Case report:
Indolent IgG4 hypophysitis with partial anterior pituitary failure

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BACKGROUND

IgG4 hypophysitis is an increasingly recognised entity causing anterior pituitary failure or diabetes insipidus (1). A series reported underestimation of its prevalence, at 4% of those with hypopituitarism or diabetes insipidus, with mean age of diagnosis at 62 years old (2). MRI imaging typically revealed pituitary enlargement or stalk thickening (3). In acute or subacute clinical cases presenting with headache and ophthalmological involvement (diplopia or visual field defect), successful treatment with immunosuppressive dose of steroid leading to radiological improvement and resolution of these symptoms (4,5). However, In the lack of agreed guideline in the matter, the necessity of immunosuppressive steroid shall be tailored according to the clinical course. We herein report a case of IgG4 hypophysitis of a more indolent clinical course, which was safely managed conservatively with hormonal replacement for 3 years.

CASE HISTORY

A 79-year-old man with presented with a fall and vomiting.

The finding of significant postural hypotension associated with severe hypotonic hyponatraemia (Na 114 mmol/L, Osmolarity 244 mmol/L) mandated a Short Synacthen Test (cortisol 98mmol/L at baseline and 238mmol/L 30-minute post synacthen) and ACTH (6ng/L) which confirmed central hypoadrenalism.
This led to revelation of multiple pituitary axes involvement:

- hypogonadism (testosterone 1.1 nmol/L, FSH 1.3U/L, LH 1.1U/L),
- growth hormone deficiency (IGF-1: 5nmol/L)
- partial central diabetes insipidus confirmed on a water deprivation test.

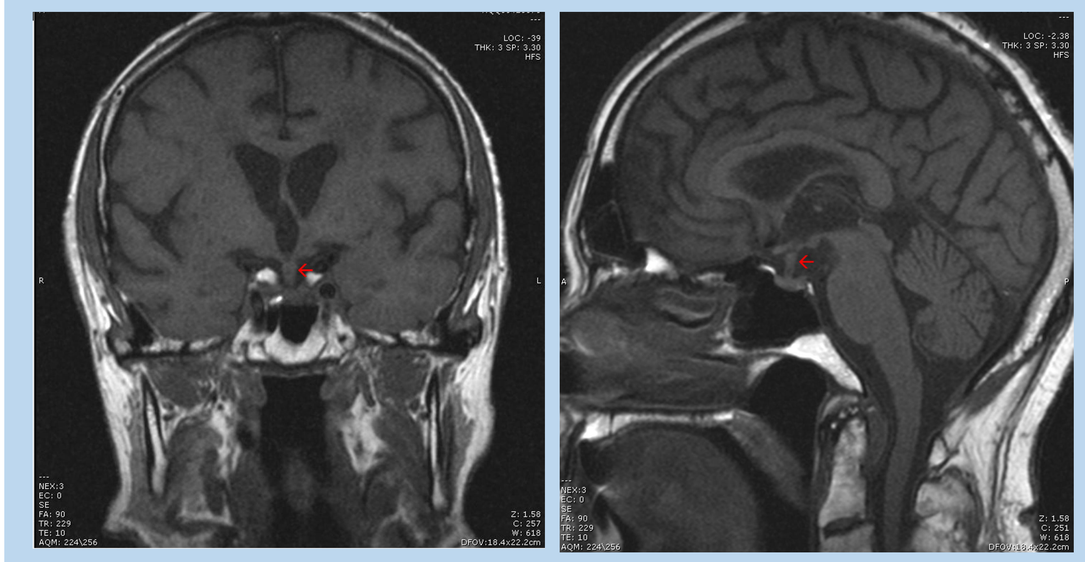
(The prolactin and thyroid test were normal.)

Confrontation visual field test was unremarkable. The MRI pituitary then revealed a normal pituitary gland with thickened stalk up to 5mm (Fig 1). A discussion at pituitary multidisciplinary meeting (MDT) raised suspicion for lymphocytic hypophysitis and led to IgG4 level measurement, which was markedly elevated at 12.7g/L (0-1.3g/L).

Granulomatous infiltration was less likely in this context especially with normal calcium and ACE level. A screening for systemic IgG4 disease with CT thorax and abdomen revealed multiple hilar and mediastinal lymphadenopathy, measuring up to 1.3cm. These findings were stable on serial imaging which precluded bronchoscopy.

On this ground, a diagnosis of IgG4 related hypophysitis was entertained. In the absence of sight-threatening radiological findings or debilitating symptoms such as headache, high dose steroid was not required as the condition was deemed to be indolent. Patient's symptoms responded to physiological hydrocortisone and testosterone replacement and remained so for the subsequent 3 years of follow-up. Serial annual MRI pituitary consecutively showed non-progressive findings.

IMAGING



A B

Fig 1. MRI brain (T1 sequence) of coronal (A) and sagittal (B) view demonstrating the thickened pituitary stalk (red arrow) with otherwise normal pituitary gland.

CONCLUSION AND LEARNING POINTS

While acute hypophysitis presenting with florid symptoms or ophthalmological involvement may require high dose steroid, this case presented as indolent "burn-out" disease was safely managed with mere hormonal replacement with no evident progression in the 3 year of follow-up.

Histological confirmation may not be possible or even meaningful in these settings.

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