Pilomyxoid astrocytoma

Pilomyxoid astrocytomas (PMAs) were first officially described in 2007. Since then, intermediate pilomyxoid tumors with histopathological features typical of both PMAs and pilocytic astrocytomas (PAs) have been described.

Previous studies have shown PMA to behave more aggressively than PA, with shorter progression-free and overall survival as well as a higher rate of recurrence and CNS dissemination. These findings suggest that PMA may be a unique and distinct neoplasm.

A review summarizes the histologic, clinical, and radiographic characteristics of PMA. In addition, the current treatment options and research endeavors involving this disease are described. Increased recognition of PMA within the medical community has the potential to affect the treatment and prognosis of pediatric low-grade astrocytomas [1].

The grading of pilomyxoid astrocytoma in the World Health Organization Classification of Tumors of the Central Nervous System 2016 has also changed. While previously designated as WHO grade II, recent studies have shown extensive histological and genetic overlap between pilomyxoid and pilocytic astrocytomas, with some of the former maturing into the latter over time and less certainty that the pilomyxoid variant always follows a more aggressive course than a more classic appearing suprasellar pilocytic astrocytoma. For these reasons, it is not clear that pilomyxoid astrocytoma should automatically be assigned to WHO grade II and the suggestion was made to suppress grading of pilomyxoid astrocytomas until further studies clarify their behavior.

Case series

Ma et al. retrospectively analyzed four cases involving a 16-year-old adolescent with a 3-cm recurring suprasellar tumor, an 11-year-old boy with a nonrecurring 3-cm mass in the left cerebellum, an 18-year-old adolescent with a mass in the suprasellar cistern who died 2 days after total tumor resection, and a 26-year-old woman with a nonrecurring 2-cm mass in the right temporal lobe. Microscopically, the tumors were a monomorphous population of small bipolar cells in a prominent myxoid/mucoid background with rare Rosenthal fibers. The tumor cells infiltrated the adjacent brain parenchyma. Findings for glial fibrillary acidic protein and oligodendrocyte transcription factor were positive, and the Ki-67 protein proliferation index was about 2 %. Our findings document the existence of tumors that are histologically like PMAs but also have Rosenthal fibers. Studies of more such cases are needed for clarification of such tumors' clinical features [2].

Case reports

A 15-month-old girl presented with a spinal pilomyxoid astrocytoma manifesting as a 3-month history of dysphagia. Magnetic resonance imaging showed an intramedullary mass of the cervical spinal cord at C1-C6 with syringobulbia. She underwent partial removal of the tumor and received postoperative chemotherapy with cisplatin and etoposide. The tumor completely responded to the treatment and has not relapsed for 64 months. Pilomyxoid astrocytoma frequently occurs in the opticohypothalamic regions but is rare in the spine. The present case suggests that surgery followed by chemotherapy with cisplatin and etoposide may be an effective therapeutic option for pilomyxoid astrocytoma of the cervical spinal cord [3].

1)
http://www.ncbi.nlm.nih.gov/pmc/articles/PMC1480592/
2)
Ma X, Wang Y, Liu H, Yu H, He J. Pilomyxoid astrocytomas with rare rosenthal fibers. Brain Tumor Pathol. 2015 Dec 15. [Epub ahead of print] PubMed PMID: 26670169.
3)
Matsuzaki K, Kageji T, Watanabe H, Hirose T, Nagahiro S. Pilomyxoid astrocytoma of the cervical spinal cord successfully treated with chemotherapy: case report. Neurol Med Chir (Tokyo). 2010;50(10):939-42. PubMed PMID: 21030812.