Primary pilomyxoid astrocytoma of the thoracolumbar spinal cord in an adult

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Full Text

Sir,

Pilomyxoid astrocytoma (PMA) is a rare tumor that was previously classified as pilocytic astrocytoma (PA) because of similar histological features. [1] Compared with PA, PMA demonstrates more aggressive biological behavior, and was assigned to World Health Organization (WHO) grade II according to the 2007 WHO Classification. [2] PMA in the spinal cord is extremely rare. We documented probably the first case of primary thoracolumbar spinal PMA.

A 40-year-old female presented with intermittent burning pain and progressive numbness in both legs of 8 months duration. On admission, mild incomplete voiding was noted. Neurological examination revealed reduced superficial sensation below T12 level. Preoperative magnetic resonance imaging (MRI) of the spine demonstrated an abnormal intramedullary lesion with cord swelling at the T11-L1 level. The lesion showed heterogeneous isointensity on the T1-weighted image (WI) with ill-defined margins from the normal spinal cord, mild hyperintensity on the T2WI, and heterogeneous speckled enhancement after gadolinium administration [Figure 1]. A T11-L1 laminectomy was performed through the posterior approach. Due to its adhesion to the cord and ill-defined margins with the conus medullaris, the intramedullary tumor was subtotally removed. Histopathologically, the tumor was composed of monomorphic bipolar piloid cells with elongated cytoplasmic processes in a prominent myxoid background without Rosenthal fibers [Figure 2]a and b. The tumor cells were arranged around the small blood vessels, showing an angiocentric pattern [Figure 2]c. Immunohistochemical staining revealed immunoreactivity for glial fibrillary acidic protein, and the MIB-1 labeling index was 3%. These findings were consistent with the diagnosis of PMA. After surgery she had relief of pain. Postoperatively she received external beam radiotherapy (40 Gy in 25 fractions, 1 fraction per day, 5 fractions per week). She had gradual improvement in numbness and bladder dysfunction in the follow-up. At 3 years follow-up MRI revealed no regrowth, or metastasis of the residual tumor was found [Figure 3].
PMAs may originate from anywhere along the neuroaxis but the most frequent locations are the hypothalamic and chiasmatic regions. [2] Spinal PMAs of spinal cord are extremely rare, especially in adult patients. Since the first description in 2005 by Komotar et al., [3] only nine cases have been reported in the literature. [1],[2],[3],[4],[5],[6] Spinal PMAs affect pediatric age group more than adult patients with a female preponderance, mostly the cervical and thoracic spinal regions. Most of the lesions were intramedullary, and only one case was intradural extramedullary lesion. [5] The MRI features of spinal PMA are variable and could be misleading. Pathological examination is the gold standard in the differential diagnosis of PMA. [1] [2] [3] [4] [5]

Because of the ill-defined margins or dense adhesions to the neural tissue, partial removal or even biopsy had to be performed to avoid unacceptable postoperative complications for the treatment of spinal PMA. The main goal of surgery is preservation of neurological function. Adjuvant therapy is recommended considering the high risk of local recurrence and cerebrospinal fluid dissemination, [5] particularly when total removal cannot be achieved. However, the efficacy of adjuvant radiation therapy for controlling the tumor is still uncertain, and cisplatin/carboplatin-based multidrug chemotherapy is recommended for the initial treatment of PMAs. [6] Cisplatin + etoposide was shown to be effective in a 15-month-old girl and the tumor has not relapsed for 64 months. [4]

Paraskevopoulos et al. reported a female child who had early recurrence and transformation within 3 months into a glioblastoma with a fatal outcome in spite of adjunctive chemotherapy. [7] This case suggests that the biological behavior of PMA may be variable. Because of possible variable biologic behavior of PMA, adjuvant therapy may be recommended at an early stage of diagnosis, particularly for the tumor with more aggressive findings, such as high expression of Ki-67, rapid progression, and malignant transformation, although overall prognosis is uncertain.

In conclusion, PMAs should be included into the differential diagnosis of intradural tumors of the spine. Multidisciplinary treatment consisting of surgical removal and adjuvant radiochemotherapy should be considered.

References