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Leptomeningeal glioneuronal tumor in a 6 year old XYY patient presenting with rapid growth velocity: A case report

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Background: Diffuse Leptomeningeal Glioneuronal Tumors (DLGNTs) are relatively novel pediatric tumor entities presenting with neurological symptoms of headache, cranial nerve palsies and motor disturbances. This tumor has been associated with BRAF mutations & chromosomes 5p/19q deletions. Here we present the case of a XYY genotype patient presenting with rapid growth velocity which is unique.

Case Presentation: A 6 year 4 month old Caucasian male was evaluated for a rapid growth velocity of 10.6 cm/year since the age of 2 years having traveled from the 25th to the 98th percentile for height at the time of admission to our hospital (Figure). He had been evaluated for Marfan's syndrome, spinal stiffness, and scoliosis and was referred to endocrinology for overgrowth. Patient was admitted to our hospital with sudden onset headaches & severe constipation. Shortly after admission, he developed projectile vomiting, lethargy & mental status changes which warranted a stat MRI of his brain. A suprasellar enhancing mass lesion involving the pituitary stalk & hypothalamus was seen with nodular lesions along the third and fourth ventricles, brain stem and upper cervical spinal cord. An MRI of the spine showed extensive leptomeningeal enhancement with drop metastases in the distal thecal sac & at T1-T2 level. Biopsy of the tumor revealed leptomeningeal thickening with cortical infiltration by monomorphic oligodendroglia-like tumor cells. Cells had uniform, round nuclei & perinuclear halos in a desmoplastic stroma. Immunohistochemistry demonstrated a mixed glial & neuronal immunophenotype. iFISH demonstrated a duplication of BRAF(7q34), but no loss of chromosome 1p or 19q. Chromosomal microarray analysis demonstrated a 47 XYY karyotype. He was started on vincristine & carboplatin and overtime his growth velocity has decreased to 7.2 cm/year. This is the first case of the tumor associated with an XYY genome & presenting with rapid growth velocity.

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