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*Brief Report of Special Case*

# Pilomyxoid astrocytoma of the spinal cord in an adult

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## Summary

We report a 45-year old woman with a pilomyxoid astrocytoma (PMA) of the cervical spinal cord with a rapid clinical course and fatal outcome. Moreover, two family members of the patient were reported to have brainstem tumours with similar histopathological features. This may be the first report of familial PMAs.

*Keywords*: Pilomyxoid astrocytoma; spinal cord.

We report a 45 year old woman with a pilomyxoid astrocytoma (PMA) of the cervical spinal cord with a rapid clinical course and fatal outcome. The patient had a family history of brainstem pilocytic astrocytoma (PA) occurring in her father and brother. She presented with a

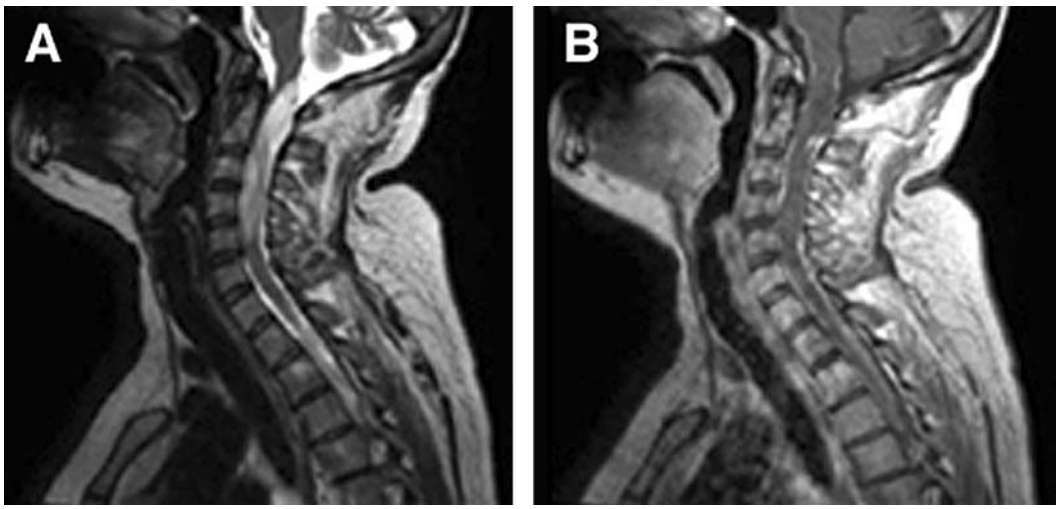


Fig. 1. (A) Cervical $T_2$-weighted MRI sequences showing an intramedullary hyperintense lesion extending from the foramen magnum to the C5 level. (B) Post-gadolinium $T_1$-weighted MRI sequences showing partial enhancement of the lesion in the posterior cord at C2 level and extensive pial enhancement in the spinal cord and the posterior fossa. (C) Vimentin staining of the patient's intramedullary tumour (100× magnification). Histopathology revealed a compact vimentin-positive tumour having a rich myxoid background, with uniform bipolar cells. The absence of Rosenthal fibers and eosinophilic granular bodies was consistent with the diagnosis of PMA. (D) Haematoxylin and eosin staining of the brainstem tumour of the patient's brother (400× magnification). Histopathology was substantially identical to (C), suggesting the diagnosis of PMA

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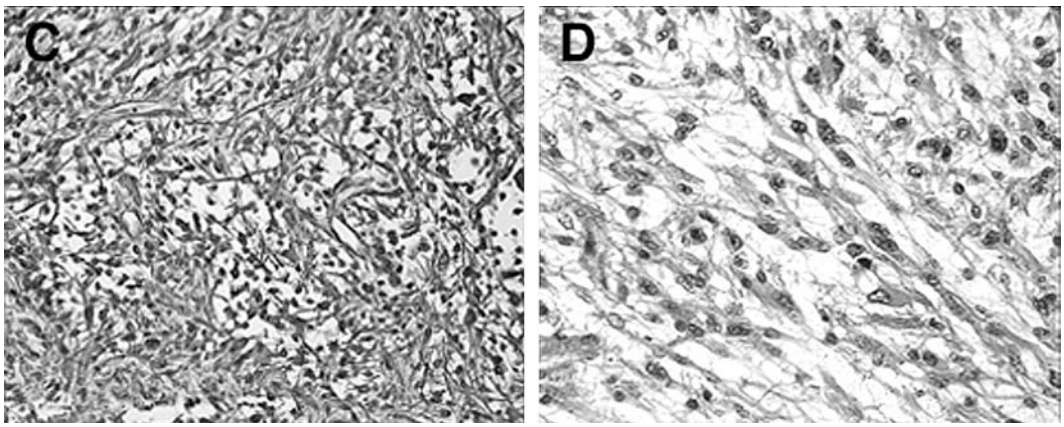


Fig. 1 (*continued*)

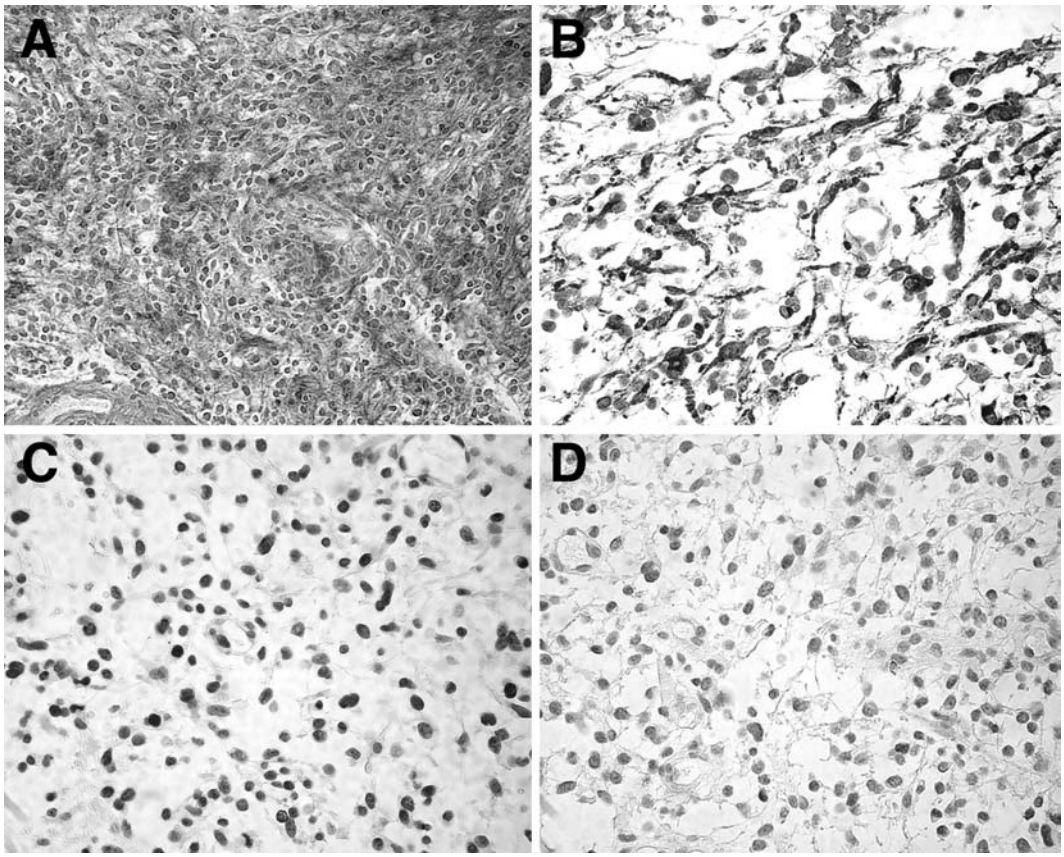


Fig. 2. (A) Alcian-Blue-staining (100× magnification). Compact tumour with high cell density composed of mostly spindle shaped cells on a fibrillary and myxoid background. (B) GFAP immunohistochemistry (400× magnification). The majority of tumour cells express GFAP showing uni- or bipolar processes. (C) P53 immunohistochemistry (400× magnification). The great majority of tumour cells are negative for p53, only less than 1% of the cells express p53. (D) EGFR immunohistochemistry (400× magnification). No expression of EGFR is seen in the tumour cells

rapidly progressive cervical myelopathy over 3 months. A MRI showed a diffuse cervical intramedullary tumour (Fig. 1A, B) with spinal and intracranial pial seeding. The patient underwent C1–C2 laminectomy and tumour biopsy. Histopathology was consistent with the diagnosis of PMA (Figs. 1C and 2).

Despite corticosteroid and radiation therapy, the patient developed a rapidly progressive tetraparesis and bulbar symptoms, and eventually died due to respiratory failure one month after her admission.

PMAs occur more frequently in the hypothalamic-chiasmatic region in children and are known to have

a worse overall outcome as compared to PAs [4]. Occurrence of PMAs in adults represents an exceptional event [1, 2] with only one example of an adult spinal cord lesion observed to date [3]. The rapidly fatal progression in our patient illustrates the aggressive nature of PMAs, despite their prior classification among PAs. Moreover, a review of the tissue sections of the originally described brainstem PAs in two family members of the patient revealed histopathological features compatible with a PMA (Fig. 1D). This may be the first report of familial PMAs, suggesting a genetic predisposition for these tumours.

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