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Presumed Idiopathic Central Serous Chorioretinopathy in a 12-Year-Old Girl

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Key Words

Central serous chorioretinopathy, idiopathic · Fluorescein angiography · Optical coherence tomography · Serous retinal detachment

Abstract

Idiopathic central serous chorioretinopathy (CSC) typically affects middle-aged males. To date, only one case of idiopathic CSC in a prepubertal subject has been reported. Atypical idiopathic CSC presentation may be challenging to diagnose. Exclusion of secondary causes of serous retinal detachment (SRD) is warranted. We describe the atypical case of a 12-year-old female with a circumscribed SRD that resolved spontaneously and with fluorescein angiography (FA) findings that were compatible with idiopathic CSC. Optical coherence tomography (OCT) and systemic assessment were performed to exclude other etiologies. FA demonstrated multiple focal leaks in early phases, with subretinal leakage and pooling in late phases. OCT showed a localized circumscribed retinal detachment. Complete blood count was within normal limits. Serum cortisol was normal (22.1 µg/dl) and mean arterial blood pressure was 100/60 mm Hg, thereby excluding secondary causes of CSC. This is the second reported case of idiopathic CSC in a prepubertal female and the first one documented by FA and OCT, as well as other studies to exclude secondary causes. Albeit rare, idiopathic CSC should be considered in the differential diagnosis of SRD in this (prepubertal) age group, after excluding secondary ocular or systemic etiologies.

Introduction

Central serous chorioretinopathy (CSC) is defined as a circumscribed neurosensory retinal detachment caused by focal retinal pigment epithelium (RPE) incompetence. It is generally a self-limiting condition, in which patients complain of metamorphopsia,

micropsia, relative central scotoma, and a moderate decrease of visual acuity [1]. Fluorescein angiography (FA) typically shows single or multiple ‘inkblot’ early leakage points at the RPE followed by progressive localized subretinal leakage [2]. Optical coherence tomography (OCT) shows a localized dome-shaped macular detachment, with a normal neurosensory retina.

CSC mostly affects middle-aged males with type A personality. Fine and Owens [3] reported the only case in the literature of CSC in a child. When the clinical presentation is typical, diagnosis is forthcoming, based on ophthalmoscopic and FA findings. In atypical patients, however, a diagnosis of exclusion and further examination are warranted.

Case Report

A healthy 12-year-old female presented with a 3-day history of acute-onset blurred vision in her right eye without any prodromal symptoms. She denied a previous history of medication, systemic or ophthalmic diseases. Initial best-corrected visual acuity was 20/40 in the right eye and 20/15 in the left eye. Anterior segment examination was normal in both eyes, without any signs of inflammation. Fundus examination of the right eye revealed a normal coloration of the optic nerve and a retinal elevation at the fovea and inferior macula secondary to subretinal fluid without any signs of intraocular inflammation (fig. 1a). Fundus examination of the left eye was normal.

Right eye OCT showed an accumulation of subretinal fluid which did not show an extension to the optic nerve head, excluding the presence of optic nerve pit (fig. 1b). Angiography demonstrated multifocal leakage at the level of the RPE at the papillomacular bundle region. Late phases of the FA showed subretinal leakage and pooling, with a very discrete hyperfluorescence of the optic nerve head (fig. 2). A B-scan ultrasound was performed and produced results within normal limits.

Based on these findings, we decided to observe this patient without any treatment or intervention. After three months, visual acuity spontaneously improved to 20/15 and fundus evaluation was without any abnormal findings. Repeated FA and OCT showed complete resolution of the multifocal leakage and subretinal fluid (fig. 3). The patient has been periodically checked for 30 months without recurrence or any signs of inflammation in either eye.

Discussion

CSC is a clinical entity affecting middle-aged patients, usually males, in whom clinical appearance and FA are usually sufficient to confirm the diagnosis [1, 4–6]. In patients outside the usual age range of this condition (20–50 years), other diagnoses have to be ruled out. Localized serous retinal detachment (SRD) in childhood could be secondary to inflammatory, neoplastic, or developmental conditions.

Infectious diseases or inflammatory conditions that produce multifocal choroiditis, which may simulate CSC, include presumed ocular histoplasmosis syndrome (POHS), punctate inner choroidopathy (PIC), and Vogt-Koyanagi-Harada (VKH) disease. POHS has characteristic ‘punched-out’ choroidal lesions in the mid-periphery and peripapillary atrophy, which were not present in our patient. PIC, considered to be one of the white dot syndromes (WDS), affects young and middle-aged myopic women (age range 15–55 years). PIC presents with blurred central vision and photopsias, and posterior pole examination reveals multiple (12–25) small (100–300 μm) opaque round lesions with an SRD occasionally overlying them. These lesions evolve into

atrophic chorioretinal scars within months [7]. The focal leaking points in our patient healed without any residual scarring. Other WDS include multifocal choroiditis with panuveitis, multiple evanescent WDS, acute multifocal posterior placoid pigment epitheliopathy, and birdshot chorioretinopathy. All of these have characteristic choroidal lesions and are mostly associated with a degree of anterior chamber inflammatory cells and vitritis, which were not associated with the lesions in our patient [7]. Based on the multifocal leakage at the site of the SRD and the discrete hyperfluorescent appearance of the optic nerve head on the late phases of the angiogram, the most important differential diagnosis is VKH disease. VKH disease can have very different clinical manifestations depending on the stage of the disease [8]. There are some reports occurring in children, and this group has been estimated to represent 3% of all patients with VKH disease [9, 10]. In our case, in contrast to the classic FA findings in CSC, a late-phase, very discrete hyperfluorescence of the optic nerve head was found. However, the course of the disease seen in this case was incompatible with any presentation of VKH disease, since all patients with VKH disease present more than one sign of inflammation. Furthermore, VKH disease always requires treatment with no reported spontaneous resolution present in any case [11–15]. Clinically observable chorioretinal pigmentary changes follow healing of the leakage spots, while in our case, no residual fundus changes were noticed after spontaneous resolution of the subretinal fluid.

Optic nerve pit, which can manifest with serous macular detachment that communicates with the optic nerve, was not clinically detected in this patient. Moreover, our patient showed multifocal leakage on FA which is not associated with optic pit maculopathy [13]. Another cause of fluid at the macular area is X-linked retinoschisis – this was ruled out since it is a bilateral condition that only affects males. Hemopoietic or intraocular neoplasia was excluded clinically and by complete blood count as the etiology of SRD in this patient.

In a report on 312 cases of CSC [1], it was demonstrated that systemic steroid use, pregnancy, stress and type A personality were risk factors as shown in previous studies [4, 5]. In a study of CSC in women only [6], it was reported that a higher than average age at onset, namely 53 years (range 40–60 years), and high levels of cortisol play important roles. An important cause-effect relationship between serum cortisol levels and CSC presentation has been established [14]. In the present case, cortisol was normal (22.1 µg/dl, normal range 6.99–25 µg/dl) [15]. Another contributing risk factor is high mean arterial blood pressure [4, 5]. Our patient had a mean arterial blood pressure of 100/60 mm Hg (measured twice/day for 1 week).

In conclusion, based on the clinical appearance and the natural course of the disease, this case is compatible with CSC. To our knowledge, this is only the second case of a presumed idiopathic CSC in a prepubertal female and the first one documented by FA and OCT, as well as other studies to exclude secondary causes. CSC may affect females at a young age and should be included in the differential diagnosis of multifocal localized SRD in that age group.

Disclosure Statement

None of the authors have any financial/conflicting interests or funding sources to disclose. The content of this case report has not been published or submitted for publication elsewhere nor has it been presented at any conference.

Fig. 1. **a** Red-free photograph showing SRD extending from the foveal region to the inferior vascular arcade. **b** OCT showing accumulation of subretinal fluid causing a circumscribed area of SRD not communicating with the nerve.

Fig. 2. **a** Early-phase FA showing multifocal leakage through the RPE. **b** Late-phase multifocal subretinal pooling and a very discrete hyperfluorescence in the optic nerve head is noted.

Fig. 3. **a** Red-free photograph showing complete resolution of the SRD. **b** FA showing complete resolution of the multifocal leakage. **c** OCT showing complete resolution of subretinal fluid.

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