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Central nervous system germ cell tumours (GCTs) are rare, accounting for 0.1-3.4% of all primary brain malignancies. Intracranial GCTs (ICGCTs) usually arise within the pineal recess (50-65%), sellar-suprasellar region (25-35%) and rarely in the basal ganglia-thalamus (5-10%). A peakonset occurs during the second decade of life with a median age at diagnosis between10-12 years and a male predominance. Histologically, ICGCTs are classified into germinomas accounting for two thirds of cases and non-germinomatous GCTs. ICGCTs present with clinical features dependent on their location and tumour size. Symptoms most often arise from compressed optic structures, increased intracranial pressure and central endocrine abnormalities, particularly diabetes insipidus (DI). Clinical findings combined with elevated tumour marker levels within the serum and/or cerebrospinal fluid (CSF) can be diagnostic.
Case: A 30-year-old lady who survived a childhood leukaemia presented as an emergency with a two-month history of headaches, dizziness and near-complete visual loss. Cranial MRI identified a large sellar mass with suprasellar extension compressing the optic chiasma and hypothalamus. Biochemical and endocrine profiles confirmed diabetes insipidus, hypocortisolaemia and hypothyroidism all requiring hormone replacement. Visual field examination revealed total left visual loss and a right temporal hemianopia. Cerebrospinal fluid analysis identified an elevated human chorionic gonadotropin (HCG) level at 16 IU/L (normal <2), with a normal alpha-fetoprotein (AFP) level. Both tumour markers were normal in the serum. Emergency Etoposide-Cisplatin (Em-EP) chemotherapy was initiated for a suprasellar GCT followed by the EPOMB-ITMTX regimen [Etoposide-Cisplatinwith intrathecal Methotrexate (ITMTX) alternating with Vincristine-Methotrexate-Bleomycin]. After four cycles EPOMB-ITMTX, the vision was fully restored, brain MRI demonstrated an excellent radiological response and the CSF HCG normalised. Our patient received volumetric-modulated arc cranial radiotherapy (VMAT) with a total 54 Gy in 16 fractions. She remains well without disease recurrence for 14 months. Tumour markers are normal with stable MRI brain and whole-spine. Endocrine follow-up confirms recovery in all pituitary axes except the gonadotrophins for which she remains on hormone replacement therapy. **Conclusion:** This case demonstrates an excellent outcome for a young adult diagnosed with a suprasellar GCT treated with chemotherapy followed by stereotactic radiotherapy. It highlights the importance in keeping a high clinical index of suspicion in young adults presenting with a midline intracranial tumour, visual dysfunction and an endocrinopathy. CSF analysis identified an elevated tumour marker level, which bypasses the need for a histological diagnosis.

Neuroendocrinology and Pituitary

NEUROENDOCRINOLOGY AND PITUITARY CASE REPORTS

A Case of Relapse of Lymphocytic Hypophysitis Triggered by the Pregnancy of the Second Child

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Lymphocytic hypophysitis is a rare immune-mediated inflammatory disorder that causes pituitary dysfunction. It has been reported that lymphocytic hypophysitis onset during pregnancy rarely relapses or exacerbates in subsequent pregnancies. We herein report a patient with relapse of lymphocytic hypophysitis triggered by the pregnancy of the second child. **Case Presentation:** At the age of 34, at 28 weeks of gestation of the first child, she became aware of left visual field disorder and was diagnosed as an upper left visual field defect. An MRI scan revealed an enlargement of the pituitary gland and the thickening of the stalk. She was referred to our hospital for diagnosis and treatment. Laboratory data showed central adrenocortical dysfunction and central hypothyroidism. Based on the course of the disease, MRI findings and laboratory data, we diagnosed her as lymphocytic hypophysitis occurred during pregnancy. With a replacement dose of hydrocortisone and levothyroxine, she gave birth by cesarean section at 38 weeks of gestation. We performed detailed assessment of anterior pituitary functions with hypothalamic hormone challenges after giving birth. It showed panhypopituitarism without diabetes insipidus. An MRI scan found the compression of the optic chiasm remained after childbirth, the patient underwent steroid pulse therapy. After that, visual field defect improved rapidly, and the patient continued to receive oral prednisolone with gradually reduced amount. An MRI scan performed over time and found the pituitary swelling gradually improved. The pituitary was completely intact 3 years after the onset of disease. At the age of 38, the patient became pregnant with her second child, showed no signs of hypopituitarism at the time of pregnancy. She still had been administrated with 3.5mg/day prednisolone. At the 21 weeks of pregnancy, she became aware of blurred vision and was diagnosed as a left paracenter scotoma. Laboratory data showed a decrease in blood glucose and neutrophil count, suggesting the occurrence of central adrenocortical insufficiency. Therefore, we suspected the relapse of hypophysitis due to second pregnancy. We started hydrocortisone supplementation in addition to prednisolone. No MRI scan was performed during pregnancy, since no progression of visual impairment was observed. She gave birth at 37 weeks of gestation, and postpartum MRI scan showed mild thickening of the stalk. Steroid pulse therapy was not performed because the visual field abnormality was spontaneously improved. Lymphocytic hypophysitis has a diverse course, and there is currently no confirmed risk factor for recurrence. In this case, hypophysitis recurred due to pregnancy despite the continuation of prednisolone administration, and the pathogenic mechanism may be different from the previously reported cases of recurrence of hypophysitis.