Hypophysitis associated with a ruptured Rathke’s cleft cyst in a woman, during pregnancy

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ABSTRACT. We report the case of a 29-yr-old woman who first presented an aseptic meningitis at the beginning of a pregnancy. She was admitted one month later with headaches and vomiting. Panhypopituitarism with diabetes insipidus was diagnosed. Magnetic resonance imaging (MRI) data suggested the existence of lymphocytic infundibulohypophysitis, with inflammation of the suprasellar area. No new symptoms were noticed until 6 months later when this patient pointed out troubles of the visual field, due to a compression of the optic chiasma. Three boluses of 1 g methylprednisolone were prescribed, with no effects. After delivery, the defects of the visual field increased. A neurosurgical intervention was decided. Diagnosis of Rathke’s cleft cyst (RCC) was made. We concluded that this patient presented a rupture of a RCC, which occurred at the beginning of pregnancy, associated later with panhypopituitarism with diabetes insipidus, due to a probable hypophysitis. The end of the pregnancy was marked by consequences of an increased volume of the RCC. To our knowledge, this case is the first described during pregnancy.

INTRODUCTION

Rathke’s cleft cysts (RCC) are benign, intra- or juxtasellar tumors derived from remnants of Rathke’s pouch. This type of tumor is considered rare, even if its frequency has been estimated to be as high as 3.7% in routine autopsies (1). RCC is usually asymptomatic. When it is not, signs are generally due to a compression of pituitary gland, optic chiasm, or hypothalamus by large cysts (2). Hypophysitis associated with RCC was described in only few cases in non-pregnant women (3-8). Hypophysitis seems to be due to a rupture of the cyst and leakage of the contents in the area. We report here a new case of hypophysitis associated with a possible rupture of an RCC in a woman, during pregnancy.

CASE REPORT

A 29-yr-old woman was admitted in October 2003 to a Breton Hospital, with a suspicion of meningitis. She was 6 weeks pregnant. She had had persistent headaches for 10 days. Hyperthermy appeared at a late time, with photophobia and stiff neck. The cerebrospinal fluid (CSF) showed 760 white cells/mm³ (84% neutrophilic cells). Proteinorachia was normal (0.6 g/l). On routine laboratory examinations, no inflammation marker was elevated. No germ was found on blood and CSF, despite cultures and PCR analysis. Computed tomography (CT) scan was described as normal. Antibiotic therapy was introduced (clamoxycycline). Headaches and hyperthermia disappeared during the first two days of hospitalization, only to reappear 10 days later. In CSF, 1590 white cells (80% neutrophilic cells) were found (proteinorachia: normal; no germs). Ceftriaxone was added. Four days later, only 43 cells (75% lymphocytes) were present at the CSF control. She then returned at home. However, in November 2003, she was again admitted. She had headaches, vomiting and was without hyperthermia. The new CSF analysis showed 26 lymphocytes (proteinorachia: 0.62 g/l). An important polyuria was found (diuresis: 17 l/day), with hypokalemia and dDAVP was introduced. Despite her pregnancy, a magnetic resonance imaging (MRI) with an injection of gadolinium was made. It revealed the presence of an enlargement of the hypophysis, isointense on T1-weighted images except a small hypointense part, hyperintense on T2-weighted images. A thickening of the pituitary stalk was seen, and a large and intense hypersignal on T2-weighted images.
images of the suprasellar area, compatible with an inflammation (Fig. 1). The bright posterior lobe was not found. Diagnosis of hypophyseal abscess was first made. A new antibiotic therapy was begun. She was then admitted to our hospital.

After high doses of hydrocortisone IV, vomiting disappeared. Hypopituitarism was confirmed: cortisol: 1.18 ng/l, ACTH: 4 pg/ml (no. >9); free T₄ (FT₄): 8 pg/ml (no. >10); TSH: 0.1 mU/ml (no. >0.5). With ddAVP, hydrocortisone, L-T₄, symptomatology slowly disappeared. In the visual field, fundoscopy and neurological examinations were normal.

In this context, a new MRI without injection of gadolinium showed a decrease of the endosellar enlargement and the inflammation of the hypothalamic area 15 days later. The bright posterior lobe of the pituitary was not seen on T₁-weighted images (Fig. 2). We then made the hypothesis of a lymphocytic infundibulohypophysitis, with partial necrosis of the adenohypophysis. Antibiotics were stopped. No biological or clinical argument was found for an infiltrative disease such as tuberculosis, sarcoidosis, histiocytosis, or for an autoimmune disease (antithyro-peroxidase antibodies were negative). Antipituitary antibodies were looked for, by an immunofluorescence technique (serum diluted at 1:10, on lengths of antithyroid and pars intermedia from the thyroidectomised guinea pig) but not found. Unfortunately, antibodies to vasopressin-cells were not searched.

No new fact during her pregnancy was noticed until April 2004, when this patient pointed out troubles of the visual field (bitemporal hemianopsia). A new MRI showed a compression of the optic chiasma by the stalk and the pituitary (Fig. 3). Three boluses of 1 g methylprednisolone were prescribed, with no effects. She gave birth to a girl 10 days later (36 weeks of pregnancy). An onset of lactation was noticed 3 days later and treated by