OBSTETRIC CASE REPORTS

A case of recurrent lymphocytic hypophysitis in pregnancy

D. SINHA, A. SINHA, & A. M. PIRIE

Department of Obstetrics and Gynaecology, Birmingham Women's Hospital, UK

Case report

A 26-year-old female in her first pregnancy at 38 weeks presented with 6 weeks history of right-sided headache and bilateral gradual deterioration of vision for the last 2 weeks. There was no significant past medical history including autoimmune disorders and her family history was unremarkable. Her periods were regular prior to conception. Pregnancy had been otherwise uneventful. She was referred to our institution after having had an MRI scan, visual perimetry and relevant blood tests. MRI scan had shown a pituitary mass that had caused enlargement of the sella turcica and was compressing the optic chiasma. Figures 1 & 2. A formal visual perimetry had shown bilateral homonymous hemianopia. Her pre-eclamptic profile was normal. Serum prolactin was mildly elevated to 4,876 milliunits (normal levels in pregnancy, up to 4,400 milliunits). Her thyroid function test was suggestive of secondary hypothyroidism TSH: 0.9 mU/l (normal, 0.4–4 mU/l) and T4: 6.0 pmol/l (normal, 10.6–21 pmol/l). Other endocrine profiles were within normal limits.

A working diagnosis of pituitary apoplexy was made and she was commenced on hydrocortisone supplementation (20 mg twice daily). In view of the progressive nature of her symptoms and impending visual loss, a decision was made to surgically decompress the pituitary gland. Prior to proceeding with this, labour was induced with prostaglandin and she delivered vaginally a live male infant weighing 3.2 kg. This was followed by trans-sphenoidal excision of the pituitary tumour by the neurosurgical team. During the postoperative period, she continued on hydrocortisone and thyroxine replacement therapy. She became polyuric for a period as a result of her surgery, but this settled with conservative management. She made a good postoperative recovery and was discharged on hydrocortisone 20 mg b.d. along with thyroxine 100 µg/day. Histology of the pituitary lesion reported a diagnosis of lymphocytic hypophysitis.

She had a repeat MRI scan after 4 months which showed only a minimum amount of residual tissue sitting within an enlarged pituitary fossa. Her pituitary function tests along with cortisol levels returned to normal and she was gradually weaned off her hydrocortisone. Visual acuity and peripheral fields improved steadily.

She subsequently had a spontaneous conception 1 year later with recurrence of headache and visual deterioration starting, this time, at 16 weeks' gestation. She had a repeat MRI scan, which showed an expanding pituitary lesion compressing the optic chiasma. However, she did not require surgical intervention in this pregnancy. Her symptoms were well controlled on increasing the dose of thyroxine and hydrocortisone. She had a spontaneous vaginal delivery of a healthy infant at term. She had a further MRI scan 4 months following her second delivery, which did not show any significant abnormalities of concern.

Discussion

Lymphocytic hypophysitis was first described by Goudie and Pinkerton in 1962 in a woman who suffered postpartum hypothyroidism and amenorrhoea and subsequently died of adrenal crisis after a routine appendicectomy (Goudie and Pinkerton 1962). It is a rare but an increasingly recognised pituitary disorder that occurs mostly in women at the end of pregnancy or in the immediate postpartum period. Of the cases reported in females, 55% were related to pregnancy (Tubridy et al. 2001). The aetiology is unknown but it is thought to be an autoimmune disease with both humoral and cell mediated components resulting in destruction of the pituitary gland. The autoimmune process is generally confined to the anterior pituitary and leads to hypopituitarism and pituitary mass. It has also been associated with other autoimmune conditions: Hashimoto's thyroiditis, adrenitis, atrophic gastritis and lymphocytic parathyroiditis. Several autoantibodies, like antihuman growth hormone, anti-pituitary gland and antihuman pituitary membrane antigens have been associated with this condition (Saiwai et al. 1998).

It usually presents with visual symptoms and headache due to the effect on the optic chiasma, although a case of lymphocytic hypophysitis has been described who presented with a normal perimetry (Goudie and Pinkerton 1962). Other common symptoms are nausea, vomiting and tiredness. Partial or complete loss of adrenocortical function is common. The majority of patients show deficiency of ACTH, TSH, FSH, LH and prolactin. Our patient additionally developed diabetes insipidus for a short period of time, which is a rarer manifestation and indicates involvement of the pituitary infundibulum. A similar process known as infundibuloneurohypophysitis may affect the neurohypophysis and infundibulum only and manifest as diabetes insipidus (Imura et al. 1993). Mild hyperprolactinaemia has been reported in cases not associated with pregnancy and breast feeding. This is thought to result from compression of the pituitary infundibulum (Saiwai et al. 1998). The other possibility is loss of inhibitory action of dopamine on prolactin release as a result of inflammatory reaction which may have altered the dopamine receptor.

The main diagnostic dilemma includes expanding pituitary adenoma or meningioma, both of which may enlarge considerably during pregnancy. Pituitary apoplexy may be associated with a pituitary adenoma but may occur spontaneously in a normal gland during pregnancy (Lee and Pless 2003). Inflammatory and infectious processes such as tuberculosis, sarcoidosis and giant cell granuloma may also present with similar symptoms. A diagnosis of lymphocytic hypophysitis should always be considered as a part of differential diagnosis of pituitary mass or deficiency presenting peripartum. In such cases, if there is no surgical emergency (i.e. impending visual loss), then conservative treatment with hormone replacement and a trial of increasing the dose of steroid should be attempted. We successfully managed our patient with the same therapeutic regimen in her second pregnancy.

Chronically, many patients require modest to complete pituitary hormone replacement. MRI or CT with contrast generally reveals a homogenously enhancing pituitary mass with supracellar extension (Saiwai et al. 1998). A definitive diagnosis can only be made by biopsy (Gagneja et al. 1999).
The course of disease varies from spontaneous resolution or resolution with cortisone treatment (Krimholtz et al. 2001) to development of progressive hypopituitarism (Saiwai et al. 1998). Death due to adrenal insufficiency was also reported (Goudie and Pinkerton 1962). In the absence of visual failure, pituitary replacement therapy and repeated visual field and radiological assessment may be all that is required. High doses of corticosteroids have been used in some patients to ameliorate visual impairment (Krimholtz et al. 2001). Progressive visual impairment and uncertain diagnoses are indications for surgical intervention (Gagneja et al. 1999).

Due to the variable natural history of lymphocytic hypophysitis, an accurate assessment of risk of recurrent disease cannot be made. Spontaneous conception has been reported following lymphocytic hypophysitis in previous pregnancy (Gagneja et al. 1999). Gonadotrophin failure has been reported in over 30% of women and HRT is required to induce withdrawal bleeding and to treat the effects of hypo-oestrogenism. Successful induction of ovulation with HMG and HCG has been reported in a woman with lymphocytic hypophysitis following resection of a pituitary mass. Our patient thankfully achieved spontaneous conception the second time, despite having had surgical intervention to the pituitary in her first pregnancy. Symptoms recurred again when she became pregnant for the second time but this time headache and visual problems were managed conservatively by hormone replacement to good effect. She had complete resolution of symptoms following her delivery and was weaned off her hormones.

References


Correspondence: D. Sinha, 14 Wellington Close, Kingswinford, West Midlands DY6 8JG, UK. E-mail: adsinha@hotmail.com

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