XANTHOMATOUS HYPOPHYSITIS AS A CAUSE OF CLUSTER HEADACHE
A CASE REPORT

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ABSTRACT
Introduction: Xanthomatosis is an inflammatory disease of the pituitary gland that may mimic pituitary tumors clinically and radiologically. Primary hypophysitis has traditionally been classified as lymphocytic (LG), granulomatous (GH), and xanthomatous (XH).

Case description: We report on a case of a xanthomatous hypophysitis initially diagnosed as pituitary adenoma. A 23-year-old man suffered from typical cluster type headache. Two years after the first symptoms, we confirmed diabetes insipidus. All the anterior pituitary hormone levels were normal. Sella MRI scan depicted a 14x10x17 mm inhomogenous mass. Transphenoidal surgery was performed; the removed tissue showed accumulation of foamy cells and xanthomatous-sphingolipoid cells. Following an unsuccessful postoperative recovery, severe cluster type headache returned after stopping the hydrocortisone therapy. The endocrine work-up revealed hypothrophydia (morning cortisol: 86 nm/L, ACTH:3.8 pm/L), hypothyroidism (T4:10.5 pm/L), hypogonadism (testosterone: 3.44 nm/L). We restarted glucocorticoid therapy, administered to one of the patients with XH has been described.

The overall prognosis for XH is good, but improvement of pituitary function after transsphenoidal surgery has been reported in less than 55% of patients. Glucocorticoid therapy has been advocated to reduce inflammation and has been temporarily effective in some patients with primary hypophysitis. However, a recent study demonstrated that methyprednisolone therapy, administered to one of four patients with XH, did not yield an efficient outcome. Steroid therapy seems to be less effective in the treatment of primary hypophysitis.

Our case:
We report a 23-year-old man with XH who presented with cluster type headache, diabetes insipidus and MRI-proven pituitary intrasellar mass.

Since 2009 our patient suffered from recurrent headache. CT scan, ophthalmological, neurological investigations revealed no obvious cause of the symptoms. In April 2011, polyuria-polydipsia occurred and endocrine investigations revealed diabetes insipidus. Anterior pituitary hormone levels were normal: TSH: 1.3 mIU/L, FSH: 2.4 IU/L, LH: 3.7 IU/L, PR-L: 197 mIU/L, ACTH: 7.78 pm/L, cortisol: 0.44 nm/L. After initiation of dexamethasone treatment, diarrhea returned to normal. The pituitary MRI scan revealed a 14x10x17 mm inhomogenous lesion with the disappearance of the hypophysis signal of the neurohypophysis.

In July 2011, transsphenoidal surgery was performed and histology proved xanthomatosus hypophysitis. We were able to manage the patient with glucocorticoid supplement without any complication. The headache resolved but the diabetes insipidus persisted. The anterior pituitary function after the surgery was normal: serum cortisol 08 h: 404-445 nm/L, ACTH:4.9 pm/L, FSH:3.1 mIU/L, LH:4.2 mIU/L, TSH:1.61 pm/L. 2 months later severe cluster type headache occurred. Endocrine investigations revealed hypothrophydia, hypothyroidism and parenchymal hypogonadism: serum cortisol: 08 h: 96 nm/L, TSH: 1.32 mIU/L, LH: 10.5 pm/L, FSH: 3.3 mIU/L, LH: 2.8 mIU/L. TSH, ACTH: 3.8pm/L. LH/HRH test results: FSH: 0 min: 2.8, 30 min: 4.7, 60 min: 5.1 mIU/L, LH: 0 min: 2.9, 30 min:13.5, 60 min:13.8 mIU/L.

The postoperative pituitary MRI scan proved the persistent presence of the inhomogenous mass. After initiation of glucocorticoid replacement the headache disappeared. With levothyroxin, testosterone supplementation and gradually lowered dosage of GC and all symptoms disappeared with the exception of diabetes insipidus.

Despite of low IGF 1 (92 ng/ml, age matched reference rate:117 ng/ml), hypogonadism (testosterone: 3.44 nm/L, testosterone: 9.8, 30 min:13.7 ng/ml), hypogonadism (testosterone: 3.44 nm/L, hypogonadism (testosterone: 3.44 nm/L, hypogonadism (testosterone: 3.44 nm/L). We restarted glucocorticoid (GC) treatment only in case of recurrent cluster type headache, but no persistent replacement is needed.

Conclusion:
Typical cluster type headache and diabetes insipidus were the two main syndromes of the XH in our case.

The patient requires GC supplementation only in case of recurrent cluster type headache, but no persistent replacement is needed. The cause of the XH is still unknown, but regular endocrine check-up can reveal disturbances in pituitary function and, as in our case, glucocorticoid replacement seems to be effective in the treatment of the disease.

References:
3. L.Aste et al.: XANTHOMATOUS HYPOPHYSITIS MIMICKING A PITUITARY ADENOMA: CASE REPORT AND REVIEW OF THE LITERATURE, J.of Oncolo...