Myxopapillary Ependymoma with Drop Metastasis in a Young Child

Athisayaraj Patrick Joshua, Murugaiyan Nagarajan, Ramesh Banu

Valavadi Narayanasamy Cancer Centre, G. Kuppuswamy Naidu Memorial Hospital, Coimbatore 641037, Tamil Nadu, India.

Abstract

Background: Myxopapillary ependymoma (MPE) is a rare variant of ependymoma. It has an indolent course in adults but in children it is aggressive. Because of its rarity there are no guidelines for management of metastatic MPE. Hence we report this case of MPE with drop metastasis treated with cranio-spinal irradiation (CSI) after subtotal resection (STR). Case Report: A 9 year old girl was evaluated elsewhere for low backache. MRI whole spine showed intra-dural mass compressing the conus and another lesion in the sacral spinal canal. She underwent STR. Post-operative histopathological examination was consistent with myxopapillary ependymoma. She was treated with CSI. Conclusion: Cranio spinal irradiation is essential for the treatment of metastatic MPE in pediatric population.

Keywords: Central Nervous System, Ependymoma, Low Backache, Magnetic Resonance Imaging, Spinal Canal.

Introduction

Myxopapillary ependymoma (MPE) is a rare variant of ependymoma constituting around 13% of ependymomas. These benign tumors are characterised by their slow indolent growth. It usually affects adult males in their 3rd or 4th decade of life. Surgery is the standard treatment of choice and adjuvant RT improves local control [1]. These tumors are rare and aggressive in children [2]. Hence we report this case of MPE in a 9 year old child.

Case Report

A 9 year old girl was initially evaluated elsewhere for complaints of sudden onset of low backache radiating to both lower limbs for two weeks. Clinically she had no neurological deficit. MRI thoracolumbar spine showed well defined enhancing intra-dural mass measuring 18×16×4 mm compressing the conus and proximal cauda equine nerve roots. There was another enhancing intra-dural mass within sacral spinal canal measuring 5×6×10 mm suggestive of drop metastasis and an intra-medullary cystic component 10×10×13 mm in the distal thoracic spinal cord and conus [Fig.1]. There were syringomyelic changes in the mid and distal thoracic cord. She underwent T11-L2 laminoplasty and subtotal excision of tumor. Her post-operative period was uneventful. Post-operative MRI spine showed 7×6 mm residual disease within spinal canal at L2 level and enhancing intra-dural lesion 13×8 mm in the spinal canal at S1-S2 level and persistent intra-medullary cystic lesion in the lower thoracic spine. She presented to our hospital for further management. Clinically she was asymptomatic with no focal neurological deficit. Slide review done in our institute was consistent with myxopapillary ependymoma and immunohistochemistry (IHC) done was positive for glial fibrillary acidic protein (GFAP) and negative for epithelial membrane antigen (EMA) [Fig. 2,3].

In view of metastatic disease at presentation and residual disease she was treated with craniospinal irradiation (CSI). She received 36 Gray (Gy)
in 18 fractions to the entire neuraxis followed by
10.4 Gy in 6 fractions boost to the residual gross
disease and metastatic lesions.

Discussion

MPE is a rare variant constituting around 13% of
ependymoma cases. It more commonly affects
adults than children. Most of the spinal MPE
patients present with low backache as in our case
[3]. Most common sites are conus medullaris and
filum terminale. Pathologically it is characterized
by presence of numerous papillary projections
and mucin between tumor cells [4]. In our case
also the slides showed transversely cut papillary
projections with core of mucin. IHC was positive
for GFAP and negative for EMA which correlates
with ependymoma. Though it has been classified
as WHO Grade I tumor, in children it has more
aggressive course.

Surgery is the mainstay of treatment and
gross tumor resection is preferred. However in the
Stephen et al. case series almost all paediatric MPE
patients recurred after gross total resection (GTR)
[5]. Hence GTR alone is not sufficient to prevent
recurrence in children. Hence adjuvant treatment
is warranted to prevent recurrence. Adjuvant RT
plays a vital role to prevent local recurrence and
also to treat disseminated disease. In our case she
had disseminated disease and underwent subtotal
resection. Hence she was planned for adjuvant
radiotherapy.

Seven cases of MPE in less than 15 years of
age treated with cranio-spinal irradiation had been
reported in the literature [6-9]. Four patients had
metastatic lesions in the spinal canal at the time of
presentation as in our case and three patients had
localized disease. Of the four patients who had
metastatic disease, one had local recurrence and one
patient died of unrelated cause. The remaining two
patients were disease free for more than a year. CSI
prevents disease recurrence and it is recommended
for MPE patients with spinal metastases following
tumor resection in paediatric age group. It has the

Fig.1: Sagittal section of T2 weighted image of the spine showing intra-dural mass in the lower thoracic region and another lesion in the sacral spinal canal.

Fig.2: High power photomicrograph shows central mucin surrounded by spindle shaped cells with fibrillary cytoplasmic process.

Fig.3: GFAP immunostain highlights the neoplastic ependymal cells.
advantage of treating the disseminated disease and also it prevents recurrence.

CSI in pediatric population can cause acute and late toxicities. Tsang et al. in their retrospective study analysed the clinical outcomes and toxicities in pediatric patients who received CSI for low grade gliomas [10]. In our case during radiotherapy, the child had grade II neutropenia which was managed with granulocyte colony stimulating factor. Otherwise she tolerated treatment well. This child will be followed up for local recurrence and late toxicities.

Conclusion

Metastatic spinal MPE in children is often a clinical challenge to treat in view of local recurrence and dissemination. Adjuvant RT is indicated even after gross tumor resection to prevent local recurrence in children. CSI is warranted in patients who present with disseminated intra-spinal disease.

Contributors: APJ: literature search, data acquisition and manuscript preparation; MN: concept and manuscript review; RB: manuscript editing. MN will act as guarantor. All authors approved the final version of this manuscript.

Funding: None; Competing interests: None stated.

References

1. Agbabiwe HC, Wharam M, Batra S, Cohen K, Terezakis SA. Management of pediatric myxopapillary ependymoma: the role of adjuvant radiation. Int J Radiation Oncol Biol Phys. 2013;85:421-427.

2. Bandopadhayay P, Silvera VM, Ciarlini PD, Malkin H, Bi WL, Berghold G, et al. Myxopapillary ependymomas in children: imaging, treatment and outcomes. J Neurooncol. 2016;126:165-174.

3. Petersen D, Lystad RP. Spinal myxopapillary ependymoma in an adult male presenting with recurrent acute low back pain: a case report. Chiropractic & Manual Therapies. Chiropr Man Therap. 2016;24:11.

4. Wang H, Xie J. Pathological features of myxopapillary ependymomas in lumbar spinal canal: report of two cases. Int J Clin Exp Pathol. 2016;9:8736-8740.

5. Stephen JH, Sievert AJ, Madsen PJ, Judkins AR, Resnick AC, Storm PB, et al. Spinal cord ependymomas and myxopapillary ependymomas in the first 2 decades of life: a clinicopathological and immunohistochemical characterization of 19 cases. J Neurosurgery: Pediatrics. 2012;9:646-653.

6. Shirasawa H, Ishii K, Iwanami A, Mikami S, Toyama Y, Matsumoto M, et al. Pediatric myxopapillary ependymoma treated with subtotal resection and radiation therapy: a case report and review of the literature. Spinal Cord. 2014;52(S2):S18.

7. AlHalabi H, Montes JL, Atkinson J, Farmer JP, Freeman CR. Adjuvant radiotherapy in the treatment of pediatric myxopapillary ependymomas. Pediatr Blood Cancer. 2010;55:639-643.

8. Merchant TE, Kiehna EN, Thompson SJ, Heideman RL, Sanford RA, Kun LE. Pediatric low-grade and ependymal spinal cord tumors. Pediatric Neurosurgery. 2000;32:30-36.

9. Chinn DM, Donaldson SS, Dahl GV, Wilson JD, Huhn SL, Fisher PG. Management of children with metastatic spinal myxopapillary ependymoma using craniospinal irradiation. Pediatr Blood Cancer. 2000;35:443-445.

10. Tsang DS, Murphy ES, Ezzell SE, Lucas JT, Tinkle C, Merchant TE. Craniospinal irradiation for treatment of metastatic pediatric low-grade glioma. J Neurooncol. 2017;134:317-324.