pp. 189-190 (2013) gineco eu

Vogt-Koyanagi-Harada disease in a patient with polycystic ovary syndrome. A case report

Alina Popa-Cherecheanu[1,2], Mirabela Ciuca[2], G. Iancu[1,3], Cristina Alexandrescu[1,2], Viorela Popescu[1]

1. UMF "Carol Davila", SUUB Ophtalmology Clinic, Bucharest (Romania)
2. Ophthalmology Department, Emergency University Hospital Bucharest (Romania)
3. Filantropia Clinical Hospital, Bucharest (Romania)

Correspondence: Dr. Cristina Alexandrescu, e-mail: alexandrescucristina76@yahoo.com

All authors contributed equally to conceiving the case-report, data collection, manuscript writing and editing and approval of the final version.

Abstract

We present the case of a young woman with polycystic ovary syndrome on oral contraceptive pills, presenting with sudden impairment of vision associated with headache, after a minor cranio-cerebral trauma. She was diagnosed with Vogt-Koyanagi-Harada disease. Steroid treatment improved the visual outcome.
Keywords: *oral contraceptive pills, polycystic ovary syndrome, serous retinal detachment, Vogt-Koyanagi-Harada disease*

Introduction

Vogt-Koyanagi-Harada (VKH) disease affects women mainly in their third and fourth decades of life. In the prodromal phase, patients with VKH disease may manifest meningeal irritation symptoms, including headache, vertigo, nausea and vomiting. In uveitic phase, bilateral exudative retinal detachment causes blurring of vision due to choroidal inflammation. After treatment with systemic corticosteroids, in the chronic phase, elevation of the neural retina gradually disappears. VKH disease may recur, typically as an anterior uveitis.

Described for the first time by Stein and Leventhal[(1)], the polycystic ovary syndrome (PCOS) associates menstrual dysfunction, hyperandrogenism (virilization), infertility, obesity and impaired glucose metabolism. The characteristic appearance of the ovaries on ultrasound consists in the presence of more than 12 follicles between 2-9 mm diameter in at least one ovary or total ovarian volume over 10 cm^3. PCOS diagnosis has to fulfil two out of the three following criteria: polycystic ovaries on ultrasound, oligo- or anovulation (manifested as oligo- or amenorrhea) and clinical or biochemical evidence of androgen excess[(2)].

PCOS is commonly diagnosed in young women, but the association with Vogt-Koyanagi-Harada disease is rare. As far as we know, there is only one similar case presented until present[(3)]. The two conditions may have a common autoimmune pathogenesis, although the role of autoimmunity in PCOS pathogenesis is still speculative[(4)].

In this study, we report a women diagnosed with PCOS that developed bilateral serous retinal detachments after a minor head trauma which has responded well to steroid treatment.

Case report

A 27-year-old female was admitted in our department for decreased vision (more severe in the right eye) for two weeks. Her past medical history revealed PCOS, treated with oral contraceptive pills (Logest®; Schering AG, Germany) and a minor head trauma in which thereafter she accused severe headache and blurred vision.

On physical examination, high body mass index (28 kg/m^2) and hirsutism were noticed.

The ocular examination found the best visual acuity in the right eye 1/20 and in left eye 20/20, with normal intraocular pressure. Slit lamp exam showed no signs of inflammation; fundus examination revealed superior retinal oedema (Figure 1). The patient was referred for optical coherence tomography (OCT) that demonstrated multiple areas of serous neuroretinal detachment, retinal oedema and intraretinal cysts, most prominent in the macular area (Figure 2).

Visual field investigation shows blind spot enlargement.

The gynecological examination revealed normal external genitalia and ovaries of normal size and mobility on bimanual palpation. Transvaginal ultrasound showed normal appearing, homogenous myometrium, endometrial thickness of 4 mm, both ovaries with multiple small follicles (>15 follicles), distributed peripherally ('string of pearls' appearance) (Figure 3).

The blood analysis was normal. All the examinations (internal, dermatological, ear nose, throat and neurological) presented no evidence of neurological deficit, as well as the cerebral magnetic resonance imaging.

The presumed diagnosis, based on ocular exam and OCT findings, was Vogt-Koyanagi-Harada disease. The patient had steroid treatment started as pulse therapy with 1g/day methyl-prednisolone infusion, followed by oral 0.8 mg/kg/day methyl-prednisolone and topical administration. Four we-

Received: *April 19, 2013*
Revised: *May 14, 2013*
Accepted: *July 05, 2013*

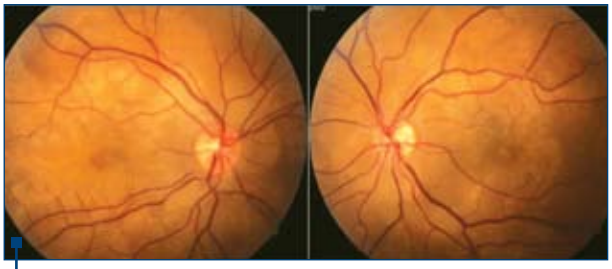

Figure 1. ***Fundus examination***

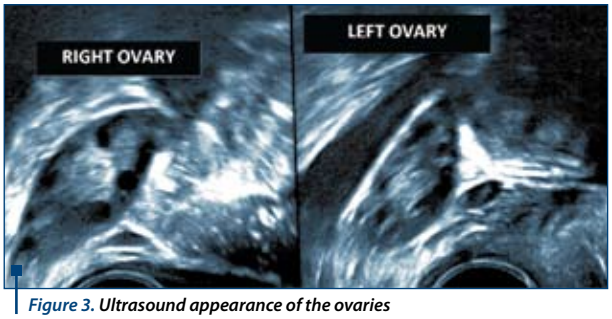

Figure 3. ***Ultrasound appearance of the ovaries***

eks after the initiation of steroid therapy, visual acuity was 20/20 in both eyes, with normal aspect of ocular fundus (Figure 4), showed also by OCT scans (Figure 5); retinal thickness reached normal range values. During the steroid treatment, the intraocular pressure increased (40 mmHg in the right eye and 35 mmHg in the left eye), probably due to the steroid responders reaction. After general and local antihypertensive treatment, the intraocular pressure normalized, with values within normal range without local treatment.

Discussion

Autoimmunity is well documented in the pathogenesis of Vogt-Koyanagi-Harada disease. Although PCOS is associated mainly with metabolic impairment, high levels of auto-antibodies (anti-histone, anti-double stranded deoxyribonucleic acid) were also found in clinical trials[4,5]. However, their significance in PCOS pathogenesis is still unclear.

The metabolic syndrome with impaired glucose tolerance is another characteristic shared by both conditions, with important role in PCOS pathophysiology and increased prevalence in Vogt-Koyanagi-Harada disease[6,7].

Conclusions

The concomitant presence of the two features in the same patient suggests a common autoimmune pathogenic mechanism. However, PCOS could be only incidental in this patient with Vogt-Koyanagi-Harada disease, considering that PCOS is a frequent condition with prevalence between 6-12% in general population. ■

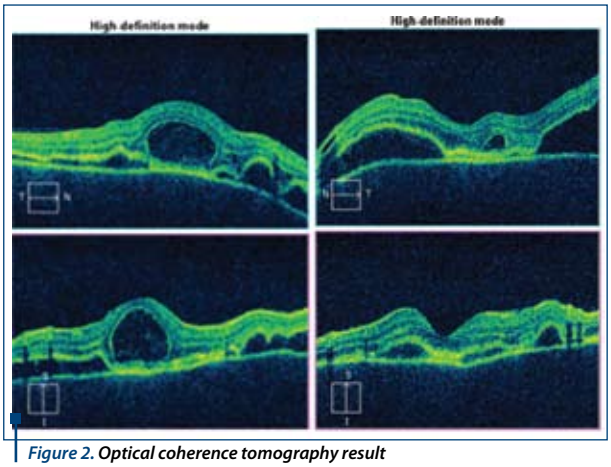

Figure 2. ***Optical coherence tomography result***

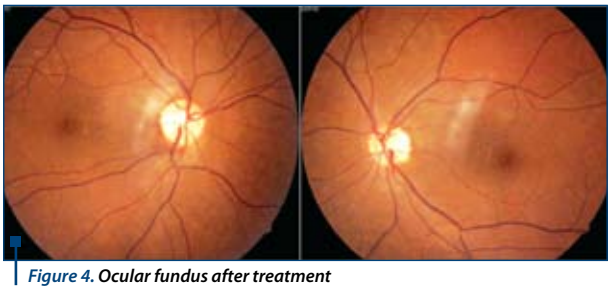

Figure 4. ***Ocular fundus after treatment***

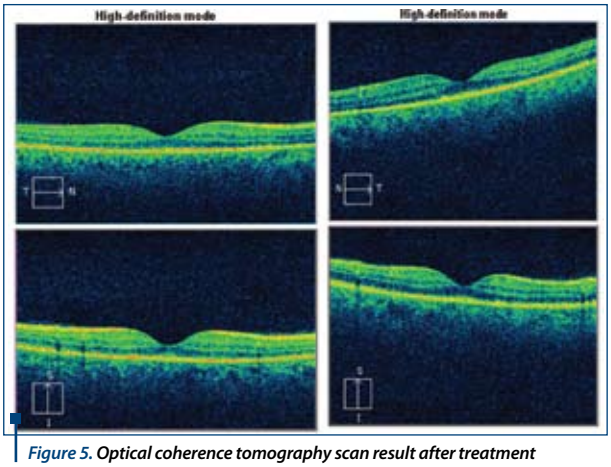

Figure 5. ***Optical coherence tomography scan result after treatment***

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