Headache secondary to corticosteroid withdrawal a case report

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INTRODUCTION AND OBJECTIVE

Headache attributed to increased cerebrospinal fluid pressure (ICP) is a syndrome characterized by an elevation of intracranial pressure without hydrocephalus, with normal cerebrospinal fluid (CSF) composition and no alterations in the brain parenchyma.
Most of these cases are considered idiopathic, several possible etiologies have arised. ICP secondary to corticosteroid withdrawal is poorly understood and rarely reported in the literature while observed primarily on pediatric population.
The following case involves an adult presentation of this unusual trigger of ICP while simulating migraine.

CASE PRESENTATION

A 28-year-old male patient with a history of Crohn's disease and glaucoma treated with stable doses of azathioprine (150mg/daily), mesalazine (2gr/daily) and meprednisone (60mg/daily) for over 6 months.

Patient had no personal history of headache, his mother has migraine without aura. In October 2018 gradual corticosteroid withdrawal was started.
At week four, while receiving 10mg/daily, the patient referred right pulsatile hemicranial headaches, 9/10 in intensity, associated to photophobia and right ear phonophobia. Headache increased with Valsalva maneuvers, did not worsen with dorsal decubitus nor awaken the patient at night. It initially improved with the use of acetaminophen; but after corticoid suspension, it became persistent and increased its intensity.
A brain and orbital magnetic resonance imaging (MRI) was performed and revealed no parenchymal alterations, intrasellar arachnoidocele and increased right optic nerve sheath liquid and tortuosity (figure 1). Intracranial magnetic resonance angiography (MRA) was normal with a slight tortuosity on the transverse sinuses (figure 2).
Ophthalmology evaluation revealed mild right hyperemic papilledema, confirmed by optical coherence tomography (figure 3). Visual field was normal (figure 4).
Lumbar puncture (LP) showed an opening pressure of 260mm, CSF analysis and cultures were normal.
After 1 week of corticosteroid therapy reintroduction (20mg/daily), with a more gradual tapering, the headache was completely resolved and the patient remained asymptomatic for 6 months after complete corticosteroid withdrawal.
Patient was interpreted as a 7.1.2 headache attributed to intracranial hypertension secondary to toxic cause, according to the IHS headache classification, third edition.

DISCUSSION

Few reports of ICP associated with a decrease in corticosteroid doses, both systemic and inhaled, have been reported; most of them in pediatric population. Numerous pathophysiological hypotheses have been proposed. Nevertheless, further studies addressing these theories are needed.

It has been proposed that corticosteroid suspension decreases CFS reabsorption and increases its flow resistance. In addition, some research has suggested an increasement of CSF production after corticosteroid withdrawal through corticosteroid and local enzymes (11-β-hydroxysteroid dehydrogenase type 1 and type 2) disbalance in the choroid plexus [3,7,8].
Another possible cause is the development of cerebral edema secondary to the loss of the "brain-shrinking" corticosteroid properties after withdrawal [2].
Furthermore, the development of ICP may be mediated by alterations in calcium metabolism generated by secondary adrenal insufficiency [2].
Possible associated etiologies include underlying anemia and autoimmune diseases with corticosteroid treatment withdrawal and ICP development [4].

Higher prevalence in pediatric population suggests either a different pathophysiology in this population, or an underdiagnosis in adult patients.

CONCLUSIONS

Our case presents an unusual presentation of ICP syndrome with unilateral symptoms associated with withdrawal of corticosteroids. Interestingly, our patient's initial pain characteristics simulated a migraine crisis.
Diagnosis is supported by time relationship between corticosteroid tapering/withdrawal and the onset of ICP, presence of papilledema and imaging showing typical changes in ICP; in addition to the symptomatic resolution with steroid administration.
Performance of an LP and fundoscopy should be considered on patients presenting a headache of recent onset while descending corticosteroid treatment.

Figure 1. Magnetic resonance imaging

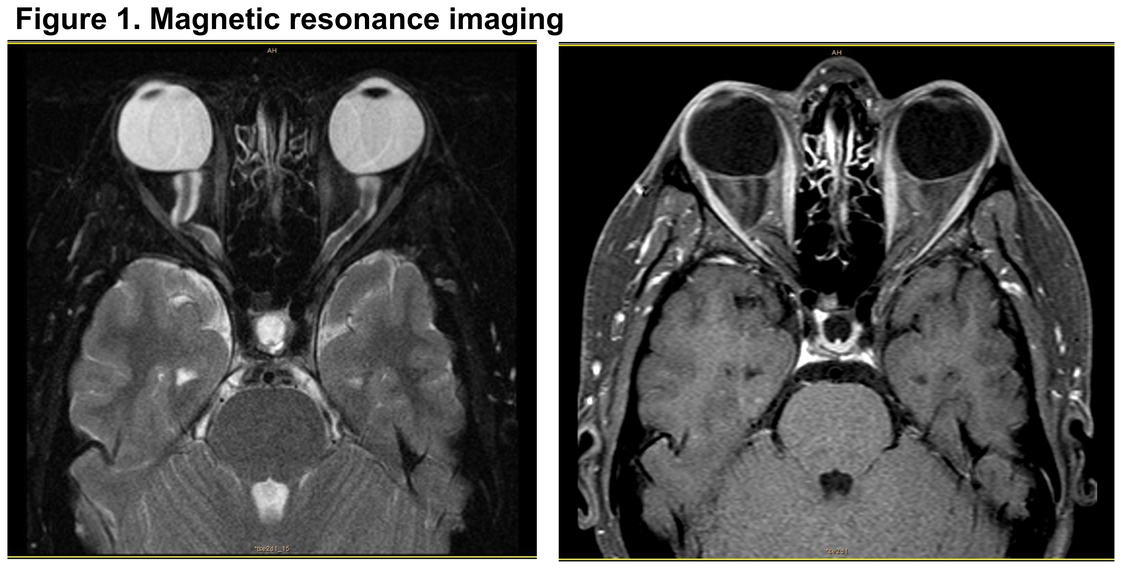


Figure 2. Intracranial magnetic resonance angiography

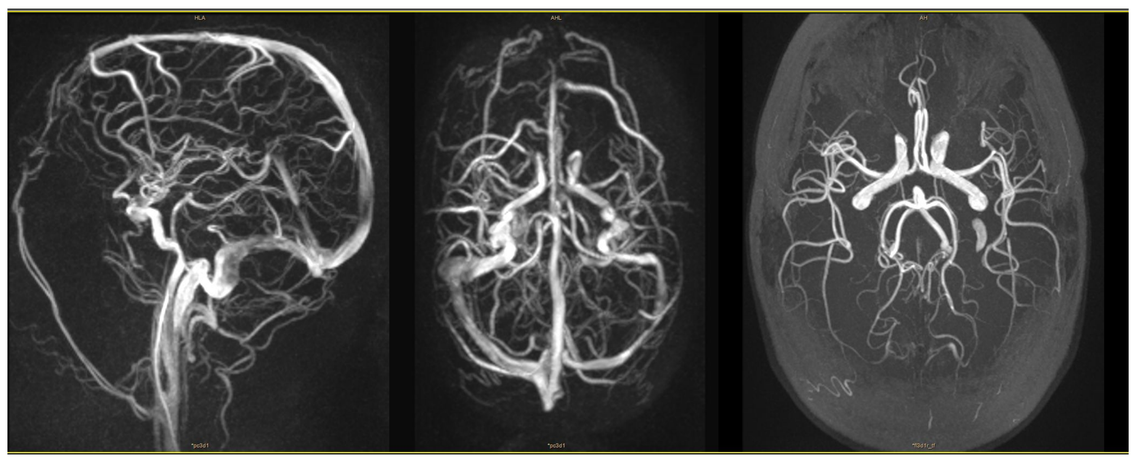


Figure 3. Optical coherence tomography (RE: right eye, LE: left eye)

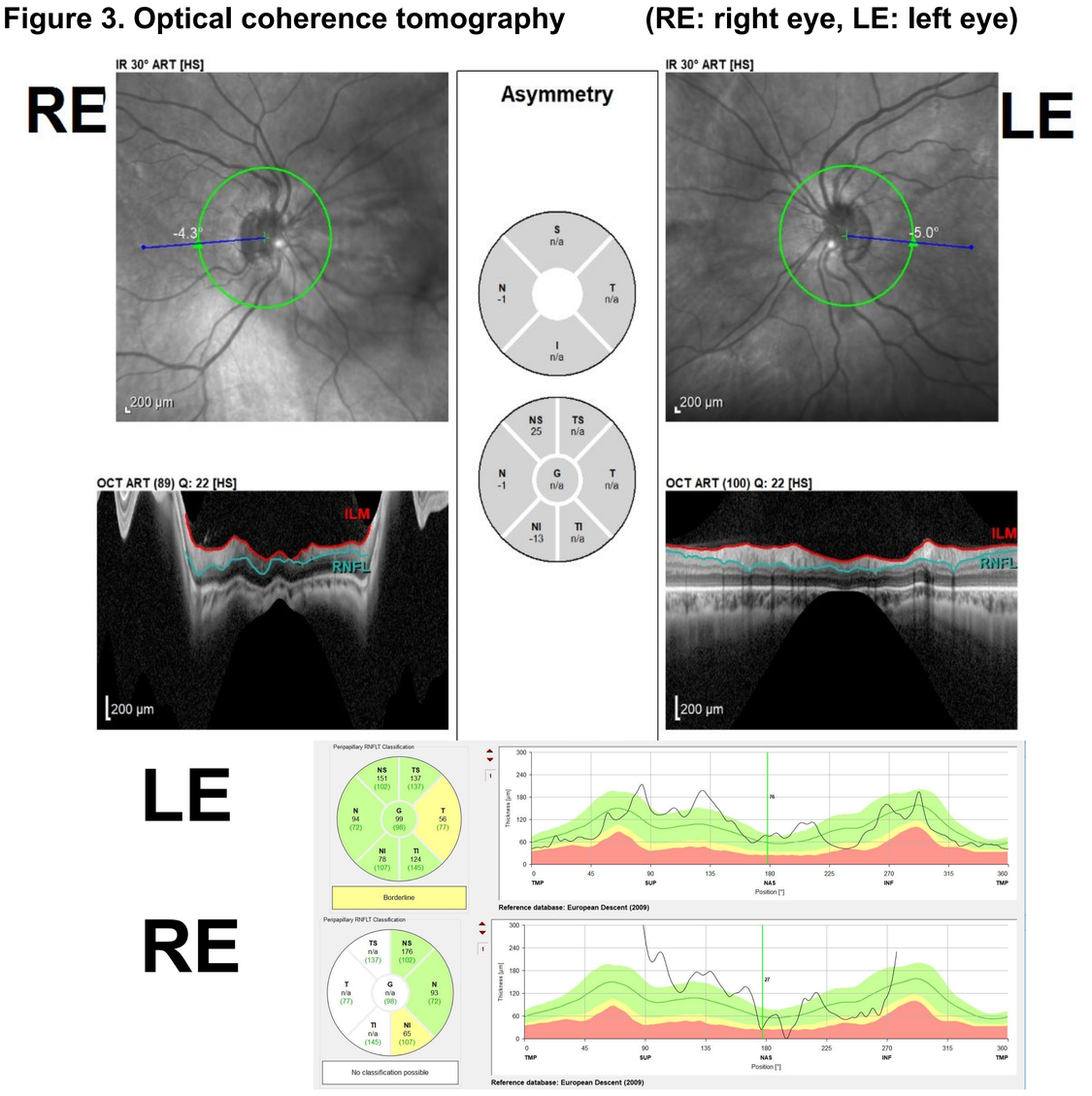
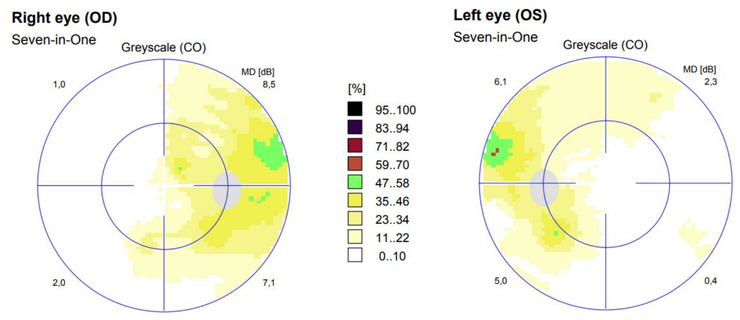


Figure 4. Computerized visual field test



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Disclosures: This poster has been previously presented at the 56th National Argentinian Congress. October, 2018

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