scalloping of the posterior surface of L5 and S1,2 vertebral body.

Introduction

Neurofibromas typically present most commonly as a cutaneous nodule, less often in a peripheral nerve, occasionally in spinal roots, and multiple neurofibromas are typically associated with neurofibromatosis (NF) ¹,1-3. Histologically, neurofibromas are composed of schwann cells, fibroblasts, and perineurial cells.²,⁴,⁵

Neurofibromas account for 16-30% of all spinal tumours and 13.7% of spinal tumors of nerve sheath origin. The thoracic region is the most common site for neurofibromas, followed by the cervical and lumbar regions. Only 1-5% of all spinal neurofibromas are localized in the sacral region.¹,²,⁷ Most nerve sheath tumors are small, solitary, and benign and rarely exceed 6 cm in diameter.

Occurrence of giant spinal neurofibroma in pediatric age group is rare.¹² Complete excision with good neurological recovery is a rule rather than exception. Neurofibroma in lower lumbar and sacral regions can enlarge significantly and can further extend into the paravertebral and presacral space. We report a case of giant extradural lumbosacral neurofibroma in an 11-year-old female child without neurofibromatosis which was treated successfully with surgical intervention without persistent neurological deficit.

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interpedicular distance at L5 level, and scalloping of the posterior surface of L5, S1 and S2 vertebral body. Magnetic resonance imaging (MRI) of the lumbosacral spine (Figure 2) showed an extradural mass against L5 to S3 vertebral bodies with significant left paravertebral and presacral component. The mass was hypo to iso intense on T1-weighted images and hyper intense on T2-weighted images. There was mild heterogeneous enhancement of the mass after contrast. Patient underwent L5 to S3 laminectomy with total excision of the mass from the posterior approach on 02.08.2016. Posterior lamina of L5 and sacral laminae were found to be thinned out. The tumour was more on the left side and caudal portion of the tumour was found in presacral space through deficient part of S3. The tumour was loosely adherent to the surroundings and was separated carefully from nerve roots. The tumour was firm, encapsulated, less vascular, and entirely extradural measuring 12 cm X 5 cm. (Figure 3,4) Patient was relieved of pain after the surgery.

Histopathological examination of the tumor revealed neurofibroma, a benign tumor consists of spindle shaped cells with wavy nuclei arranged in fascicles. On 2nd postoperative day she developed retention of urine, relieved by catheter and on 5th POD she noticed faecal incontinence. She was discharged on 38th POD after resolving faecal incontinence but bladder dysfunction still persisting which was resolved after 3 months. Patient was remained asymptomatic after 1 year of follow-up.

Fig.-2 (a,b,c): MRI of the lumbosacral spine (a) showed an extradural mass against L5 to S3 vertebral bodies with significant left paravertebral and presacral component.

Fig.-3: a- Intraoperative photographs. b- Tumour measuring 12 cm X 5 cm after total removal
Discussion:
Neurofibromas are composed of a mixture of cell types including schwann cells, perineurial-like cells, and fibroblasts. To distinguish neurofibromas from schwannomas and neuromas, careful and precise pathological examinations should be performed. Immunohistochemical staining for s-100 protein has been used classically as a marker in the differential diagnosis of neurofibromas from schwannomas. However, occasionally, s-100 protein might not reliably distinguish these tumors. Recently, the usefulness of immunohistochemical staining for Cd34 has been reported to differentiate neurofibromas from schwannomas.

The incidence of giant neurofibroma in pediatric age group is rare. Initial symptoms depend upon the location of the tumor and are due to irritation of the involved sensory nerve. Giant neurofibroma commonly present with local pain and neurologic deficit develops late. Neurofibroma of the lower lumbar and sacral regions, because of the wide canal available at these locations and relatively mobile nerve roots can enlarge significantly and can extend vertically and in the paravertebral regions.

Giant neurofibroma, because of their growth in all directions, may extend:

1. Anterolaterally into extra spinal space via the foramen which they may erode and widen,
2. Posteriorly, thinning and attenuation of dura and posterior elements and occasionally may extend posterior myo-fascial planes, and
3. Anteriorly, erode the vertebral bodies to varying extents.

No radiographic findings are pathognomonic for intraspinal neurofibroma. Plain radiographs may reveal widening of the neural foramen and the spinal canal, erosion of the pedicle, and scalloping of the adjacent vertebral body. On MRI, signal characteristics of a typical neurofibroma are: T1W: Iso to hypo intense to that of spinal cord. T2W: Typically hyper intense and often heterogeneous in pattern of intensity. T1+Contrast: Shows heterogeneous enhancement with evidence of hemorrhage, calcification, or cyst formation.

Complete excision with good neurological recovery is a rule rather than exception. Although neurofibroma arises from the nerve tissue, only in 50% cases a direct relationship with a nerve is found. It is usually possible to preserve some fascicles of the nerve root, although sometimes section of the entire nerve root is required, usually in invasive giant varieties. Kim et al. reported that sacrificing an involved nerve root does not cause increase in the neurological deficits as the involved nerve root is nonfunctional.

Conclusion:
Giant neurofibromaoma are rare in children, complete excision of the tumor should be the goal of surgery. We recommend instrumentation only if the spine is unstable otherwise a wait and watch protocol be followed as spontaneous remodeling of the scalloped vertebral bodies is well known.

References:
15. Hirose t, Tani t, Shimada t, Ishizawa K, Shimada s, Sano t: immunohistochemical demonstration of emA/ Glut1 positive perineurial cells and Cd34-positive fibroblastic cells in peripheral nerve sheath tumors. Mod Pathol 16: 293–298, 2003