

Case report of Pilomyxoid astrocytoma of the thoracic spinal cord: Literature review with presenting an extremely rare Case of over 70 year's old patient

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Abstract

Case summary: 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 preformed and a biopsy was taken, followed by postoperative radiotherapy.

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Literature review with presenting an extremely rare Case of over 70 year's old patient

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Background

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 managment outcomes.

Case summary: 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 preformed and a biopsy was taken, followed by postoperative radiotherapy.

Methods: Only a few adult cases of spinal PMA have been reported. Relevant papers were selected for the literature review.

Objective: Our case shows that thoracic PMA spinal cord tumors 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.

Conclusion: 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.

Keywords : Case report, pilomyxoid astrocytoma, thoracic spinal cord, elderly.

Introduction

PMA is a rare tumor in adults, and most certainly it is extremely rare in the spinal cord .It was first reported in a pediatric group of patients [1].It has been added under pilocytic astrocytoma PA subtype according to WHO CNS tumor classification in 2007 as grade II, with variant histological characteristics and a 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].

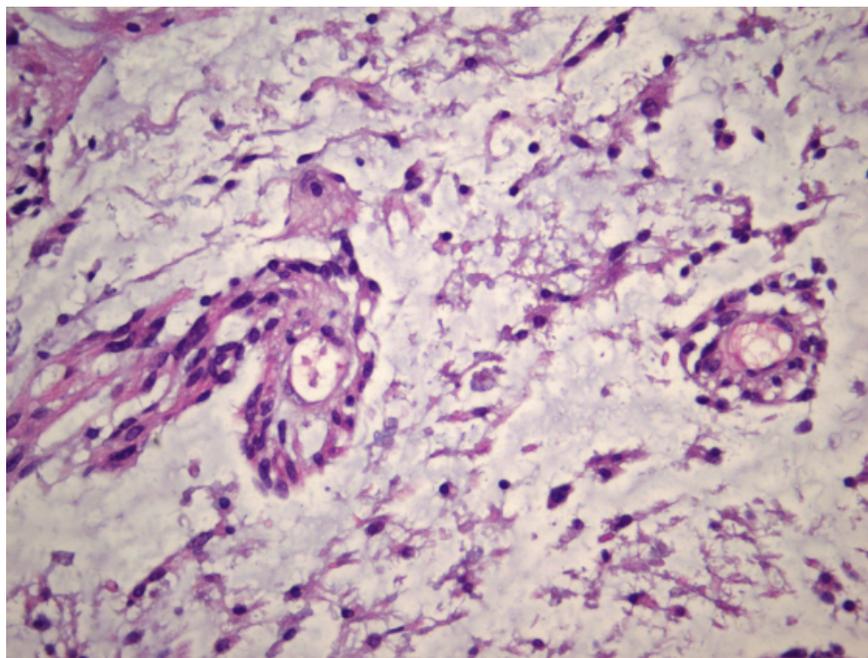
Case

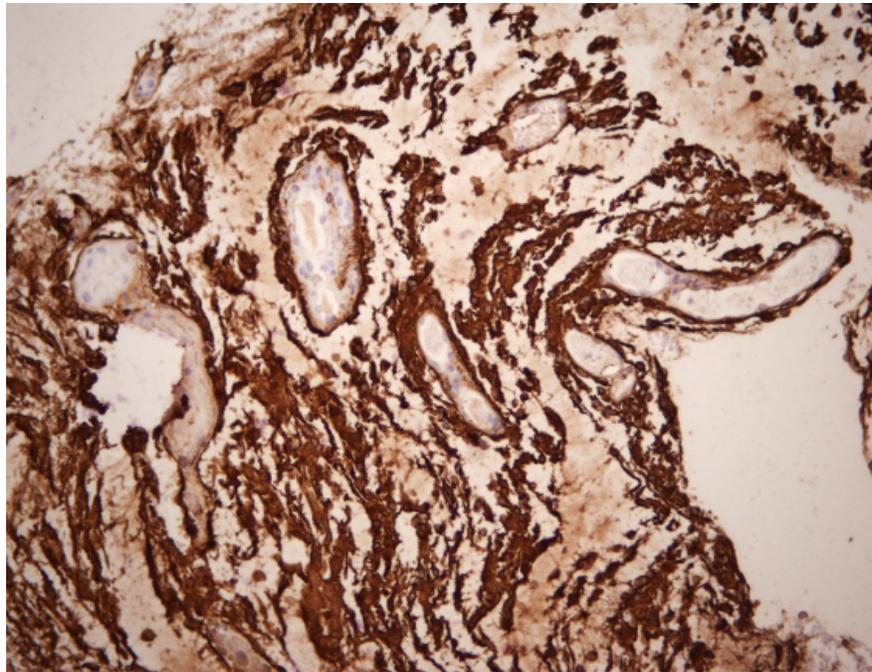
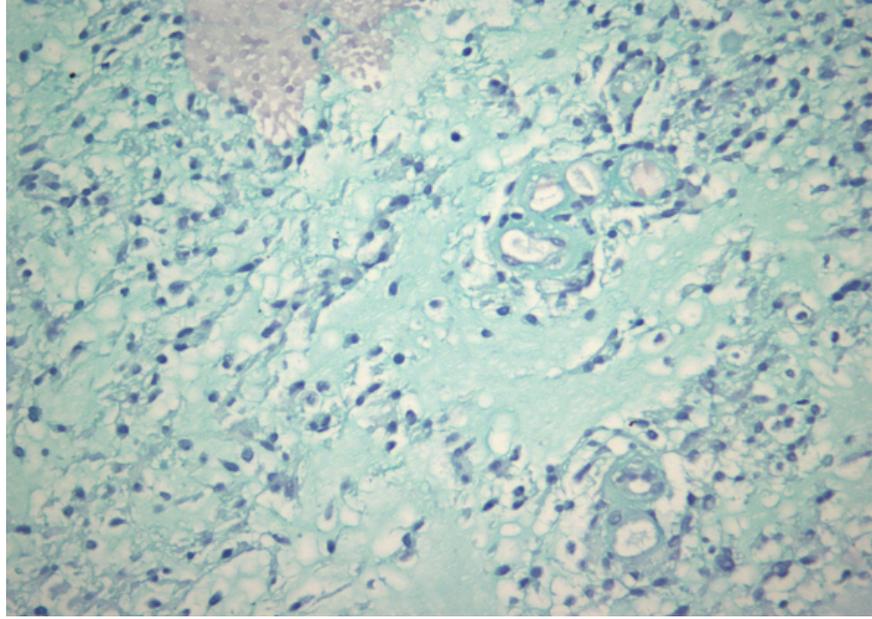
A 73-year-old male patient with a two month history of sudden onset numbness, gradual weakness in the lower extremities accompanied by walking difficulty was examined. 10 days before admission his symptoms progressed to paraparesis. Magnetic resonance imaging (MRI) showed an intramedullary mass of 73 x 9 x 8mm at the thoracic spinal cord extending from T6 to T8. On T1 weighted images hypointense intramedullary lesion. T2w weighted images 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 tumors were excluded. Moreover, there was no evidence of any other spinal or intracranial lesion. LP was negative for malignant cells.

He underwent unilateral T7 Hemilaminectomy through posterior approach. Intraoperatively the tumor appeared noncystic and gelatinous. Partial surgical resection was performed, as well as a biopsy was taken for pathological analysis. A histopathological laboratory disclosed that the tumour tissue consisted of bipolar cells with tendency of perivascular arrangement, embeded in myxoid matrix, Rosenthal fibers were not found. On immunohistochemistry, tumour cells were positive for S-100, GFAP, OLIG2, SOX10 and focally for Synaptophysin. Tumour cells were negativne for panCK (AE1/AE3) and EMA. Mitoses were rare. Proliferative Ki-67 index ranged up to 4%. [figure 1&2].

After surgery the patient had no improvement. He was refered to recieve external beam radiation therapy





- a)
- b)
- c)

Fig 1. (a) : Tumorous cells show tendency for the erivascular arrangement (HE, x400). (b) Myxoid matrix is visualised by Alcian blue histology.

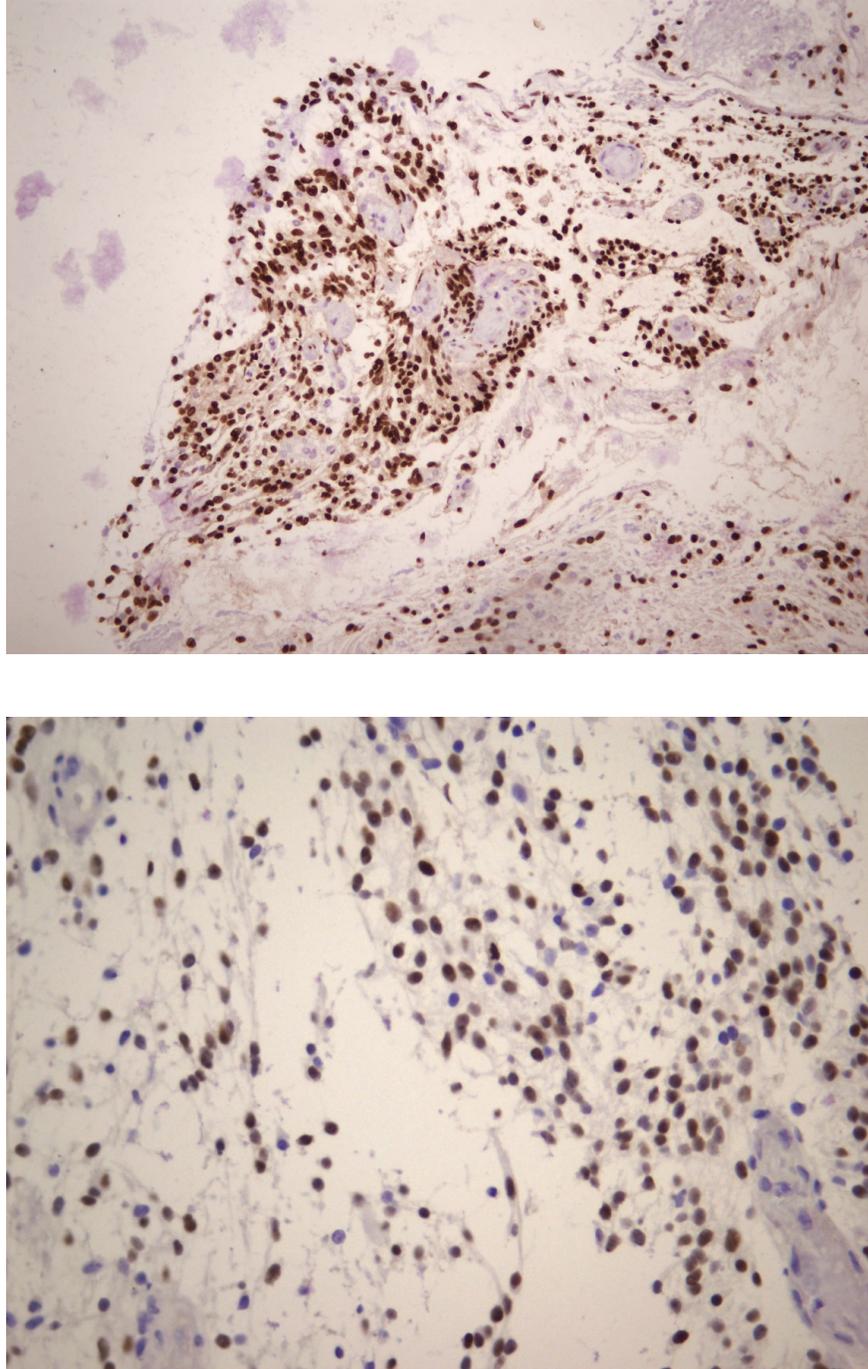


Fig 2. OLIG2 (left, x20) and OSX10 (right, x40) immunopositivity confirms glial origin and excludes the diagnosis of myxopapillary ependymoma.

Discussion

PMA 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 isolated lesion in the thoracic spine without any evidence of other primary systemic or CNS neoplasm [4,5,6,7,8].

PMA has showed more aggressive behaviour and higher recurrence rate than PA[3]. Radical surgical resection of the tumor showed more favorable survival outcomes within the pediatric population of patients. Restricted surgical resection in the case of spinal PMA was performed in most of reported cases [9].

We report, which what appears to be the second case, of isolated PMA located in the thoracic spine level. MRI findings in our case compare to the only one thoracic adult case reported, MRI has shown similar radiological features in both cases of the thoracic intramedullary PMA. Our lesion on MRI contrast enhanced T2 weighted images showed hyperintense, nonhomogeneous, spindle shape, intramedullary lesion. The T1 weighted images showed hypointense lesion [figure 3]. [5].





b)

Fig 2: Thoracic intramedullary PMA extending from T6 to T8 vertebral body. a) Sagittal T2w weighted (MRI) showed hyperintense le

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

Conclusion

PMA as a spinal cord tumor in adults, especially over 70 years, is extremely rare and lacks treatment guidelines [11]. The oncological treatment is recommended, and follow-ups are essential. However, a total surgical removal was only reported in a single case; in addition, the radiological therapy duration and its advantages were not well reported [3].

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Table 1

Age	Gender	Location	Surgery	Tretment	Outcome
73	Male	T6-T8	Subtotal removal	RT	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 tretment at the time of publishing
40	Male	T11-L1	Subtotal removal	RT 4 Weeks	Gradual improvement of numbness,Bladder dysfunction,3years follow up no regrowth

Age	Gender	Location	Surgery	Tretment	Outcome
45	Female	C1-C2	Surgical biopsy	RT 4weeks	Tetraparasis then died. RDS

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