Case Report

International Journal of Neurology and Neurosurgery
Volume 9 Number 1, January - June 2017
DOI: http://dx.doi.org/10.21088/ijnns.0975.0223.9117.6

Isolated Sacral Nerve Root Ependymoma

Nitin Dange*, Vikas Singh, Achal Gupta***, Aniruddh Agarwal******

Abstract

Ependymomas commonly arise from conus medullaris, cauda equina and filum terminale. Ependymoma involving single nerve root is rare. We report a case of young male with disabling low back pain without motor- sensory deficits whose magnetic resonance imaging (MRI) showed a intradural lesion opposite L2-3 vertebral body which was confined only to the S1 nerve root and was resected without incurring any post operative deficits. Pathologic examination was suggestive of myxopapillary ependymoma. Patient was asymptomatic postoperatively and MRI did not show residual disease. Ependymomas can cause central nervous system dissemination (CNS), recur and undergo malignant transformation, knowledge of which is essential for management and follow up. We believe this is the first reported case of S1 nerve root Ependymoma.

Keywords: Ependymoma; Nerve Root; Sacral.

Introduction

Ependymomas account for 60% of glial spinal cord tumors and comprise 90% of primary tumors in the filum terminale, conus medullaris and cauda equina [1,2]. Although the filum terminale is of neuroectodermal origin, these tumors are often categorized as extramedullary from the anatomical and surgical point of view [3]. Spinal ependymomas arise from ependymal cells lining the central canal. In the lumbosacral region, the majority of ependymomas arise from the ventriculus terminalis of filum terminale and can grow to involve adjacent nerve roots [3]. Isolated nerve root ependymomas without concomitant or contiguous involvement of the filum terminale are exceedingly rare and isolated sacral nerve root ependymomas have not been reported in literature till date. Here we report which is probably the first reported case of isolated sacral nerve root ependymoma. Although rare, it is important to keep ependymomas in the differential for lumbosacral nerve root tumors as they have a propensity for recurrence and malignant transformation even with gross total resection.

Case History

Twenty nine year old male was seen with progressive low back pain especially at night for two months with no motor or sensory deficits. MRI evaluation revealed intradural lesion opposite L2-L3 vertebral body, iso intense on T1 and iso- hypo intense on T2 **(Figure 1)** with enhancement on contrast **(Figure 2)**. Rest of the neuraxis evaluation was normal. L2-L3 laminectomy was done and dura opened. Expansile grey-white lesion was seen arising from single nerve root **(Figure 3)**. Intraoperative neurostimulation indicated the nerve root to be right S1 motor root. Gross total excision of the tumour was performed with preservation of nerve root. Post operatively patient did not develop any deficits and was relieved of pain. Histopathologic examination was suggestive of WHO grade 1 myxopapillary ependymoma with perivascular pseudorosettes **(Figure 4)** and was glial fibrillary acidic protein, S100 and vimentin positive. Follow up MRI was performed that confirmed gross total excision of the tumour.

Author's Affiliation: *Consultant Neurosurgeon **Clinical Associate ***DNB Neurosurgery Trainee, Lilavati Hospital and Research Centre, Mumbai, India. ****Medical Student, Topiwala National Medical College, Mumbai, India.

Reprint Request: Nitin Dange, Consultant Neurosurgeon, Department of Neurosurgery, Lilavati Hospital and Research Centre, Bandra Reclamation, Bandra west, Mumbai 400050.
E-mail: docdnitin@yahoo.com

Received on 31.12.2016, **Accepted on** 09.01.2017

© Red Flower Publication Pvt. Ltd.

Discussion

Ependymomas are thought to originate from rests of ectopic ependymal cells which are believed to be excluded from the neural tube during closure [4].

Ependymomas are the most common intradural spinal cord tumors in adults [5]. Myxopapillary ependymomas are most common subtype and occur almost exclusively in the filum terminale and conus medullaris [6] . Ependymomas arising outside the

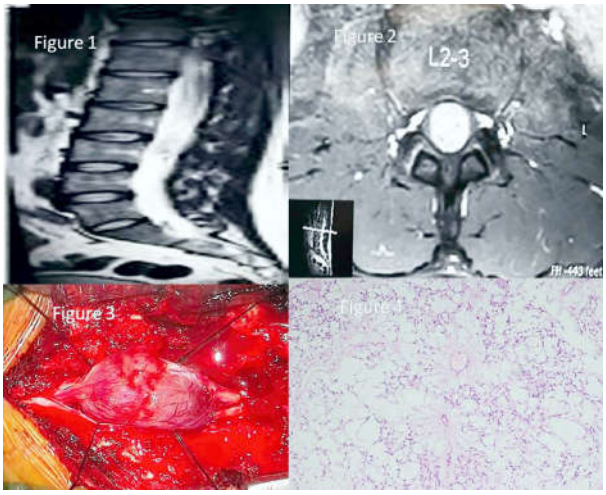

Fig.1: T2 saggital section showing iso-hypointense lesion opposite L2-3 vertebral body. **Fig. 2**: Contrast axial image showing enhancement on contrast. **Fig. 3**: Intra operative picture showing attachment to S1 root only. **Fig. 4**: Haematoxylin and eosin staining showing perivascular pseudorossette

region of the conus medullaris, cauda equina, and terminal filum, or developing from ectopic ependymal cells are highly unusual [7]. They are usually located within the filum terminale and can extend into conus medullaris. Small tumors tend to displace the lumbosacral nerve roots, whereas large tumors may encase them. Intradural lumbosacral ependymomas can spread throughout the CNS through cerebrospinal fluid pathways but rarely metastasize beyond it [8]. These tumours are benign tumours but can rarely undergo malignant transformation.

Most nerve root tumours are meningiomas or schwanommas. Metastatic adenocarcinoma, melanoma, lymphoma, and sarcoma have been reported. There are only 3 reported cases of isolated lumbar nerve root ependymomas [9]. This we believe is the first reported case of sacral nerve root ependymoma.

It is essential to keep in mind that ependymoma can undergo malignant transformation and spread throughout the CNS. Thus the need for complete neuraxis evaluation and gross total excision of the tumour cannot be overemphasized.

Key Messages

In young patients with back pain, nerve root lesions should be considered as diffential and treatment goal should be maximal resection without deficits.

References

1. Fassett DR, Schmidt Benzagmout M, Boujraf S, Oulali N: Intradural extramedullary ependymoma: Is there constantly a hormonal relationship?. Surg Neurol. 2008; 70:536-8. 10.1016/j.surneu.2007.05.055.
2. Cerase A, Venturi C, Oliveri G, De Falco D, Miracco C. Intradural extramedullary spinal anaplastic ependymoma. Case illustration. J Neurosurg Spine. 2006; 5:476.
3. El-Mahdy W, Kane PJ, Powell MP, Crockard HA. Spinal intradural tumours: Part I – extramedullary. Br J Neurosurg, 1999; 13:550–557.
4. Norstrom CW, Kernohan JW, Love G (1961) One hundred primary caudal tumors. JAMA, 1961; 178:1071–1077.
5. Koeller KK, Rosenblum RS, Morrison AL. Neoplasms of the spinal cord and filum terminale: radiologic-pathologic correlation.RadioGraphics, 2000; 20:1721–1749.
6. Kernohan J: Primary tumors of the spinal cord and intradural filum terminale. Cytology and Cellular Pathology of the Nervous System. Penfield W (ed): Hoeber, New York; 1932; 3:993–999.
7. Osborn AG. Diagnostic neuroradiology, Chap 21: Tumors, cysts, and tumour like lesions of the spine and spinal cord. Mosby, Boston, 1994.p.876–918.
8. Fassett DR, Schmidt MH. Lumbosacral ependymomas: a review of the management of intradural and extradural tumors.Neurosurg Focus2003; 15(5):E13.
9. Tripathy P, Mohapatra D, Mohapatra S: Primary intradural extramedullary ependymoma: report of two cases and review of the literature. Neurol Neurochir Pol. 2011, 45:397-401.