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CASE REPORT

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Pilomyxoid astrocytoma of the thoracic spinal cord: Extremely rare case report of over 70-year-old patient

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Abstract

The Pilomyxoid is rare tumor in elderly population, in addition to the occurrence of an isolated lesion in spinal cord is extremely rare in non-pediatric population. Taking biopsy and subtotal resection is the starting point in essential approach for the treatment. After defining the histopathological nature of the tumor and specified that is Pilomyxoid, the next step is the combination of reoperation and adjuvant therapy.

KEYWORDS

case report, elderly, pilomyxoid astrocytoma, thoracic spinal cord

1 | INTRODUCTION

Pilomyxoid astrocytoma of the spinal cord in adults (PMA) is a rare neoplasm without definitive grade assignment in the revised WHO 2016 classification of tumors in the central nervous system. We report here a case of an adult patient with a spinal intramedullary T6-T8 PMA, its clinical manifestation and management outcomes. The MRI of a 73-year-old male patient with paraparesis showed an intramedullary mass at the thoracic spinal cord extending from T6 to T8. Partial surgical removal was performed and a biopsy was taken, followed by postoperative radiotherapy. Only a few adult cases of spinal PMA have been reported. Relevant papers were selected for the literature review. Our case shows that isolated thoracic PMA in spinal cord can occur in elderly age, even though PMA is a predominantly pediatric tumor where it has been reported for the first time 20 years ago. Because of its extremely rare occurrence, especially in this age group, there is a need for treatment guidelines that take into consideration the tumor features and occurrence in old age.

Pilomyxoid astrocytoma is a rare tumor in adults, and most certainly, it is extremely rare in the spinal cord. It was first reported in 1999 in pediatric group of patients.[1] It has been added under pilocytic astrocytoma PA subtype according to the WHO CNS tumor classification in 2007as grade II, with variant histological characteristics and more aggressive behavior.[2] PMA predominantly occurs in infants and young children and is most commonly located in the hypothalamic/chiasmatic region and shares other locations with pilocytic astrocytoma like the thalamus, temporal lobe, brain stem, and cerebellum.[3]

2 | CASE

A 73-year-old male patient with a two-month history of sudden onset numbness, gradual weakness in the lower extremities accompanied by walking difficulties. 10 days before admission his symptoms progressed to paraparesis.

Magnetic resonance imaging showed an intramedullary mass of 73x9x8mm at the thoracic spinal cord extending from T6 to T8. On T1 weighted image hypointense intramedullary lesion. T2-weighted image showed hyperintense lesion, signal intensity higher than cerebrospinal fluid.

The patient was transferred from a primary healthcare facility with a past medical history of controlled hypertension and DM II. History of chronic pelvic pain syndrome was reported. Any evidence of prostate cancer or other primary systemic tumor was excluded. Moreover, there was no evidence of any other spinal or intracranial lesion. CSF exam was negative for malignant cells.

He underwent only unilateral T7 Hemilaminectomy through posterior approach. Intraoperatively, the tumor appeared noncystic and gelatinous. More than third of the tumor was resected at the level of T7, and biopsy was taken for pathological analysis.

A histopathological laboratory disclosed that the tumor tissue consisted of bipolar cells with tendency of perivascular arrangement and embedded in myxoid matrix. Rosenthal fibers were not found. On immunohistochemistry, tumor cells were positive for S-100, GFAP, OLIG2, SOX10, and focally for Synaptophysin. Tumor cells were negative for panCK (AE1/AE3) and EMA. Mitoses were rare. Proliferative Ki-67 index ranged up to 4% (Figures 1 and 2).

After surgery, the patient had no improvement. He was referred to rehabilitation center and to receive external beam radiation therapy for 4 weeks. The patient had no improvement and no progression of his symptoms nor expansion of tumor mass in 2-year follow-ups after surgery.

3 | DISCUSSION

Pilomyxoid astrocytoma is a relatively rare tumor, it occurs in pediatric groups of patients commonly in the brain.[1] PMA originating in spinal cord is very rare, with over all about 10 reported cases of which 5 cases involving the thoracic region (Table 1), only one young adult case was reported with an

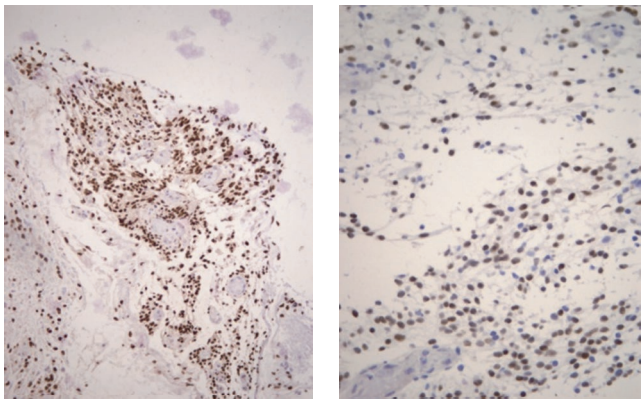

FIGURE 2 OLIG2 (left, ×20) and OSX10 (right, ×40) immunopositivity confirms glial origin and excludes the diagnosis of myxopapillary ependymoma

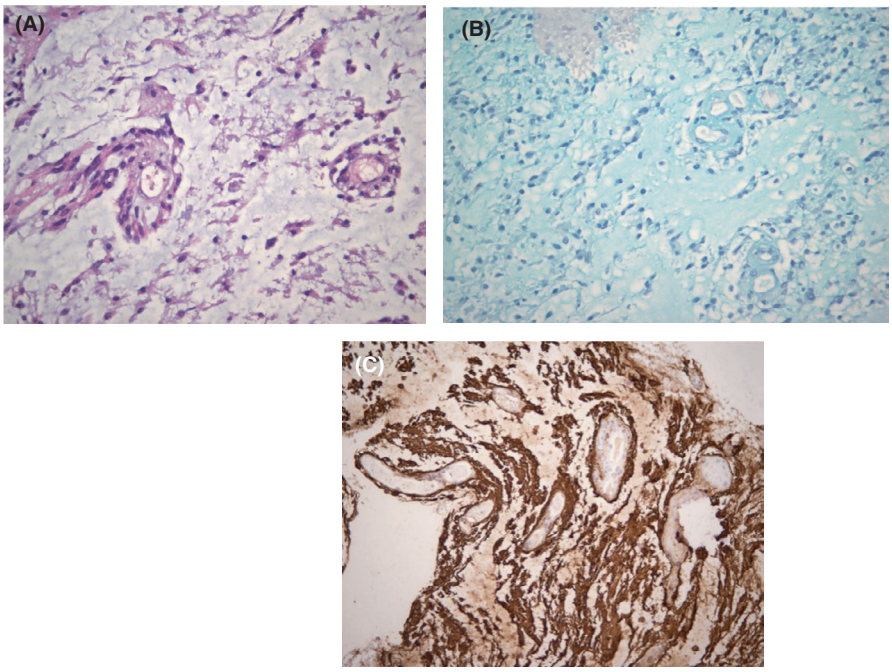

FIGURE 1 (A) Tumor cells show tendency for the perivascular arrangement (HE, ×400) (B) Myxoid matrix is visualized by Alcian blue histochemical stain (×400) (C). Immunohistochemistry for GFAP emphasizes perivascular arrangement of tumor cells (×200)

TABLE 1 Summary of spinal thoracic PMA cases reported in the literature

Age	Gender	Location	Surgery	Treatment	Outcome
73 Our case	Male	T6-T8	Subtotal removal	RT 4 weeks	No improvement
23	Female	T1-T2 NF1	Subtotal removal	Carboplatin 4 weeks no RT NF1	11 months then return to baseline deficit
29	Female	Mid cervical -lumbar	Subtotal removal	RT 4weeks	No follow-up was done or documented
35	Male	T10-T11	Total removal	RT 6 weeks	Under treatment at the time of publishing
40	Male	T11-L1	Subtotal removal	RT 4 Weeks	Gradual improvement of numbness, Bladder dysfunction,3-year follow-up no regrowth
45	Female	C1-C2	Surgical biopsy	RT 4 weeks	Tetraparesis then died. RDS

Abbreviations: NF1, Neurofibromatosis type 1; RDS, Respiratory Distress Syndrome; RT, Radiotherapy.

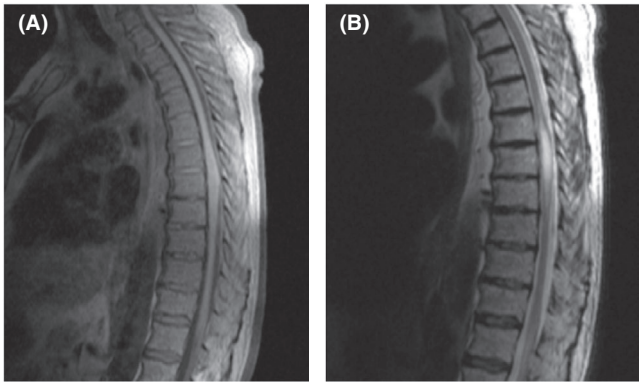

FIGURE 3 Thoracic intramedullary Pilomyxoid astrocytoma extending from T6 to T8 vertebral body (A) Sagittal T2w weighted (MRI) showed hyperintense lesion (B) Sagittal T1w weighted (MRI) showed hypointense lesion

isolated lesion in the thoracic spine without any evidence of other primary systemic or CNS neoplasm.[4-8]

Although radical surgical resection of the tumor showed more favorable survival outcomes within reported the pediatric cases[9]; radical surgical resection was only once reported (Table 1). Because PMA has showed more aggressive behavior and higher recurrence rate than PA,[3] 4–6 weeks of radiotherapy were recommended in all cases reported below in the table and restricted surgical resection was performed in most of reported cases. Because of aggressive nature of the tumor and to avoid radiation-induced myelopathy; we suggest a second surgery after the first subtotal resection in case of regrowth or expansion of tumor aiming further reduction of the tumor mass effect.

Our case appears to be the second case reported in the literature of isolated PMA located in the thoracic spine level with no brain involvement. MRI findings in our case compare to the only one isolated thoracic adult case reported has shown similar radiological features in both cases of the thoracic intramedullary PMA which can help for diagnosis. MRI contrast enhanced T2-weighted images showed hyperintense, nonhomogeneous, spindle shape, and intramedullary lesion. The T1-weighted images showed hypointense lesion (Figure 3).

Since PMA is a recently discovered tumor, reported cases in the literature suggesting difficulty distinguishing radiological imaging features for this extremely rare tumor compare to PA.[10] On the other hand, the histological finding of PMA has dependable difference features compared to PA.[3]

4 | CONCLUSION

Pilomyxoid astrocytoma as spinal cord tumor in adults especially over 70 years is extremely rare and lacks diagnostic and treatment guidelines.[11] Regardless of the recommendation of radiological oncological treatment after surgery that suggested on the cases reported in the literature, the radiological therapy duration and its advantage were not well reported.[3] Reoperation and more tumor resection of tumor mass in case of expansion could be taken in consideration in the process of incoming treatment guidelines.

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CONFLICTS OF INTEREST

I have read and agree to abide by the ScholarOne Conflict of Interest policy. I acknowledge that I must disclose any conflict of interest, along with a description of any personal business interest, affiliation, or activity with any entity, whether or not active in the neurosurgery field, which may give rise to a conflict of interest. I acknowledge that I understand that the neurosurgery Conflict of Interest policy extends to my partner, a business or other entity with which I am associated and all members of my immediate household. I also understand that it is my obligation to promptly reveal any changes in my personal business interests, affiliations or activities which may give rise to a conflict of interest, by updating my Disclosure Statement. On behalf

of myself and all other coauthor, we declare that we have no conflicts of interest.

AUTHOR CONTRIBUTIONS

Dr Abousabie A.Z and Dr Almzeogi M conceived of the presented idea. Dr Abousabie A.Z. developed the theory and drafted the manuscript. Dr Almzeogi M. verified the literature and designed the alignment. Dr Kotic J was coordinating between Oncology treatment and histopathological results, Dr Janicijevic A. coordinated and helped to draft the manuscript. Prof. Dr Tasic.G encouraged all the coauthors to investigate the case and supervised the findings of this work. All authors discussed the results and contributed to the final manuscript.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

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