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Case Report

Thoracolumbar pilomyxoid astrocytoma concomitant with spinal scoliosis: A case report and literature review

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ABSTRACT

Background: Pilomyxoid astrocytoma (PMA) is a variant of pilocytic astrocytomas but exhibits more aggressive behavior. Further, it is more prevalent in the hypothalamic/chiasmatic regions and is only rarely encountered in the thoracic spine.

Case Description: A 9-year-old male presented with severe spastic paraparesis (motor/sensory) attributed to a thoracic cord PMA and scoliosis. The magnetic resonance (MR) showed an intraaxial ill-defined expansile lesion with heterogeneous enhancement extending from the cervicothoracic junction to conus medullaris. A multilevel decompressive laminectomy was performed with restricted tumor debulking; an expansile duraplasty was also effected. Two years later, the patient has moderately improved and has not shown any symptom progression.

Conclusion: We recommend the early performance of a thoracic MR in children with idiopathic scoliosis presenting with the onset of a significant spastic paraparesis.

Keywords: Magnetic resonance imaging, Pilomyxoid astrocytoma, Radiotherapy, Scoliosis, Spinal cord, Thoracic

BACKGROUND

Intramedullary pilomyxoid astrocytoma (PMA) is a variant of pilocytic astrocytoma (PA).[2,5,6] That is categorized as the WHO Grade 2.[6] However, PMA is more aggressive than PA and is usually located within the spinal cord of younger patients.[9] The treatment options for PMA include surgical resection which is often limited to debulking (e.g., extent restricted to minimize neurological deficit) followed by radiotherapy with or without chemotherapy.[5,10] Here, we are presenting a 9-year-old with a thoracic PMA and scoliosis who successfully underwent laminectomy with partial tumor debulking/resection plus duraplasty and exhibited an adequate 2-year postoperative neurological outcome.

CASE PRESENTATION

A 9-year-old male with spinal scoliosis recently diagnosed/brace (e.g., 1 month's duration) presented with a severe motor/sensory spastic paraparesis. The holospinal magnetic resonance (MR) revealed an intraaxial/intramedullary ill-defined expansile lesion with heterogeneous enhancement extending from the cervicothoracic junction to conus medullaris [Figure 1].

Table 1: All reported cases of pilomyxoid astrocytoma of thoracic spine in the literature.

Author/year of publication	Age/sex	Imaging features	Surgical treatment	Adjuvant therapy	Follow-up	Scoliosis
Komotar *et al.*[6]/2005	3 w*/male	An heterogeneous T2 hyperintensity of entire cord with meningeal enhancement and fourth ventricle and medulla involvement	Biopsy and decompressive laminectomy	No	3.5 years with disability	No
Komotar *et al.*[6]/2005	8 y†/male	An enhancing lesion at T9-T12 with syrinx above it	GTR‡	No	9 months with recurrence	No
Komotar *et al.*[6]/2005	6 y/male	An enhancing lesion at T9-T11 with extension of abnormal heterogeneous signal in whole thoracic cord	STR‴ and then decompressive laminectomy	Radiotherapy and Chemotherapy	5 years with some residue	Yes
Mendiratta-Lala *et al.*[7]/2007	29 y/female	An enhancing intradural extramedullary lesion in cervical-thoracic-lumbosacral region with associated syrinx	STR	Radiotherapy	N/A	No
Garber *et al.*[5]/2013	11 y/male	An enhancing T5-T10 cord lesion with associated syrinx	NTR¶	Radiotherapy and chemotherapy	20 months with multiple recurrences	Yes
Wu *et al.*[11]/2013	40 y/female	A T11-L1 mass with cord swelling and heterogeneous enhancement	STR	Radiotherapy	3 years with no recurrence	No
Chaudhuri *et al.*[3]/2014	35 y/Male	An intramedullary T10-T11 lesion with extensive enhancement	Laminectomy and tumor excision	Radiotherapy	N/A€	No
Dunn-Pirio *et al.*[4]/2016	23 y/female	An enhancing T1-T12 lesion with cystic change	STR	Chemotherapy	6 months with partial response	Yes
Aghajan *et al.*[1]/2016	3 m£/female	An extensive nodular enhancement of cervicomedullary, cervical, and thoracic cord with diffuse leptomeningeal disease with concomitant intracranial involvement	Biopsy of Intracranial lesion	Chemotherapy	Died at 20 months old	No
Tabibkhooei *et al.*/2019	9 y/male	An intraaxial ill-defined expansile lesion with heterogeneous enhancement from cervicothoracic junction to conus medullaris	STR	Radiotherapy	2 years with no recurrence	Yes

*Week-old, †Year-old, ‡Gross total resection, ‴Subtotal resection, ¶Near total resection, £Month-old, €Not available. GTR: Gross total resection, STR: Subtotal resection, NTR: Near total resection

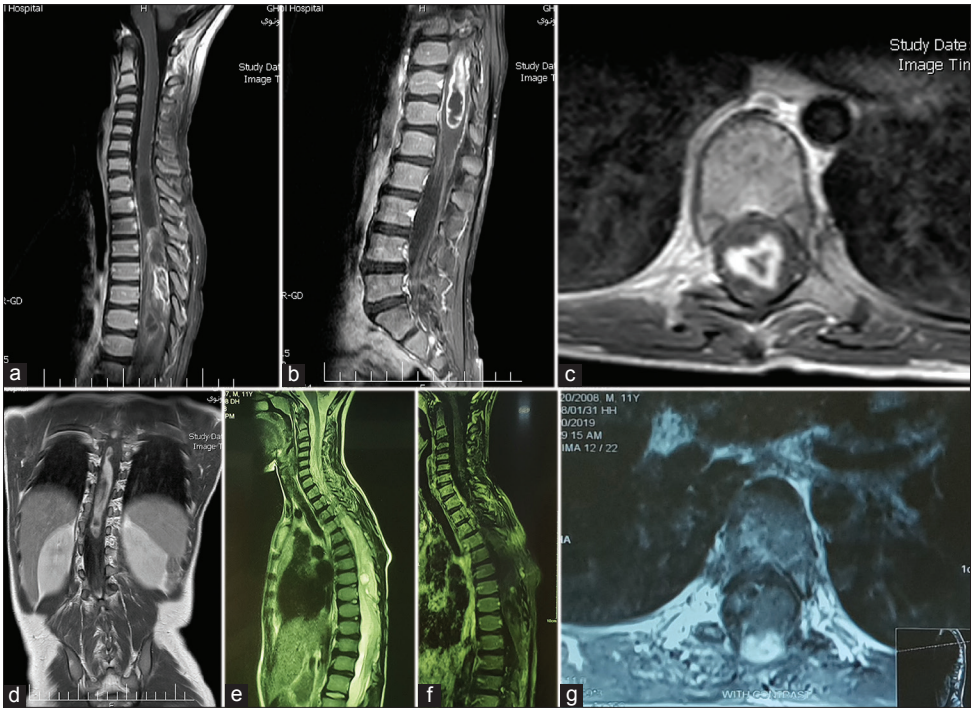

Figure 1: Preoperative magnetic resonance imaging (MRI) with gadolinium showed an expansile intramedullary tumor of thoracic cord and conus medullaris with irregular heterogeneous enhancement and some cystic changes on sagittal (a and b) and axial (c) views in a 9-year-old patient with severe thoracolumbar scoliosis (d). After surgical treatment, 2 years follow-up MRI reveals an evidence of minimal cord herniation toward laminectomy sites and intramedullary cystic changes (sagittal T2 sequence), (e) with minimal enhancement on sagittal (f) and axial (g) views.

He underwent a T5-T11 laminectomy with tumor debulking (e.g., poor plane between tumor/cord) followed by expansile duraplasty. Postoperatively, he exhibited significant improvement of the paraparesis. As the histopathological exam showed a PMA, adjuvant therapy included spinal radiation [Figure 2]. Two years later, the patient improvement has continued and the follow-up MR showed no further progression [Figure 1].

DISCUSSION

PMA is the more aggressive type of astrocytoma that usually seen in child <4-year-old age and in hypothalamic/chiasmatic region.[8,9] PMA in spinal cord region is rare in the thoracic cord; this case is the tenth reported in the literature [Table 1].

On imaging, we have no any distinct criteria for differentiation of PA from PMA.[6,7] However, histological findings include; a mucopolysaccharide matrix with monomorphic piloid cells, the absence of Rosenthal fibers, biphasic architecture, and eosinophilic granular bodies.[5,6]

Usually, in cases of intramedullary astrocytoma, we recommend only debulking the center of tumor (e.g., not beyond the ill-defined border between tumor/normal tissue), and then performing an expansile duraplasty, if indicated.

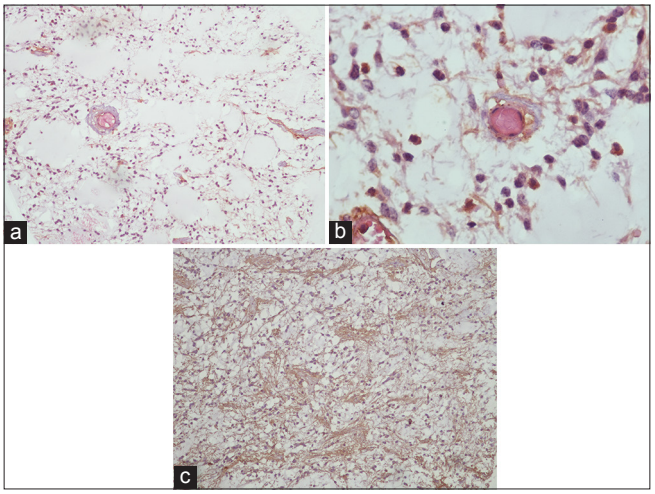

Figure 2: This shows histopathology of resected tumor. We could see hypocellular glial tumor, in which monomorphic piloid tumor cells are dispersed in a myxoid matrix (Hematoxylin and Eosin stain, ×100) (a). Furthermore, tendency of piloid perivascular cells to form a rosette-like structure without any Rosenthal fiber could be seen (Hematoxylin and Eosin stain, ×400, b). (c) Reveals a strong immunoreactivity of the tumor cells for glial fibrillary acidic protein (GFAP immunostaining, ×100, c).

The addition of adjuvant radiation therapy is controversial. Many clinicians recommend safe surgical resection of the

tumor followed by both radiotherapy and chemotherapy to improve survival.[5,10] In this case, the patient did receive radiation.

CONCLUSION

We present the rare case of a 9-year-old with a PMA of the thoracic cord and scoliosis who was effectively treated with a T5-T11 laminectomy, tumor debulking, duraplasty, and later adjuvant radiation therapy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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