ATYPICAL SPECTRUM OF FUNDUS FLUORESCEIN ANGIOGRAPHY FEATURES IN CENTRAL SEROUS CHORIORETINOPATHY

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ABSTRACT

Background: Central serous retinopathy (CSR) is an idiopathic disorder of the macula characterized by serous elevation of the neurosensory retina at the posterior pole caused primarily by leaky choriocapillaries. On fundus fluorescein angiography (FFA), two typical patterns of this leakage are defined as inkblot and smoke stalk appearance. But atypical and unusual patterns also occur that can produce difficulty in diagnosis. **Objective:** To determine the spectrum of atypical presentations of CSR on FFA. **Patients and Methods:** All patients presenting with a clinical diagnosis of CSR and referred for FFA were included in this descriptive study. After taking consent, FFA was performed and the results were analyzed for atypical features of this disorder. Where FFA features were not conclusive an optical coherence tomography (OCT) was also performed to aid further evaluation. **Result:** 12 out of 23 patients had atypical FFA features. These include multiple inkblots in one eye, multiple inkblots in both eyes, ink blot and smoke stalk in the same eye, CSR with pigment epithelial detachment in which only the pigment epithelial detachment shows up on FFA, CSR with choroidal eovascularisation in which the CNV only shows on FFA and CSR with no leakage on FFA. **Conclusion:** Atypical presentations of CSR can cause diagnostic problems. The ophthalmologist must be aware of these features. Furthermore, these cases should be worked up for associated systemic / ocular disease.

Keywords: Atypical CSR, FFA, OCT

INTRODUCTION

Central Seruos Retinopathy (CSR) is an idiopathic exudative disorder of the macula where the neurosensory retina is detached from the underlying retinal pigment epithelium. The resultant cavity is usually filled with clear fluid. It was Donald Gass that first studied the FFA characteristics of CSR, establishing the underlying pathology to be active leakage into the subretinal space through the retinal pigment epithelium (RPE) from abnormally leaky choriocapillaries.

The two typical patterns of leakage in FFA in idiopathic CSR are described as inkblot appearance and smoke stalk appearance of leakage. However, atypical forms of CSR have also been described by onset at an older age, the presence of multiple and bilateral foci, more frequent recurrences and a poorer prognosis. Although an exact cause of CSR has not been proven, various disorders and conditions have been found to be associated with the development of CSR. These include disorders that increase the corticosteroid / catecholamine levels in the body, namely, type A personality, Cushing's syndrome, pregnancy etc.

CSR also has certain ocular associations like choroidal neovascularisation (CNV), bullous retinal detachment and optic disc pit. Moreover, although spontaneous recovery is the rule in the clinical course of CSR about 45% of cases are seen to recur. All these factors may alter the typical FFA appearance of CSR. The objective of this study was to determine the spectrum of atypical presentations of Central Serous Retinopathy (CSR) on fundus fluorescein angiography (FFA).

PATIENTS AND METHODS

This study was carried out in alliance in two clinical ophthalmic set ups; the department of Ophthalmology, Sheikh Zayed Medical College and Hospital in Rahim Yar Khan and the Amanat Eye Hospital in Rawalpindi. Both centers are equipped with state of the art FFA machinery and are the hub of peripheral referrals in their respective geographic areas. Duration of study was 12 months from 1st November, 2012 to 31st October, 2013.

Inclusion criteria: All patients with clinical diagnosis

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of CSR and who were referred for FFA were included in the study. Exclusion criteria: Renal failure, hypersensitivity with fluorescein.
Informed consent was taken from all patients prior to inclusion in this study. A detailed history was obtained from all patients of CSR. The demographic data was recorded and a clinical examination was carried out. They were then educated about fluorescein angiography and an informed consent was taken. FFA was performed and in atypical cases, an OCT was also performed for further evaluation. The data was entered and analyzed in SPSS version 15.

RESULTS

A total of 23 study subjects were included with mean age of 36 ± 5.1 years. 16 (69%) were male. Typical presentation of single inkblot (one eye) and single smokestalk (one eye) was present in 11 (48%) cases. Single inkblot in one eye was present in 6 (26%) and single smokestalk in 5(22%) of cases. Mean age of atypical presentation was 36 ± 4.7 years. Atypical presentations was found in 12 (52%) of cases with 8(67%) males. Break up of atypical presentation is shown in Table I.

Table I: Frequency of different presentations (N=23)

Presentation	Subtypes	No (%)
Typical 11 (48%)	Single inkblot in one eye	6 (26%)
	Single smoke stalk in one eye	5 (22%)
Atypical 12 (52%)	Inkblot and smoke stalk (same eye)	2 (8.6%)
	Multiple inkblots (same eye)	4 (17.3%)
	Multiple inkblots (both eyes)	3 (13%)
	CSR with PED	1 (4.3%)
	CSR with CNV	1 (4.3%)
	CSR with no leakage	1 (4.3%)

Figure I: Combined smokestalk and inkblot leakage in the same eye

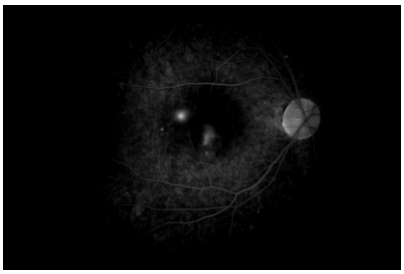


Figure II: Multiple leakage points in the same eye

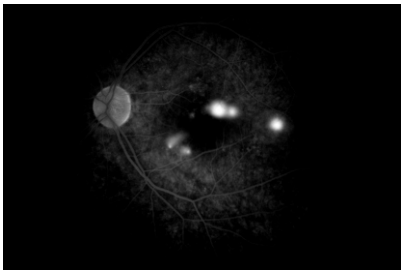
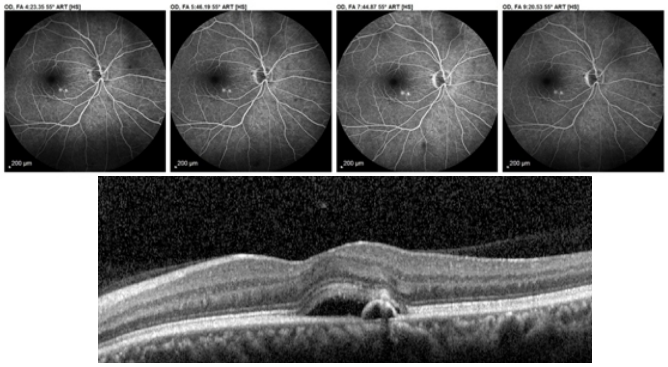


Fig. III. FFA features suggest PED alone where as OCT shows CSR along with PED



DISCUSSION

In a total of 23 cases, 70% were males and 30% were females. This is consistent with the known pattern of CSR's being more common in male individuals. The mean age of patients presenting with typical CSR on FFA was 36yrs with standard deviation of 4.7 yrs. The mean age of those presenting with atypical features on FFA was slightly higher (38yrs $\pm$ 4.5 yrs). Out of the two typical presentations of CSR the inkblot appearance was seen slightly more frequently (26%) than the smoke stalk appearance. This is consistent with a local study conducted by Imtiaz et al. and another study conducted in Srinagar.

Conflicting evidence as to which of the two typical appearances are more common are being reported from different parts of the world suggesting an influence of ethnicity and genetics on this FFA presentation. However, since this was not the primary objective on which our study was based, we can comment that the study group of our study is too small to aid or refute such evidence. What is however, more remarkable is that a very significant number of individuals presented with atypical features of CSR on FFA rather than a typical single leakage point (52% atypical vs. 48% typical). A possible explanation for this is the infrequent referral for FFA in cases of typical idiopathic CSR especially without any specific risk factors. Since the natural course of a classic CSR is spontaneous resolution in 90% of the cases, so ophthalmologists tend to feel reluctant to refer such patients for a costly and invasive investigation. So usually, the patients of CSR that we received in both the ophthalmic setups were those in which such angiogram was warranted due to any of the following: confusion in diagnosis, atypical features on clinical examination using the stereoscopic biomicroscopy or with accompanying atypical features like concomitant suspicion of choroidal neovascular membrane, a pigment epithelial detachment, a history of steroid intake or other known systemic association with CSR.

There is a common understanding among ophthalmologists and eye care providers that a typical patient of CSR is bound to have either an inkblot or a smoke stalk in his/her eye. During the course of this study, we have come across two cases in which inkblot and smoke stalk patterns were seen in the same eye. Now this really questions the relevance of discrimination of these patterns since both have the same pathology and that both can occur in the same eye (Fig. I)

Multiple inkblots have also been reported as an atypical presentation of CSR. In our study also, the bulk of atypical cases comprised multiple inkblots in either one eye or both. These were also those cases in which RPE changes were more commonly reported suggesting recurrences. This pattern indicates that a complicated underlying pathology may result in these atypical features which also makes it more likely for the CSR to recur as apparent by the RPE changes(Fig. II).

Various reports have been published where an atypical presentation of CSR is associated with systemic conditions/ disorders. There are reports in which multifocal CSR with underlying PED's have been investigated and found associated with Tuberculosis. It is also reported that systemic anti tuberculosis treatment resulted in the complete resolution of the serous macular detachment. In our setup, TB is epidemic, however, no local study was found in which such link was investigated. We report a case in which only a pigment epithelial detachment (PED) is seen on FFA. Subsequently, once the same patient underwent an optical coherence tomography (OCT) examination, he was found to have a concurrent CSR (Fig III).

Another case was identified which was silent on FFA with no leaking points, and on OCT a serous retinal detachment was found. We speculate that this was a CSR in remission where active leakage had ceased and the residual fluid in the subretinal space had not been completely absorbed.

There have also been reports of cases where other ophthalmic conditions may mimic an atypical CSR. Examples of adult vitelliform dystrophy have been cited to be misdiagnosed as atypical CSR in which further investigations e.g. electro oculogram (EOG) were performed only after the CSR's were found unresponsive to intervention.

Elias Reichel describes a central serous form of age related macular degeneration (AMD) stating that in this case, AMD can resemble a CSR on FFA. Indeed, during the course of our study we encountered some cases in which the appearance was so peculiar that we could not detect the presence of a serous retinal elevation on FFA. FFA features were suggesting the presence of a choroidal neovascular membrane (CNV). Only upon a subsequent OCT did it become apparent that there was a serous lift of the retina. Above are classic examples where a dynamic study like FFA was complemented by a static cross sectional analysis of the posterior pole by OCT.

Alternate treatments are now being investigated for atypical cases of central serous chorioretinopathy eg, transpupillary thermotherapy with some success. The limitation of our study is that we were not able to intervene in any kind to treat atypical cases and establish if any treatment option was more effective in countering this ocular condition.

Finally, our goal in this study was to establish atypical patterns of CSR that might be confusing to

the treating ophthalmologist. It was to give confidence to the clinician to look 'outside the box' and be aware that indeed CSR may present with a pattern other than a single smoke stalk and inkblot in one eye only. Our categorization of these atypical patterns is merely arbitrary and is only to introduce them as possibilities. Furthermore, we suggest that atypical features may mean atypical pathology and / or systemic associations and that such probabilities should be sought in unusual presentations.

CONCLUSION

Atypical presentations of CSR can cause diagnostic problems. The ophthalmologist must be aware of these features. Furthermore, these cases should be worked up for associated systemic / ocular disease. OCT can provide valuable support in evaluation of these atypical cases.

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