

Observations on Six Rare Cases of Chorioretinitis Sclopetaria

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ABSTRACT

Introduction: Chorioretinitis sclopetaria (CS) is a rare consequence of ocular injury. Its association with open globe injury (OGI) and retinal detachment (RD) is controversial. This study evaluates patterns of chorioretinitis sclopetaria and its association with open globe injury and retinal detachment.

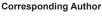
Case: This is an electronic review of records of a tertiary eye care institute of south India with descriptive analysis of six cases. Cases where fundus findings of chorioretinitis sclopetaria were available were included. Information regarding mode of injury, visual outcomes, follow up and causes of poor visual outcomes were obtained. Presence of open globe injury, retinal detachment, vitreous hemorrhage (VH) and orbital foreign body (FB) was also noted. Ultrasound scans of the eyeball were reviewed for presence of signs of chorioretinitis sclopetaria.

Observations: All the injured patients were male (age range 23-52 years). Bullet and blast injuries were the modes of injury. The duration since injury to the last follow up ranged from 0.25 to 12 years. The final visual acuity was <20/200 in 3/6 cases. Open globe injury and retinal detachment was noted in one case each, while 3/5 cases had orbital foreign body. Extensive facial and neuro-surgery were required in 2/6 cases. Sonography showed signs of chorioretinitis sclopetaria in 2/3 cases where scans were available for review.

Conclusions: Though manifestations of chorioretinitis sclopetaria evolve with time with visual improvement, final visual acuity is generally poor. Cases of chorioretinitis sclopetaria may have accompanying retinal detachment or open globe injury. Sonography should be evaluated with a high degree of suspicion for chorioretinitis sclopetaria in typical cases.

Key words: Chorioretinitis sclopetaria, Globe injury, Ocular blast injury, Ocular gunshot injury, Ocular trauma, Traumatic chorioretinal rupture..

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INTRODUCTION

Chorioretinitis sclopetaria (CS) is a rare entity, described variedly in literature. The term refers to a pathology where the choroid and the retina rupture following a "coup" injury due to non-penetrating trauma, resulting in fibrotic proliferation over the impacted area (Ludwig et al, 2019). Despite being described initially in the middle of the 19th century, less than 100 cases are known so far. Apart from its rarity, management of CS is complicated by varied presentations and injury mechanisms. The long term visual prognosis is largely poor (Ahmadabadi et al, 2010; Ludwig et al, 2019; Papakostas et al, 2013).

CS has been considered as a posterior manifestation of closed globe injury (CGI) by most previous authors (Ludwig et al, 2019; Papakostas et al, 2013, Williams et al, 1990). In most of the previously documented series on CS, RD was not considered an association of CS. This was reasoned due to the strong adhesions between the choroid and the retina at the borders of the CS lesions. However in 2014, 3 cases of RD with CS were described, thus raising serious concerns over long term follow up of CS (Ludwig et al, 2019; Papakostas et al, 2014). Another major controversy is the possibility of simultaneous presence of open globe injury (OGI). Though CS is associated with nonpenetrating trauma and traditionally discussed as CGI, there are several reports mentioning immediate management with globe exploration and corneal laceration repair (Ludwig et al,

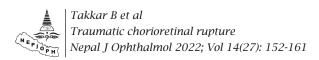
2019; Papakostas et al, 2014). While existing literature describes cases with CS, there is still poor understanding with regards to the similarities in its pathogenesis and sequelae, especially regarding RD and OGI.

The purpose of this series is to collate rare cases of CS and interpret the commonalities between them. Further, we evaluate the occurrence of RD and OGI in these cases, and explore the use of ultrasound in predicting its occurrence.

CASE

This is a retrospective, consecutive, noncomparative case series from review of electronic records of patients diagnosed as CS at our tertiary eye care center. The study is in accordance with the tenets of the Declaration of Helsinki. The protocol was reviewed and approved by the Institute Review Board (LEC BHR-R-05-20-447). Records were searched for cases where diagnosis of CS had been made. Cases where fundus picture was not present or where diagnosis had not been made by a retinal surgeon were excluded. Overall, 9 cases were identified in the period of 2011-May 2020, of which 3 were excluded.

Six cases were included in the study. General demography of all cases was noted, including mechanism of injury. Presenting and final visual acuities were recorded along with duration of last follow up since initial injury. Anterior segment and orbital features of trauma were identified. Presence of orbital foreign body (FB)



if any, was noted for all cases. Fundus findings and images were evaluated for location of CS, its clinical features and other relevant signs of trauma. Ultrasound B scan images of all patients were reviewed for presence of signs of CS and RD. For the six cases selected, all findings were tabulated (Table 1) for interpretation. Only descriptive analysis was performed due to low numbers.

RESULTS

Mean age of the sample was 32.6 ± 10.6 years (Range: 23-52). All were male, and left eye (LE) was injured in 5/6 cases. Bullet injury was the cause in 3 cases, while blast injury was involved in the rest of the 3 cases. The presenting visual acuity was better than 20/200 in 3/6 cases. Two patients had poor vision in the fellow eye related to blast injury. At last visit, the mean duration since injury was 2.7 ± 4.5 years (Range: 0.25-12). IOP was less than 10 mm Hg in 2 patients at presentation. Two patients maintained follow up after the primary injury, and visual acuity improved in both patients but remained below 20/20. The causes of vision loss in 5 patients with final visual acuity below 20/20 were corneal scar, hypotony, and neurological in one case each, and directly due to CS involving the posterior pole in 2 cases. OGI was noted in 1 case requiring urgent repair. VH was seen in 5/6 cases, while one case had localized RD. Five of the 6 cases had been evaluated extensively for orbital FB, which was determined in 3 of them. Two cases required extensive neurosurgical interventions. B-scan images were available for

review in 3/6 cases, and 2 of these showed signs of CS in retrospect (detailed in discussion).

Case 1: A 23-year-old male presented with a history of gunshot injury to LE 3 months back. Both eyes had visual acuity of 20/20, and anterior segments and IOP were normal. Retinal examination of LE revealed a large area of bare sclera with chorio-retinal adhesions at its borders along with retinal and pre retinal hemorrhages (Fig 1). Fellow eye was normal. A metallic FB was detected in the posterior orbit of the LE (Fig 2). Hence a diagnosis of CS was made. Ultrasound B scan of the eye was done for documentation and was also suggestive of CS (Fig 3, features discussed later). The patient was given the option of laser around the CS lesion but opted for follow up (Akhila & Takkar, 2020).

Case 2: A 33-year-old male presented to the clinic 1 day after repair of a paracentral inferior full thickness corneal tear following trauma with a plastic bullet to the left eye (LE). Visual acuity at presentation was counting fingers close to face. There was minimal subluxation of lens along with central vitreous hemorrhage with attached peripheral retina. There were no signs of orbital injury. Ultrasound B scan revealed only vitreous hemorrhage (VH) (Fig 3). The patient had been prescribed oral steroids by the primary surgeon. One week later vitreous had cleared centrally with only inferior residual hemorrhage. There was a 6X2 mm area of bare choroid with surrounding multi layered hemorrhage below the inferior arcade

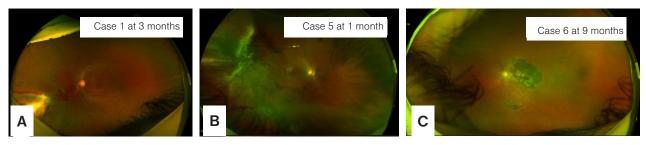


Figure 1: (A) Fundus photo of case 1 at 3 months follow up showing bare sclera with surrounding hemorrhages in nasal periphery. Pigmentation and fibrosis is also noticeable suggesting CS lesion. (B) Fundus photo of case 5 at 1 month of injury showing CS lesion in superior-temporal quadrant. Media is hazy due to resolving vitreous hemorrhage, but bare sclera can be seen with surrounding area of altered sub retinal pigmentation at its nasal border suggestive of self-absorbed retinal detachment (corroborating to location of sonography findings in Figure 3). (C) Fundus picture of case 6 at 9 months of injury showing CS lesion at macula with hypertrophic RPE and fibrosis.

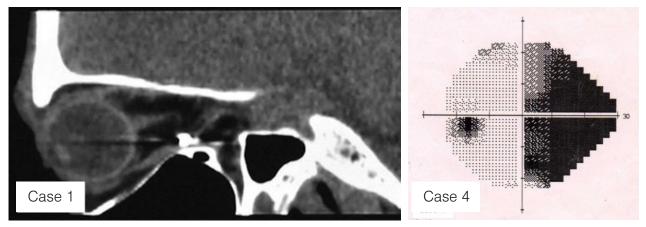
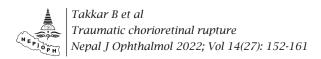


Figure 2: (Above) Orbital computerized tomography section of case 1 showing a metallic foreign body in posterior orbit with accompanying linear attenuation tract. (Below) Standard perimetry of case 4 showing hemianopia in the involved line respecting the vertical midline.

along with commotio retinae. The patient was diagnosed to have CS. The intraocular pressure (IOP) was normal. Three months into follow up there was suture related corneal infection which was managed with topical antibiotics. The patient remained in follow up for 2 years, with healing of the CS lesion along with fibrosis. The VH gradually settled inferiorly during follow up, and the eye gained a vision of 20/320. The vision loss was attributed to corneal scar. *Case 3:* A 23 year-old male presented with bilateral eye injury following a bomb blast 1 year ago. There was no history of orbital injury and orbital computed tomography (CT) scan was already done. He was referred as a case of peripheral RD in LE. Right eye (RE) was a prosthetic eye with history of OGI and history of some ocular surgery immediately after injury. LE had a visual acuity of 20/100, with normal anterior segment and IOP at presentation. On



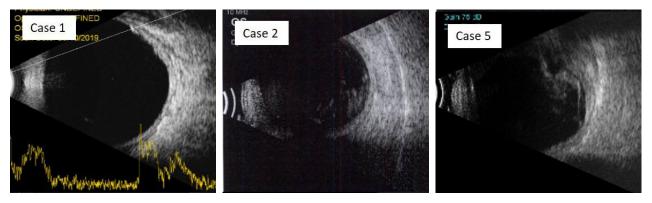


Figure 3: Sonographies of cases 1, 2 and 5 with transverse sections through the areas involved by CS. Case 1 shows a hyperechoic border (intersection of vector scan) corresponding to the chorioretinal adhesion followed anteriorly by a depression in the outer coats of the eye. Case 2 shows only vitreous hemorrhage with no signs of CS. Case 5 shows choroidal swelling with central hollow, overlying vitreous hemorrhage and membrane corresponding to the area of retinal detachment seen in Figure 1.

fundus examination, fibrous traction bands were noted all over the posterior pole along with macular atrophy and overlying epiretinal membranes. There was old VH inferiorly along with a lesion showing bare sclera and pigmented borders suggestive of CS. As per history, the vision was stable in LE, so surgical intervention was deferred and low vision aids were prescribed.

Case 4: A 46 year-old male presented for cosmetic rehabilitation of right orbit. He had sustained injury to both eyes following a bomb blast 12 years ago. He was detected to have multiple intracranial hemorrhages with bilateral orbital foreign bodies and fractures on CT scan. Subsequently he underwent immediate neurosurgical intervention and RE globe repair. He was detected to have nil vision in RE with LE hemianopia on visual field testing then (Figure 2), along with dense retinal and pre retinal hemorrhages in RE. At presentation, RE

had no perception of light, and LE had visual acuity of 20/40 and normal IOP. LE showed disc pallor and bare sclera with intense scarring at its edges in temporal peripheral fundus, suggestive of a CS lesion. The patient was advised regular monitoring and no surgical intervention was done.

Case 5: A 31 year-old male presented with low vision in RE due to air conditioner related blast injury. CT scan of brain and orbits showed no FB. RE had visual acuity of hand motions close to face, while LE was 20/20. An iris sphincter tear was detected along with hyphema measuring 2 mm and an IOP of 7 mm. Subluxation of lens was noted and B scan ultrasound done in the immediate period was reported to have VH (Figure 3). LE was normal. Medical management was prescribed with steroid eye drops. At the one month follow up, visual acuity had improved to 20/200, media had cleared and CS was noted in the superiortemporal quadrant along with localized minimal RD (Figure 1). Choroidal folds were also noted at the posterior pole though IOP became 10 mm Hg. At the final follow up 5 months after injury, visual acuity was stable though IOP was 7 mm Hg. Spontaneously attached RD and fibrotic membranes were noted.

Case 6: A 52-year-old male presented with a history of gunshot injury to LE and face 9 months ago. He had undergone multiple facial and neuro-surgeries for removal of FB, facial palsy and face repair. At presentation visual acuity in RE was 20/20, while LE visual acuity was counting fingers close to face. LE anterior segment and IOP were within normal limits. Fundus examination revealed bare sclera with hypertrophic pigmentation and scarring in the macular region of the LE suggestive of CS (Figure 1). RE was within normal limits. No intervention was done and monitoring suggested.

DISCUSSION

Ludwig et al reported 6 cases of CS and performed analysis on literature reported till last year (Ludwig et al, 2019). The authors found CS to be commoner in males (>80%), and gunshot or bullet related injury as the commonest mechanism (>60%). Around 65% of the cases had orbital foreign bodies, and nearly the same number present with VH. RD however was found to be rare and seen in around 10% of the cases only. Globe exploration for OGI was required in around 10% of cases in their analysis. The authors also reported macular and superior CS to be the most predictive of poor visual outcome. Although most patients showed initial improvement, over all less than 20% attained vision of 20/20 Most of our findings (Table 1) are consistent with the analysis by Ludwig et al (Ludwig et al, 2019).

Traumatic RD is a well-researched entity, with multiple causes and etiopathogenesis (Johnston, 1991; Williams et al, 1990). We found 1 of our 6 cases to develop RD. The CS was in superior-temporal quadrant in that case (Case 5). Traditionally, RD has not been considered a consequence or association of CS due to strong fibrotic adhesions at the edges of the ruptured retina, uncompromised vitreous cortex and healthy medullary vitreous in young patients (Martin et al, 1994). Earlier experimental studies have shown that rupture of the choroid and retina can occur without their separation following trauma where shear stress in more, and both can displace from initial position as a single unit (DeGuillebon & Zauberman, 1972; Papakostas et al, 2013). However, there are some rare reports showing occurrence of RD with CS. In the report by Martin et al, RD occurrence following CS was delayed in onset (1 year post trauma) and occurred due to breaks located at sites different from the CS. This prompted Martin et al to recommend initial observation for CS. Papakostas et al reported 3 cases of RD associated with CS, though all their 3 cases had undergone previous ocular or orbital surgery in relation to the trauma before RD actually

Case	Age (yrs)	Sex	Lat-erality	Mechanism of injury (duration)	Vision at present-ation	Vision at last Visit	Duration at last visit since injury (yrs)	IOP at First Visit (mmHg)	Cause of vision loss	OGI noted	RD	НЛ	Sign of CS on B scan*	FB in orbit
1	23	Male	Left	Gun shot (3 months)	20/20	20/20	0.25	10	-	No	No	Yes	Seen	FB noted, not removed
2	33	Male	Left	Plastic bullet (1 day)	CF 1/2m	20/320	2	very low	corneal scar	Yes	No	Yes	Not seen	-
3	23	Male	Left	Bomb blast (1 year)	20/100	20/100	1	12	macular atrophy with fibroses	No	no	Yes	-	No FB
4	34	Male	Left	Blast injury (12 years)	20/40	20/40	12	15	Brain or Post-chiasmal injury	No	no	Yes	-	Multiple FB, removed by NS
5	31	Male	Right	Air conditioner blast (1 day)	HM	20/200	0.5	7	Hypotony, choroidal folds	Suspected but absent	yes	Yes	Seen	No FB
6	52	Male	Left	Gun shot (9 months)	CF	CF	0.75	normal	Macular CS	No	no	no	-	Yes, removed by NS

Table 1: Description of cases with chorioretinitis sclopetaria.

CF: counting fingers, HM: Hand motions, CS: chorioretinitis sclopetaria, OGI: open globe injury, RD: retinal detachment, VH: Vitreous hemorrhage, FB: Foreign body, NS: Neurosurgeon *Interpretation done in retrospect

developed (Ludwig et al, 2019; Papakostas et al, 2014). Ahmadabadi et al also noted RD in 1 /13 cases in their series of CS, though that case had another retinal break adjacent to CS with vitreous detachment (Ahmadabadi et al, 2010). In our case 5 (Figure 1), RD appeared to be self-absorbed at 1 month when media was clear enough to allow fundus examination (Figure 1). The self-absorbed RD appeared to be adjacent to the nasal border of the CS, and in retrospect was also seen adjacent to the border of the CS on ultrasound (Figure 3). Since fibrosis and chorioretinal adhesion will take weeks to set in, it is possible that localized RD had occurred in our case initially but settled on its own with ensuing severe fibrosis well proven histologically in CS (Dubovy et al, 1997). Our findings, and those discussed previously by Papakostas et al, Ahmadabadi et al and Martin et al show the requirement of continuous and meticulous follow up as RD may develop in the immediate or even late period after injury, though it may not be directly linked to CS in some cases.

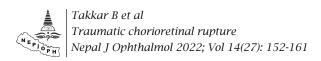
Kuhn's classification of trauma as OGI and CGI has gained wide acceptance as it has direct bearing on management perspectives

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(Kuhn et al, 1996). Traditionally, CS has been associated with CGI (Ludwig et al, 2019; Papakostas et al, 2013, Williams et al, 1990. In our series, 1/6 cases were found to have an OGI requiring urgent repair. The same has been noted by Papakostas et al in their series of CS (Papakostas et al, 2014). In the analysis by Ludwig et al, 11% of the cases had required globe exploration and corneal lacerations were also reported (Ludwig et al, 2019). However, in the series by Ahmadabadi et al, none of the 13 cases of CS had OGI (Ahmadabadi et al, 2010). Though it appears that OGI is not a common accompaniment of CS, presence of one should not rule out the other. This is important especially in blast injuries, 50% in our series. Blast injuries can result in simultaneous orbital and globe injury (Morley et al, 2010), and multiple projectiles may result in mixed features.

As most cases of CS have associated VH at presentation apart from other anterior segment findings, immediate clinical detection of CS is challenging. Ultrasound may be helpful as seen retrospectively in our series (Figure 3). Of the 3 cases where B scans were available for review, 2 showed signs of CS. Ultrasound done 3 months after injury in case 1 showed thickened edge at the site of CS, with an accompanying depression in the outer coats of the globe anteriorly. In contrast, sonography done immediately following trauma in case 5 showed choroidal thickening with surrounding echoes due to hemorrhage with discontinuous outer coats. Thus, findings on ultrasound will depend on timing of imaging in relation to injury. The ultrasound images should be observed carefully with high suspicion of CS in a typical setting of CGI with orbital foreign body in gunshot or blast injury. Other described findings on sonography are layered VH and intense choroidal swelling.^[1]

Both figures 1 and 2 depict crucial clinical findings. Though the images shown in figures are of different cases, it can be seen that immediately following trauma CS may be difficult to realize due to presence of concomitant VH and edema (Case 5). With healing, the hemorrhages will resolve and bare sclera and chorio-retinal adhesion will become clearly visible with typical appearance of CS (Case 1). In the long term, fibrosis will be more evident with pigmentation at the borders of the CS (Case 6). Dubovy et al studied an eye previously diagnosed to have CS 20 years ago in a post-mortem clinic-pathologic correlation. ^[9] The authors showed loss of neural tissue in macula (as clinically in Case 3 of our series), hypertrophy and hyperplasia of RPE, defect in the choroid and retina, with dense fibrous connective tissue (as clinically in Case 3 and 6 of our series).^[9] Similar findings have also been summarized by Ludwig et al and Papakostas et al.^[1,2] Unfortunately, we could not record serial fundus images of all cases to ascertain this clinic-pathologic correlation. Figure 2 depicts the essence of orbital and neurological trauma that may accompany CS due to the mechanism of injury. Two of our cases had required serious



facial and neurosurgeries, while 3/6 had definite signs of orbital trauma (Table 1). These findings may overshadow the presentation of CS and may require earlier management and attention soon after injury, and may also have direct impact on the visual outcomes independent of CS lesion (Case 4 of our series).

CONCLUSION

CS is a rare occurrence following ocular and orbital trauma. The manifestations of CS will evolve over time as seen in our series, but vision loss is commonly significant and irreversible. Though most previous literature documents CS as an association of CGI which does not lead on to RD, we found CS to be seen in a case each of OGI and RD. As RD is a rare possibility in cases with CS, long term surveillance and appropriate patient counseling is crucial. We also found immediate and late signs indicative of CS on ultrasound, which should be kept in the correct perspective while dealing with injuries typically associated with CS.

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