Recurrent Lymphocytic Hypophysitis during Two Pregnancies: A Very Rare Care

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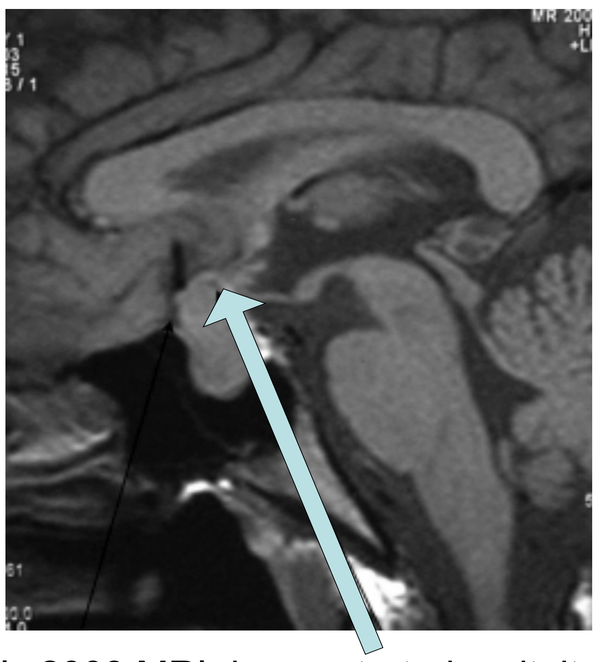
1st Presentation July 2003

- **PC**: 25 y/o Afro-Caribbean woman presented in July 2003 with a 6/52 history of headache and a 2/52 history of visual deterioration whilst 38/52 pregnant.
- **PMHX**: Nulligravida, Appendectomy 1986 and Iron Deficiency Anaemia.
- **Investigations**: Formal vision testing demonstrated bi-temporal hemianopia. Pituitary MRI revealed a sellar mass with suprasellar extension into the chiasm. Initial blood tests showed secondary hypothyroidism.
- **Treatment**: She was started on hydrocortisone, total dose 20mg daily and had an induction of labour delivering a healthy boy. Following delivery she had an urgent trans-sphenoidal removal of the mass; pathology was consistent with Lymphocytic Hypophysitis (LyH).
- **Post-Treatment**: Post-operatively, she made good recovery with marked visual improvement. Short synacthen test was normal and she was breastfeeding. A follow up pituitary MRI (November/2003) revealed a small amount of tissue within an enlarged fossa with no significant suprasellar extension.

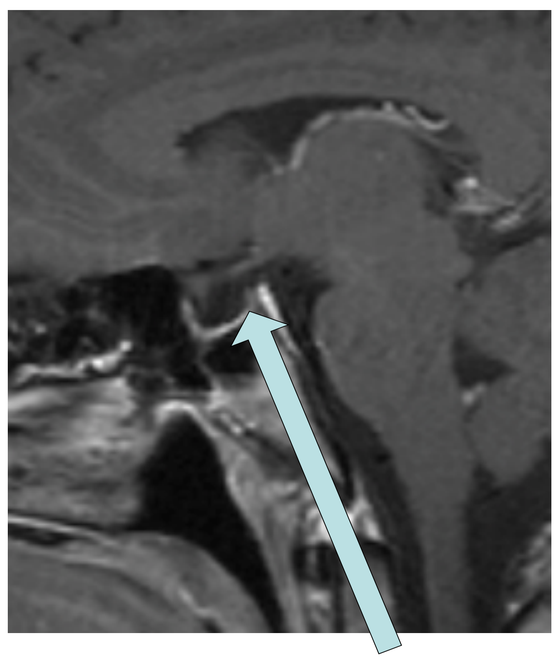
2nd Presentation April 2004

- **PC:** In January 2004, she conceived again and at the 14th week of pregnancy she was experiencing increasingly generalised headaches and bilateral visual disturbance.
- **Investigations**: A repeat pituitary MRI showed a bulky pituitary with a recurrence of swelling elevating the chiasm. Her visual fields once again presented with bi-temporal field defects. She had normal thyroid function.
- **Treatment**: Restarted on hydrocortisone (final dose: 30 mg am, 15 mg pm). In the subsequent weeks, she had close visual monitoring and no further deterioration was detected.
- **Post-Treatment**: In September 2004, she delivered a healthy boy. Post-partum, her vision returned to normal and follow-up imaging showed gradual resolution of the LyH. Her periods returned and adequate ACTH reserve was confirmed.
- **How she is today**: She had no further pregnancies. In her latest follow-up December 2016, she is on levothyroxine and GH replacement. No further recurrence of LyH was detected.

Investigations



July 2003 MRI demonstrated a pituitary mass with suprasellar extension, compressing the chiasm



June 2009 MRI demonstrating an empty sellar, normal pituitary infundibulum and hypothalamus

Hypophysitis

- Hypophysitis is a rare (Incidence 1 in 7-9 million) inflammation of the pituitary gland. There are four main histologic subtypes, (Lymphocytic, Granulomatous, Xanthomatous, Necrotizing and IgG4 Plasmacytic) each with their own variation of presentation and prognosis.
- LyH is the most common subtype (over 390 cases reported) characterised by diffuse infiltration of pituitary by B and T lymphocytes forming lymphoid follicles and distorting normal architecture which often occurs during pregnancy.
- LyH usually affects women in last month of pregnancy or in first two post-partum months. Clinical manifestations include headaches, visual deterioration and pituitary dysfunction.
- Shared antigens of placenta and pituitary suggested as causative
- Management includes observation, surgery or steroid treatment and replacement of the pituitary hormone deficits.

Conclusion

- Documented cases of recurrent hypophysitis in women experiencing multiple pregnancies are extremely rare.
- Although, it has been suggested that a history of LyH in pregnancy does not increase the risk of developing LyH in subsequent ones, our case demonstrates the variable natural history of this condition.

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