A Case of Gonadotropin-independent precocious puberty due to germ cell tumor in the frontal lobe

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Introduction

- It is known that gonadotropin-independent or peripheral precocious puberty (PPP) may develop due to tumors that secrete beta human chorionic gonadotropin (Beta-HCG).
- These tumors can be located in gonads, liver, mediastinum or central nervous system. HCG-producing tumors of the central nervous system are rare, most commonly seen in the suprasellar and pineal regions. However, any germ cell tumor (GCT) containing the syncytiotroblastic giant cell may produce HCG and potentially lead to PPP.
- Here, we present a male patient who was diagnosed as PPP due to GCT in the unusual location of central nervous system.

Case report

- A 9-year-old male patient was admitted to another hospital with headache, vomiting and double vision for 6 months ago. He was operated with a 7x6x5 cm mass in his right frontal region.
- The patient underwent total excision and was diagnosed as mixed GCT on histopathological examination and admitted to our hospital for further treatment. He was consulted to the pediatric endocrinology upon detection of macrogenitalia. At the time of admission his age was 9 years and 8 months.
- The weight of the patient was 45 kg (+ 1.8 SDS) and 145.5 cm (+1.65 SDS). On physical examination, testicular volume was 8/8 ml, Tanner stage 4 pubic hair was observed and macrogenitalia was present (Figure 1). In laboratory evaluations, LH was 0.02 mIU/ml and FSH was <0.05 mIU/ml and total testosterone value was 839 ng/dl. Beta-HCG value in the serum and cerebrospinal fluid (CSF) were 106 IU/L (N: <5), 631 IU/L respectively.
- The bone age was 13 years and 6 months. Craniospinal magnetic resonance imaging (MRI) showed residual frontal mass without spinal seeding metastasis. He received chemotherapy and craniospinal irradiation. CSF and serum beta-HCG levels were normal after 3 cycles of chemotherapy.
- After accomplished of the treatment his LH 1.48 mIU/ml and FSH was 2.25 mIU/ml and total testosterone value was 25 ng/dl. We decided to initiate a gonadotropin releasing hormone analogue due to the fact that the bone age was too advanced.

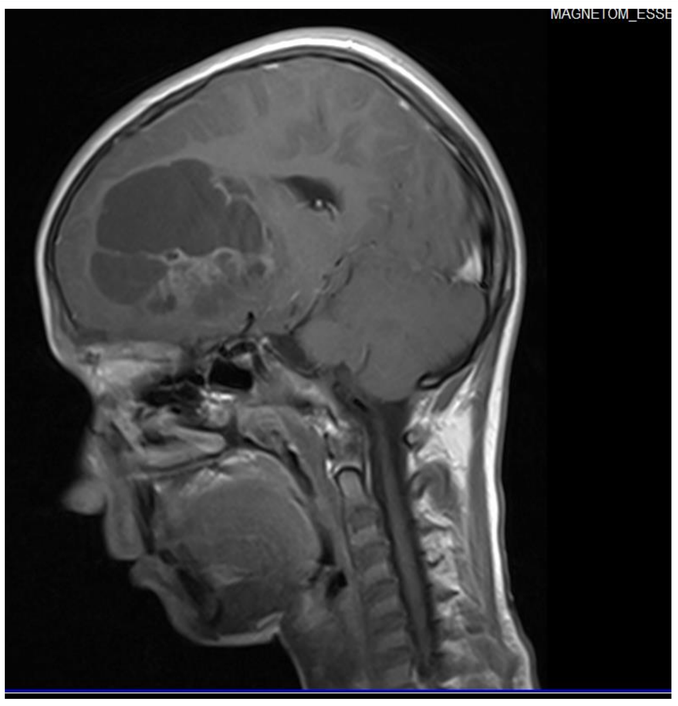

Figure 1; Preoperative radiological image of the mass in frontal region

Conclusion

- Intracranial GCTs are rarely seen as a cause of PPP.
- Gonadotropin-independent precocious puberty due to GCT located in the frontal lobe in the brain has not been previously reported in the literature.
- In these patients, the bone age advance rapidly and after the oncological treatment and normalization of beta-HCG values, the hypothalamus-pituitary-gonad axis can be activated. Therefore, close follow-up is required.