Primary Melanocytic Suprasellar Tumor Presenting with Growth Hormone Deficiency

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A 13-year-old male patient presented with the complaint of short stature. He had a history of headache for 6 months, recently accompanied by vomiting in the morning. On admission, his height standard deviation score (SDS) was -2.5, he was prepubertal and his bone age was 10 years. Ocular examination did not reveal papiledema or any defect in the field of vision and visual acuity. Complete blood count and routine biochemical tests were within the normal limits. Thyroid hormones, prolactin, adrenocorticotropic hormone and cortisol levels were all within the normal range. Serum insulin-like growth factor-1 (IGF-1) level was < -3 SD and IGF binding protein-3 (IGFBP-3) level was between -1 and -2 SD. Due to headache and morning vomiting accompanying short stature, cranial and pituitary MRI was performed. It showed a suprasellar lobulated mass of 2.2x2.1x2 cm occupying the postchiasmatic area, reaching to the floor of the third ventricle, infiltrating the posterior part of the optic chiasma, left optic tractus and left hypocampus. Growth hormone (GH) stimulation tests were performed. Peak GH response values to L-dopa and clonidine stimulation test were 4.5 ng/mL and 0.6 ng/mL, respectively. The levels of other pituitary hormones were within normal range. The result of water deprivation test was normal and central diabetes insipidus was ruled out. Because of the infiltration of the tumor, it could not be removed and only biopsy was performed. GH treatment was decided not to be started until ascertaining the stability of the tumor. The pathological evaluation revealed melanocytoma. The patient did not develop any neurological deficits after the biopsy, but developed hypothyroidism and diabetes insipidus, thus thyroid hormone and desmopressin replacements were started. Radiotherapy was given because of the risk of transformation to malignant forms and risk of enlargement of the tumor. After it was shown that the tumor was stable, the GH treatment was commenced and the height velocity during the first and second year of treatment was 13.5 cm and 13 cm, respectively. The GH replacement was stopped after 2 and a half years of treatment when the patient's height was 167 cm (-1.0 SDS). His final height reached the target height which was 168 cm (-1.0 SDS).

Primary melanocytic tumors of the central nervous system arising from the melanocytes of the leptomeninges (i.e. melanotic meningioma, melanotic schwannoma, malignant melanoma, melanotic neuroectodermal tumor and meningeal melanocytoma) are extremely rare. Melanocytomas are even rarer in childhood period with only twelve cases described in the literature. These twelve cases were located in different places in the central nervous system but none in the suprasellar/sellar region. Primary sellar melanocytic tumors are also exceedingly rare and 8 melanomas and 2 melanocytomas have been described so far, but none of these were in the childhood period. Our patient is the first case reported in the literature with suprasellar localization of primary melanocytic tumor presenting with GH deficiency in childhood. This case has been treated with GH for two and a half years without any complication related to GH treatment and there was no change in the size of the tumor.