Case Report

Pilomyxoid astrocytoma of the thoracic spinal cord in an adult: A case report and review of literature

Tamojit Chaudhuri, Kamlesh Jadava¹

Departments of Radiotherapy, and ¹Pathology, Sanjay Gandhi Post Graduate Institute of Medical Sciences, Lucknow, Uttar Pradesh, India

ABSTRACT

We present a case of pilomyxoid astrocytoma (PMA) in a 35-year-old Asian male with history of paraparesis for last 6 months. A contrast-enhanced magnetic resonance imaging of the spine revealed an intramedullary mass lesion occupying most of the thecal sac at the level of 10th and 11th dorsal vertebrae, with extensive contrast enhancement. Spinal PMA in an adult is an extremely rare entity, with only two reported cases in the literature, until date. This appears to be the first reported case of spinal PMA in an adult with isolated thoracic spinal cord involvement.

Key words: Pilocytic astrocytoma, pilomyxoid astrocytoma, thoracic spine

INTRODUCTION

Pilomyxoid astrocytoma (PMA) is a recently described central nervous system (CNS) tumor of the pediatric age group found mainly in the chiasmatic-hypothalamic region.^[1] It has been considered to be pilocytic astrocytoma (PA) until Tihan *et al.*,^[1] first summarized its features from a series of pediatric cases in 1999 and distinguished it from PAs. In the recent World Health Organization classification of CNS tumors 2007, PMA has been added into the category of histological variants of PA.^[2] As per the published reports, this tumor behaves more aggressively, has an early age of onset, higher rate of metastases throughout CNS, and poorer prognosis with shorter progression-free survival (PFS) and overall survival times, compared with PA.^[3-6] Now-a-days little is still known about its histological origin, and there is no consensus about its management.

To the best of our knowledge, there are only five adult cases of PMA being reported until date,^[6-10] and among them only

Access this article online	
Quick Response Code:	Website: www.ccij-online.org
	DOI: 10.4103/2278-0513.134497

two are of spinal origin; one involving the cervical spinal cord^[8] and the other one extending from the mid-cervical up to the lumbosacral region.^[9] Hence, probably this is the first reported case of spinal PMA in an adult with isolated thoracic spinal cord involvement.

CASE REPORT

A 35-year-old Asian male, presented with the complaints of paraparesis for last 6 months. Neurological examination on admission revealed no other abnormality, except paraparesis. Magnetic resonance imaging (MRI) showed an intramedullary mass involving the thoracic spinal cord, extending from 10th to 11th dorsal vertebrae, with extensive contrast enhancement; signal intensity on MR was uniformly hyperintense on T2 and fluid attenuated inversion recovery sequences and hypointense on T1 sequence [Figure 1]. He underwent laminectomy and tumor excision. Histological examination revealed a glial neoplasm consisting of monomorphous and piloid cells with elongated cytoplasmic precesses in a rich myxoid background [Figure 2]. Rosenthal fibers or eosinophilic granular bodies were absent. Immunohistochemistry showed these tumor cells were positive for glial fibrillary acidic protein [Figure 3]. The MIB-1 labeling index was 3%. Based on these findings, a histological diagnosis of PMA was made. In view of absence of any other lesion on screening MRI of the neuraxis, he was planned for adjuvant external beam radiation 55.8 Gy in 31 fractions

Address for correspondence: Dr. Tamojit Chaudhuri, Department of Radiotherapy, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow - 226 014, Uttar Pradesh, India. E-mail: tamojit.cnmc@gmail.com



Figure 1: Magnetic resonance imaging of spine showing an intramedullary mass involving the thoracic spinal cord, extending from 10th to 11th dorsal vertebrae, with extensive contrast enhancement



Figure 2: Photomicrograph of the tumor showing monomorphous bipolar astrocytic cells and piloid cells with elongated cytoplasmic processes in a rich myxoid background (H and E, ×400)



Figure 3: Photomicrograph showing diffuse immunoreactivity of the tumor cells for glial fibrillary acidic protein (×400)

over 6 weeks, to the tumor bed with appropriate margins, and now on treatment in our department.

DISCUSSION

Pilocytic astrocytoma is the most common pediatric CNS neoplasm with excellent long-term prognosis.^[11,12] However, not all patients with PA, experience good outcomes. A small subset of children with this tumor experience a variable clinical course with shorter disease-free survival and overall survival rates.^[1] Although it has long been recognized that particular subset of PAs behave aggressively, the reasons for this have not yet been elucidated.

Pilomyxoid astrocytoma, a recently described tumor entity of the CNS,^[1,2] may be responsible for this clinical variability. When compared with the typical biphasic histology of PA, this neoplasm demonstrates monomorphous piloid cells in a loose fibrillary and myxoid background. Moreover, PMA does not display Rosenthal fibers or eosinophilic granules, both characteristic of PA.^[1,13] This tumor entity has been designated PMA on the basis of its typical histologic appearance.

Similar to PAs, PMAs may occur anywhere along the neuraxis and affect individuals throughout childhood. PMA, however, exhibits a predilection for the chiasmatic-hypothalamic region and tends to affect very young children.^[3-5] It has also been described in the posterior fossa, diencephalon and spinal cord.^[14,15] The mean age at diagnosis for patients with PMA has been documented to be 18 months. In comparison, the mean age at diagnosis for patients with PA is 58 months.^[4]

Pilomyxoid astrocytoma generally behaves more aggressively than PA. In the first publication to clinically describe this novel tumor, long-term outcomes of 21 patients with hypothalamic PMAs were compared with those of 42 patients with PAs in the same region.^[4] Patients with PMA experienced a higher rate of local recurrence than those with PA (76% and 50%, respectively), despite having equal rates of gross total resection. Of note, the PMA group demonstrated a substantial rate of cerebrospinal fluid dissemination (14%), whereas none of the tumors in the PA group showed such spread. Patients with PMA experienced significantly shorter PFS than those with PA (mean PFS of 26 and 147 months, respectively; P<0.001). Patients with PMA also demonstrated significantly shorter overall survival than those with PA (mean overall survival of 63 and 213 months, respectively; *P* < 0.001).^[4]

The optimal treatment strategy for PMA has not yet been established. As a recently discovered tumor entity, PMA is treated in a manner similar to PA. The known aggressive biological behavior of this tumor, however, may alter its management in the future. Considering the unfortunate clinical course of many patients with PMA, the demonstrated survival advantage related to extensive resection may prompt neurosurgeons to implement more radical operative techniques when dealing with this tumor. This approach has been shown to be beneficial in children with recurrent malignant glial tumors, for which radical resection predicted a more favorable survival.^[16] The decision to use postoperative adjuvant therapy also depends on a lesion's perceived malignant tendencies, as the potential benefits of radiation therapy and chemotherapy must be weighed against their morbidity.^[17]

CONCLUSION

Pilomyxoid astrocytoma is a histological entity related to PA with a greater trend to re-growth and cerebrospinal fluid dissemination, therefore strict follow-up and oncological treatment is recommended. Until date, there are no published investigations regarding PMA genomics. This confers a tremendous potential for basic science studies aimed at further characterizing the molecular biology of these tumors.

REFERENCES

- 1. Tihan T, Fisher PG, Kepner JL, Godfraind C, McComb RD, Goldthwaite PT, *et al.* Pediatric astrocytomas with monomorphous pilomyxoid features and a less favorable outcome. J Neuropathol Exp Neurol 1999;58:1061-8.
- Brat DJ, Scheithauer BW, Fuller GN, Tihan T. Newly codified glial neoplasms of the 2007 WHO Classification of Tumours of the Central Nervous System: Angiocentric glioma, pilomyxoid astrocytoma and pituicytoma. Brain Pathol 2007;17:319-24.
- 3. Farwell JR, Dohrmann GJ, Flannery JT. Central nervous system tumors in children. Cancer 1977;40:3123-32.
- Komotar RJ, Burger PC, Carson BS, Brem H, Olivi A, Goldthwaite PT, et al. Pilocytic and pilomyxoid hypothalamic/chiasmatic astrocytomas. Neurosurgery 2004;54:72-9.

- Clark GB, Henry JM, McKeever PE. Cerebral pilocytic astrocytoma. Cancer 1985;56:1128-33.
- 6. Bhargava D, Sinha P, Chumas P, Al-Tamimi Y, Shivane A, Chakrabarty A, *et al.* Occurrence and distribution of pilomyxoid astrocytoma. Br J Neurosurg 2013;27:413-8.
- Toyoda K, Tsutsumi K, Ono T, Takahata H, Toda K, Ito M, et al. Pilomyxoid astrocytoma presenting in an elderly patient: A case report. No Shinkei Geka 2009;37:1123-8.
- Sajadi A, Janzer RC, Lu TL, Duff JM. Pilomyxoid astrocytoma of the spinal cord in an adult. Acta Neurochir (Wien) 2008;150:729-31.
- Mendiratta-Lala M, Kader Ellika S, Gutierrez JA, Patel SC, Jain R. Spinal cord pilomyxoid astrocytoma: An unusual tumor. J Neuroimaging 2007;17:371-4.
- Omura T, Nawashiro H, Osada H, Shima K, Tsuda H, Shinsuke A. Pilomyxoid astrocytoma of the fourth ventricle in an adult. Acta Neurochir (Wien) 2008;150:1203-6.
- Pollack IF, Claassen D, Al-Shboul Q, Janosky JE, Deutsch M. Low-grade gliomas of the cerebral hemispheres in children: An analysis of 71 cases. J Neurosurg 1995;82:536-47.
- Brat DJ, Burger PC. Cerebral pilocytic astrocytoma: Distinction from infiltrating fibrillary astrocytomas. Pathol Case Rev 1998;3:290-5.
- Rekate HL, Rakfal SM. Low-grade astrocytomas of childhood. Neurol Clin 1991;9:423-40.
- Komotar RJ, Mocco J, Jones JE, Zacharia BE, Tihan T, Feldstein NA, et al. Pilomyxoid astrocytoma: Diagnosis, prognosis, and management. Neurosurg Focus 2005;18:E7.
- Burger PC, Cohen KJ, Rosenblum MK, Tihan T. Pathology of diencephalic astrocytomas. Pediatr Neurosurg 2000;32:214-9.
- Wisoff JH, Boyett JM, Berger MS, Brant C, Li H, Yates AJ, et al. Current neurosurgical management and the impact of the extent of resection in the treatment of malignant gliomas of childhood: A report of the Children's Cancer Group trial no. CCG-945. J Neurosurg 1998;89:52-9.
- Fisher BJ, Leighton CC, Vujovic O, Macdonald DR, Stitt L. Results of a policy of surveillance alone after surgical management of pediatric low grade gliomas. Int J Radiat Oncol Biol Phys 2001;51:704-10.

Cite this article as: Chaudhuri T, Jadava K. Pilomyxoid astrocytoma of the thoracic spinal cord in an adult: A case report and review of literature. Clin Cancer Investig J 2014;3:329-31.

Source of Support: Nil, Conflict of Interest: None declared.