Relapsing lymphocytic hypophysitis with progressive hypopituitarism and permanent neurological deficit

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Lymphocytic hypophysitis is a rare autoimmune disease of the pituitary gland. A 39-year-old woman presented with a 6-month history of general malaise, anorexia, nausea and vomiting followed by a 3-month history of secondary amenorrhoea. Basal pituitary function testing revealed: fT4 5.1 picomoles per litre, TSH 1.6 milliunits per litre, cortisol 32 nanomoles per litre, prolactin 1167 microinternational units per millilitre, LH 2.8 units per litre and FSH 5.1 units per litre. Dynamic pituitary function testing confirmed severe ACTH, TSH and GH deficiency. MRI scan of the pituitary revealed typical appearances of lymphocytic hypophysitis. Treatment with hydrocortisone, thyroxine and growth hormone restored general well-being and normal menstruation. Repeat MRI scanning at 18 months demonstrated normal pituitary appearances and pituitary function testing confirmed persistent ACTH, TSH and GH deficiency. 41 months after presentation she became pregnant and underwent a termination. Six months later she developed secondary amenorrhoea which has persisted. A repeat MRI revealed similar appearances to that at presentation. At 57 months she returned with headache and spontaneously resolving right third cranial nerve palsy. MRI appearances were unchanged. One month later she developed headache, relapse of the right third cranial nerve palsy with additional fourth and fifth cranial nerve palsies and rapidly developed blindness of the right eye. MRI scanning demonstrated similar appearances to those at initial presentation with extension of the inflammatory lesion to the right cavernous sinus and orbital apex. Treatment with high dose prednisolone rapidly corrected symptoms and neurological defects other than the blindness. Repeat MRI scans at 62 and 65 months were normal but panhypopituitarism and blindness of the right eye persisted. This case demonstrates the unpredictable nature of lymphocytic hypophysitis with additional significant extra pituitary involvement.