Abstracts

HG-92. IMAGING CHARACTERISTICS OF PEDIATRIC DIFFUSE MIDLINE GLIOMAS BASED ON THE PRESENCE OF A POOR PROGNOSTIC MARKER HISTONE H3 K27M MUTATION

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The molecular basis underlying pediatric gliomas is distinct from adult gliomas. One unique molecular alteration that has been identified in pediatric gliomas is K27M missense mutation in histone H3 variants and presence of this mutation correlates with poor prognosis. The forthcoming 2016 WHO Classification will include “diffuse midline gliomas with histone H3 K27M mutation” as a new diagnostic entity. We identify imaging

characteristics of these diffuse midline gliomas in 43 pediatric patients based on the presence of histone H3 K27M mutation. We evaluated the imaging patterns of these diffuse gliomas based on their location, enhancement pattern, and necrosis. In these 43 patients, 74.4% of the tumors were midline with 75% of these had K27M mutation. All of the tumors arising supratentorially in the cerebral hemispheres had wild type histone H3 and were associated with a high amount of necrosis ($p = 0.043$). Both of the two cervical spine tumors were K27M positive and demonstrated distal subependymal metastases within the lateral ventricles. When comparing histone H3 mutant to wild type midline diffuse gliomas, K27M gliomas were more likely to result in mass effect ($p = 0.01$) and more likely to extend to the border of the fourth ventricle ($p = 0.038$). The majority of diffuse midline gliomas originating in the thalamus, pons, or spinal cord were found to harbor histone H3 K27M mutation. Tumors arising in the cervical cord showed propensity for distal metastatic spread. In contrast, diffuse gliomas arising in the cerebral hemispheres were uniformly negative for K27M mutation and were more likely to demonstrate necrosis.