We report a 23-year-old man with XH who presented with cluster type headache, diabetes insipidus and pituitary MRI proven 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 polynya-polyposis occurred, the endocrine investigations revealed diabetes insipidus. The anterior pituitary hormone levels: TSH: 1.3 mIU/L, FSH: 2.4 UI/L, LH: 3.7 UI/L, PRL: 197 mUI/L, ACTH: 7.78 pmol/L, cortisol 08 h: 444 nmol/L despite low testosterone level (7.36 mIU/L). After initiation of ddKVP treatment, diabetes returned to normal.

The pituitary MRI scan revealed a 14x10x17 mm inhomogenous lesion with the disappearance of the hypophysial signal of the neurohypophysis. In July 2011 transsphenoidal surgery was performed. The histology proved xanthomatous hypophysitis.

Without having any perioperative complication we could stop the glucocorticoid (GC) treatment. The headache resolved but the diabetes insipidus persisted.

After the surgery the anterior pituitary function was normal: serum cortisol 08 h: 404-445 nmol/L, ACTH: 6.49 pmol/L, FSH: 3.1 mUI/L, LH: 4.2 mUI/L, TSH: 6.61 mIU/L. 2 months later severe cluster type headache occurred. The endocrine investigations revealed hypoadrenia, hypothyroidism and peripheral hypogonadism: serum cortisol 08 h: 96 mIU/L, TSH: 1.32 mUI/L, hLH: 10.5 mIU/L, testosterone:3.44 pmol/L, FSH: 3.3 mUI/L, LH: 2.8 mIU/L, ACTH: 3.38 pmol/L.

LHRH test results: FSH: 0.8 mIU/mL, 20 min: 4.6 mIU/mL, 60 min: 5.5 mIU/mL, LH: 0.5 mIU/mL, 20 min: 2.3, 60 min: 15.9, 60 min: 13.8 mIU/mL.

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

Despite of low IGF 1 (92 ng/ml age matched reference rate:117-329 ng/ml) and GH (0.08 ng/mL) levels GH therapy was not introduced. Autoimmune screen: ANA, antiCL, antihB2GP, anti transglutaminase, anti TPO, anti parietal cell antibody was negative.

Regullarly performed sella MRI scans showed no change in tumor size and appearance after the the surgery and after the introduction of hormone replacement therapy. The patient requires GC supplementation only in case of recurrent cluster type headache, but no persistent replacement is needed.

In January 2013 we had the possibility to measure the patient's serum IgG4 level, which was markedly increased (serum IgG4 concentration: 815 mg/dl) suggesting the xanthomatous hypophysitis to be IgG4-related disease.

Conclusion:

In our case typical cluster type headache, diabetes insipidus and severe, persistent hypogonadism were the main symptoms of the xanthomatous hypophysitis (XH). The patient requires GC supplementation only in case of cluster type headache, but permanent testosterone replacement

The patient's elevated serum IgG4 concentration is suggesting the XH to be IgG4 related disease.

The cause of the XH is still unknown, but as our data suggests it could be IgG4 related disease.