Unusual presentation of more common disease/injury

Partial hypopituitarism and Langerhans cell histiocytosis

S Balaguruswamy,¹ P D Chattington²¹Department of Endocrinology and Diabetes, Royal Liverpool University Hospital, Liverpool, Merseyside, UK;²Department of Endocrinology and Diabetes, Warrington Hospital, Warrington, Cheshire, UK

Correspondence to S Balaguruswamy, drbsaravanan@yahoo.com

Summary

A case of multisystem Langerhans cell histiocytosis with pituitary involvement nearly 20 years after initial presentation. A 48-year-old man had histiocytosis X 22 years ago initially involving the groin; subsequently his external auditory meatus, scalp, gum, mandibular bone, perineum and axilla were involved and treated. The pituitary gland was involved 4 years ago. A thyrotropin-releasing hormone test showed delayed response suggestive of hypothalamic disease. Prolactin levels were normal. A gonadotropin-releasing hormone test showed impaired testosterone and gonadotrophin response in keeping with pituitary disease. A glucagon stimulation test showed an impaired growth hormone response but a normal cortisol increase. MRI pituitary showed an empty sella. There was no evidence of diabetes insipidus. Bone mineral densitometry was normal. He has partial hypopituitarism needing thyroxine and testosterone replacement. He also developed type 2 diabetes mellitus 9 years ago. He is closely monitored for any development of diabetes insipidus and the need for growth hormone supplementation.

BACKGROUND

Langerhans cell histiocytosis (LCH) is a rare disease. The granulomatous deposits occur at multiple sites within the body and often involve the hypothalamo-pituitary axis. The LCH cases reported in the literature had diabetes insipidus and most had pan-hypopituitarism. Our patient currently does not have diabetes insipidus. We are currently monitoring for this developing and for the occurrence of pan-hypopituitarism.

CASE PRESENTATION

A 48-year-old man had histiocytosis X initially involving the groin in 1987 and he responded poorly to treatment with topical nitrogen mustard and radiotherapy. In 1988, there was involvement of the external auditory canals requiring several meatoplasties. Scalp involvement in 1990 required chemotherapy. He then developed gum and mandibular disease in 1992, which was resected. Perianal disease was treated with pelvic radiotherapy in 1993. He had a relapse in axilla in 1995 and pituitary involvement was diagnosed in November 2006. He also has hypothyroidism and type 2 diabetes mellitus (diet controlled)—both diagnosed in 2001. In 2006, he presented with erectile dysfunction. He has good glycaemic control.

INVESTIGATIONS

Initial investigations showed low luteinising hormone, follicle stimulating hormone and testosterone. A thyrotropin-releasing hormone test showed delayed response suggestive of hypothalamic disease. Prolactin levels were normal. Gonadotropin-releasing hormone test showed impaired testosterone and gonadotrophin response in keeping with pituitary disease. A glucagon stimulation test showed an impaired growth hormone response but a normal cortisol increase. MRI pituitary showed an empty sella. There was no evidence of diabetes insipidus. Bone

mineral densitometry was normal. Recently, he was diagnosed to have diet controlled type 2 diabetes mellitus.

TREATMENT

Our patient has partial hypopituitarism. He is being treated with levothyroxine 75 µg a day and intramuscular testosterone injection 1 g every 12 weeks.

OUTCOME AND FOLLOW-UP

Currently our patient does not have symptoms of growth hormone deficiency and does not fit the criteria for growth hormone supplementation. He is also being monitored for any development of diabetes insipidus and cortisol deficiency.

DISCUSSION

Despite treatment, nearly 20% of patients with multisystem involvement have a progressive disease course.¹ The LCH cases with pituitary involvement reported in the literature had diabetes insipidus and most had pan-hypopituitarism.² Our patient currently does not have diabetes insipidus. Ongoing monitoring is needed to check for this developing and for the occurrence of pan-hypopituitarism.

Learning points

- ▶ LCH X often involves the hypothalamo-pituitary axis.
- ▶ Diabetes insipidus is usually associated with LCH.
- ▶ Despite treatment, nearly 20% of the patients with multisystem involvement have a progressive disease course.

Competing interests None.

Patient consent Obtained.

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