Case Report

DOI: 10.6966/EDMJ.202406_11(2).0004

Spinal Pilomyxoid Astrocytoma of Thoracic Spinal Cord Successfully Treated with Surgical Total Resection: A Case Report and Literature Review

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Pilomyxoid astrocytoma (PMA) in the intramedullary region of the spinal cord is rare, especially in adults. Total resection of PMA with good outcomes has rarely been reported. We report the case of a 27-year-old female patient who presented with thoracic spinal pilomyxoid astrocytoma. Magnetic resonance imaging showed a 2 cm intramedullary mass in the thoracic spinal cord at the T4-5 level. The patient underwent T4-5 laminectomy with tumor excision. No recurrent tumor was noted after two years of follow-up. Pilomyxoid astrocytoma frequently occurs in the hypothalamic/chiasmatic region but is extremely rare in the spine. Our case suggests that surgery with total resection may be an effective therapeutic option with good outcomes for pilomyxoid astrocytomas of the thoracic spinal cord.

Key words: pilomyxoid astrocytoma, thoracic spinal cord, spine, surgery

Introduction

Pilomyxoid astrocytoma (PMA) is a rare tumor of the central nervous system. In the past, pilomyxoid astrocytoma was regarded as a variant of pilocytic astrocytoma (PA). However, compared with PA, PMA has a more aggressive clinical course, a high recurrence rate, and early cerebrospinal fluid dissemination.[1] PMA in the intramedullary region of the spinal cord is extremely rare, especially in adults.[2-7] Here, we report a case of thoracic intramedullary PMA wherein the patient underwent successful laminectomy with total tumor resection and displayed good postoperative neurological outcome after two years.

Case Report

A 27-year-old female patient with a history of myasthenia gravis who had previously undergone thymectomy presented with voiding dysfunction and recurrent urinary tract infections during the preceding year. In addition, mild intermittent bilateral lower-limb weakness was noted. Magnetic resonance imaging (MRI) showed a 2 cm mass with

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intramedullary T2WI high signal intensity, faint contrast enhancement, and suspicious calcification at the T4-5 level (Fig. 1). Based on the imaging findings, a diagnosis of astrocytoma or other glial tumors was considered. The patient underwent a T4-5 laminectomy with total tumor excision. The tumor was soft, gray-yellowish, and gelatinous. There was a demarcation plane between the tumor and spinal cord, but the tumor attached to the spinal cord with fiber-like tissue which was excised as well. Histopathological examination showed a moderately cellular neoplasm composed of monomorphous round to oval cells in a myxoid stroma (Fig. 2). Focal vascular proliferation was identified, but mitotic activity was low. Necrosis and calcification were not observed. Rosenthal fibers and eosinophilic granular bodies were absent. Immunohistochemical staining showed diffuse positivity for glial fibrillary acidic protein (GFAP). Some tumor cells were positive for synaptophysin. The MIB-1 labeling index was 1.87%. Based on these findings, the patient was diagnosed with pilomyxoid astrocytoma. Brain MRI was performed after the cerebrospinal fluid dissemination survey, and no tumors were found. After the operation, the symptoms of voiding dysfunction improved, and no further episodes of urinary tract infection were noted. Lower limb weakness improved after 3 months. After 2 years, a follow-up MRI showed no recurrence (Fig. 3).

Discussion

PMA and PA may occur anywhere in the central nervous system, but they are usually

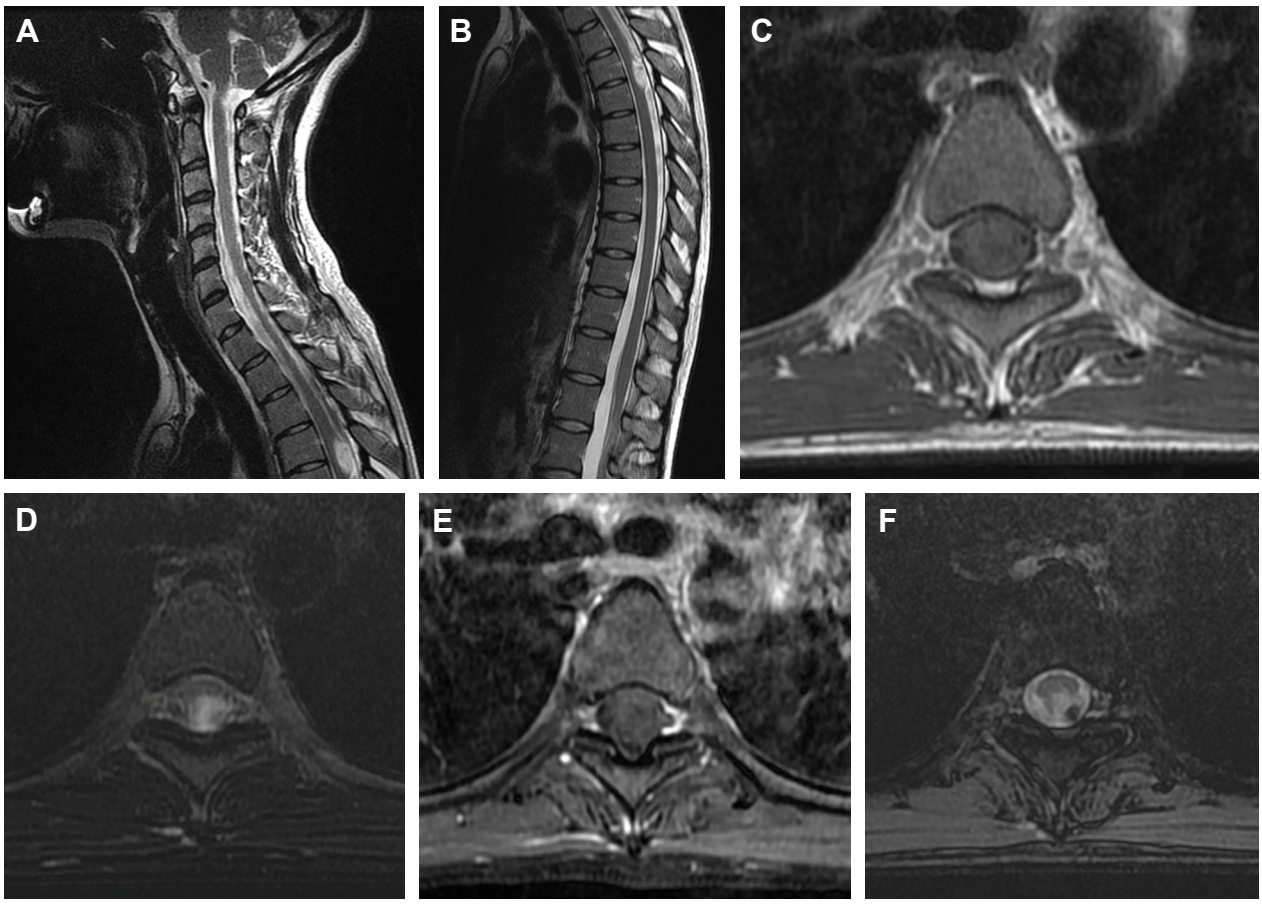

Fig. 1 (A) T2 weighted. (B) T2 weighted. Showing a 2 cm intramedullary mass with high signal intensity at the T4-5 level. (C) T1 weighted. Showing low signal intensity. (D) T2-weighted gradient-echo sequence. Low signal over the left dorsal part similar to the T2 weighted image, so calcification was suspected. (E) T1 weighted with contrast-enhancement. Showing a faint contrast enhancement.*

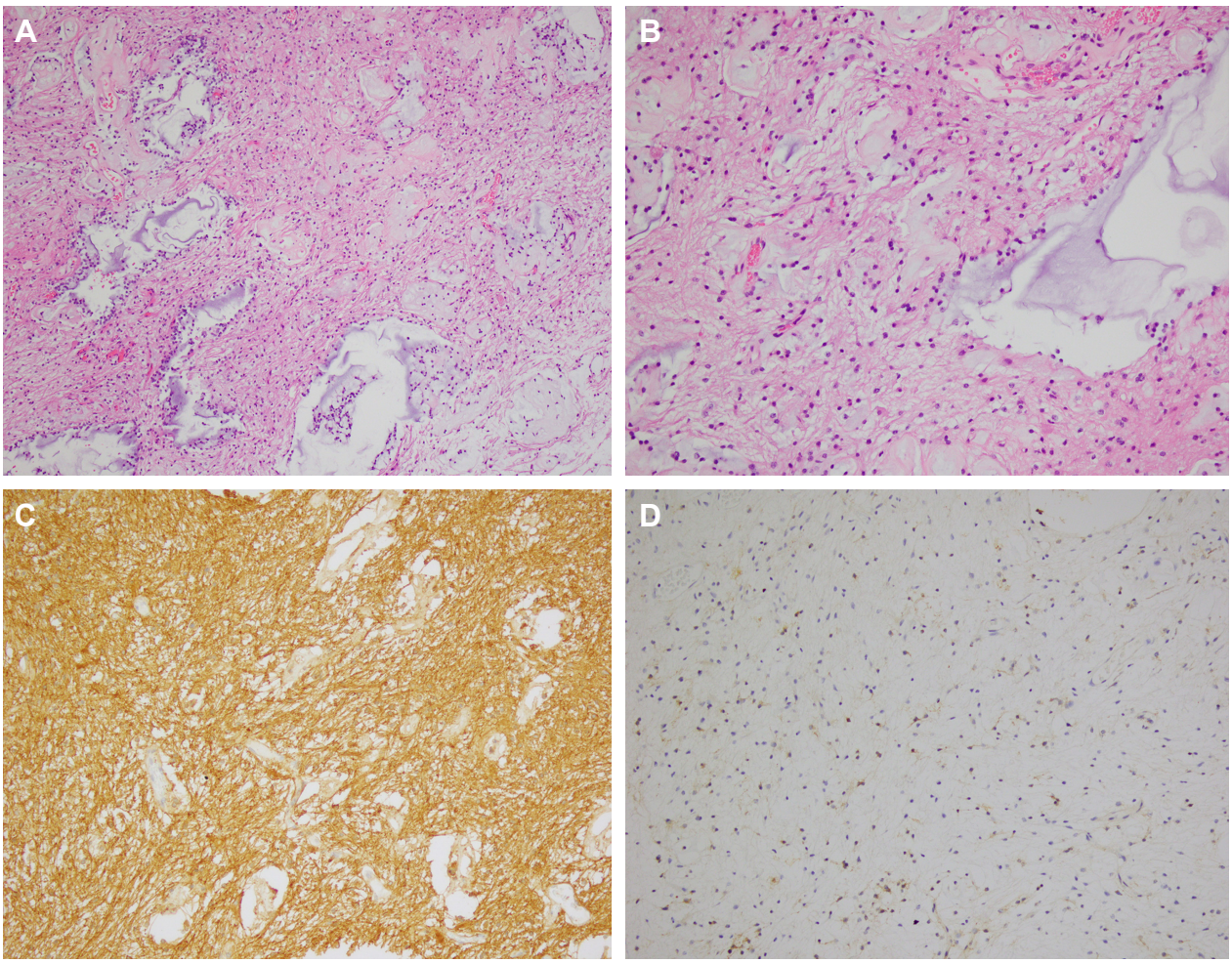

Fig. 2 (A) Hematoxylin and eosin stain; original magnification × 100. (B) Hematoxylin and eosin stain; original magnification × 200. A moderately cellular neoplasm composed of monomorphous round to oval cells in a myxoid stroma. Focal vascular proliferation was identified but mitotic activity was low. No necrosis or calcification was apparent. Rosenthal fibers or eosinophilic granular bodies were absent. (C) GFAP immunostaining; original magnification × 200. Immunohistochemical staining showed diffuse positivity for glial fibrillary acidic protein (GFAP). (D) Synaptophysin immunostaining; original magnification × 200. Some tumor cells are positive for synaptophysin. The MIB-1 labeling index was 1.87%.

seen in children of age 4 and lower and in the hypothalamic/chiasmatic region.[1] Adult patients with PMA in the spinal region are extremely rare. Compared with PA, PMA appears more aggressive and has a less favorable prognosis.[1] According to the 2016 World Health Organization (WHO) classification, it is not clear whether pilomyxoid astrocytoma should automatically be assigned to WHO grade II, and the suggestion has been made to suppress grading of pilomyxoid astrocytomas until further studies clarify their behavior.[8]

Only six adult cases of spinal PMA have been reported in the literature (Table 1).[2-7] One of these tumors involved the entire spine, one involved the cervical cord, one involved the T-L spinal junction, and three were located in the thoracic cord. In the present case, the tumor was located in the thoracic spinal cord. MRI of the brain and the whole spine did not reveal any tumors in other regions. Thus, this tumor originated in the thoracic cord, even though PMAs favor metastasis into the cerebrospinal fluid space. Optimal treatment for PMA has not yet been determined. Previous case reports showed patients undergoing subtotal resection or biopsy with or without radiotherapy or chemotherapy, and the outcomes were varied.[2-7] The addition of adjuvant radiation therapy is controversial. Some clinicians recommend safe

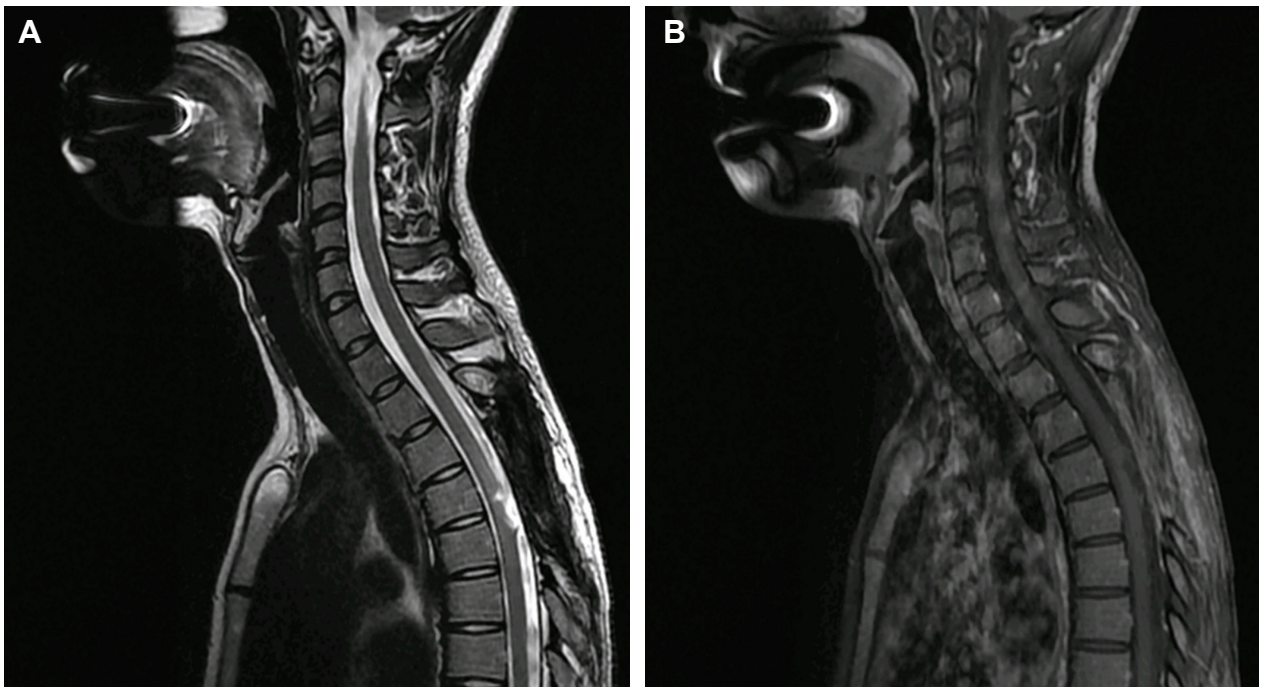

Fig. 3 (A) T2 weighted. (B) T1 weighted with contrast enhancement. No recurrent lesions were apparent in either image.

surgical resection of the tumor plus both radiotherapy and chemotherapy to improve survival; however, currently no strong evidence supports this management.[9] We did not treat our patient with radiotherapy or chemotherapy. For newly diagnosed glial tumors, gross total resection is the most powerful predictor of favorable outcomes in children if there are no unacceptable sequelae.[10] This may also be suitable for adult patients. In addition, considering the relatively poor outcome of PMA compared with that of PM, more radical or total excision techniques should be considered for survival advantage when managing this tumor. Based on our case, total resection of PMA can result in good postoperative outcome at the two year mark. Long-term follow-up is required for final evaluation of the treatment.

Author Contributions

Chi-Chih Hsieh and Hao-Kuang Wang conceived of the presented idea. Chi-Chih Hsieh wrote the manuscript with support from Tsung-Hsien Lin, Wan-Chin Lin, and Hao-Kuang Wang. Hao-Kuang Wang supervised the work. All authors discussed the results and contributed to the final manuscript.

Funding

This research received no external funding.

Institutional Review Board Statement

Not applicable.

Informed Consent Statement

Informed consent was obtained from all subjects involved in the study.

Data Availability Statement

Not applicable.

Conflicts of Interest

The authors declare no conflict of interest.

Table 1. Summary of spinal PMA cases reported in the literature

Reference	Age (years old)/ gender	Tumor region	Clinical presentation	Surgery	Radio-therapy/ chemo-therapy	Follow up	Recurrence/ progression	Follow up outcome
Mendiratta-Lala[2], 2007	29/Female	Cervico lumbosacral spine	Back pain and weakness of bilateral upper extremities	Partial resection	+/−	Nil	Unknown	Upper extremity weakness improved, but new progressive paraparesis developed after resection
Sajadi[3], 2008	45/Female	Cervical spine	Rapidly progressive cervical myelopathy	Biopsy	+/−	1 month	Yes	Died due to respiratory failure
Wu[4], 2013	40/Female	Thoracic lumbar junction	Burning pain and progressive numbness in both legs	Partial removal	+/−	3 years	No	Improvement in numbness and bladder dysfunction
Chaudhuri[5], 2014	35/Male	Thoracic spine	Paraparesis	Tumor excision	+/−	No	Unknown	No follow up
Dunn-Pirio[6], 2016	23/Female	Thoracic spine	Back pain, numbness in her lower extremities, gait difficulty	Subtotal excision	−/+ (Carbo-platin)	11 months	No	Returned to her neurological baseline
Mohamed[7], 2021	73/Male	Thoracic spine	Numbness and weakness in lower extremities, gait difficulty	Subtotal excision	+/−	2 years	No	No improvement and no progression of his symptoms
Our case	27/Female	Thoracic spine	Voiding dysfunction, recurrent urinary tract infection, bilateral lower limbs mild weakness	Total excision	−/−	2 years	No	Improvement in limb weakness and voiding dysfunction

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