Original Research Article

A prospective study of clinical profile of iridocorneal endothelial syndrome in a tertiary care hospital

M.Ramesh Chandra[1], P.Sanjeeva Kumar[2*]

Associate Professor, Department of Opthalmology, Santhiram Medical College, Nandyala, Andhra Pradesh, India

Received: 28-10-2020 / Revised: 13-11-2020 / Accepted: 23-12-2020

Abstract

Introduction: Irido-Corneal Endothelial (ICE) syndrome is an uncommon clinical entity which includes corneal endothelial damage, obstruction of anterior chamber angle, iris atrophy and polycoria.1 these proliferative and structural abnormalities are the hallmark of this disease. Corneal decompensation and glaucoma are the commonest causes of vision loss in patients with ICE syndrome.**Materials and Methods:** All patients diagnosed with ICE syndrome as mentioned in our inclusion criteria diagnosed between the October 2019-October 2020. A total of 50 patients were included in our study after ruling out exclusion criteria and the data was retrospectively analysed. Data pertaining to age, gender, clinical spectrum of disease, laterality, Visual acuity, corneal and iris findings, anterior chamber (AC) depth, gonioscopic findings, reaction in AC, association with glaucoma, medical and surgical management was collected, number of AGMs used, duration of follow up collected.**Results:** We studied 51 eyes of 50 patients with age range of 7-65 years with median age at presentation was 40.72 years. 28 (56%) were female and 22 (44%) were male. ICE syndrome was unilateral in 49 (98%), of which right was involved in 25 (50%) and left was involved in 24 (48%). Mean IOP of involved eye was 23.70± 11.7 mm of Hg. PIA and CS variant of ICE syndrome were equal in frequency of 20 (40%). CRS was found as least variant of ICE syndrome and was 10 (20%) patients. Out of 51 eyes of study group we had 43 (84.3%) eyes were phakic and 7 (13.7%) eyes were pseudophakic. There was no aphakic eye in our study group patients. **Conclusion:** In our study, ICE syndrome was unilateral, and middle-aged females were more commonly affected. Glaucoma is strongly associated with ICE syndrome. Surgical management is required in majority of the affected patients. ICE patients may require more than 1surgery for IOP control. They are commonly associated with corneal complications which may require penetrating keratoplasty.

Keywords: ICE, PIA, CRS, pseudophakic.

Introduction

Irido-Corneal Endothelial (ICE) syndrome is an uncommon clinical entity which includes corneal endothelial damage, obstruction of anterior chamber angle, iris atrophy and polycoria.[1] These proliferative and structural abnormalities are the hallmark of this disease. Corneal decompensation and glaucoma are the commonest causes of vision loss in patients with ICE syndrome[1].

**Correspondence*
Dr. P. Sanjeeva Kumar
Associate Professor, Department of Opthalmology, Santhiram Medical College, Nandyala, Andhra Pradesh, India.
E-mail: drsanjiv77@gmail.com

The aetiology of ICE syndrome is still largely unknown. Inflammation at the level of corneal endothelium due to viral aetiology like Herpes simplex or Epstein-Barr virus and a uveitic pathology have been described earlier. High percentages of HSV DNA has been found in the endothelial cells of ICE patients compared to controls[2]. It is considered to be sporadic, usually unilateral and diagnosed in young adults, females and occasionally in children. Syndrome is typically presented with diminution of vision in the morning and change in the shape and position of pupil [3]. The "membrane theory" was used in 1978 by Campbell to explain the pathogenesis of ICE syndrome. They hypothesized that corneal endothelial cell can proliferate and may develop structural defects

with the ability to migrate causing the changes noted in ICE syndrome[4].Similar to Fuchs' dystrophy, the slit lamp picture in ICE shows "Hammer silver" or "Beaten bronze" appearance of the endothelium. Also, Iris changes (such as heterochromia, ectropion uveae, corectopia, whole formation, and iris atrophy) are seen on slit-lamp examination. Gonioscopic examination may show high PAS extending above Schwalbe line, which is pathognomonic for ICE syndrome. Slit lamp examination, gonioscopy, tonometry, specular microscopy and confocal microscopy form an essential part of the examination framework used to diagnose the subclinical variants of ICE syndrome that may manifest itself asymptomatically in the unaffected eye[5,6].

Materials and methods

All patients diagnosed with ICE syndrome as mentioned in our inclusion criteria diagnosed between the October 2019-October 2020. A total of 50 patients were included in our study after ruling out exclusion criteria and the data was retrospectively analysed. Data pertaining to age, gender, clinical spectrum of disease, laterality, Visual acuity, corneal and iris findings, anterior chamber (AC) depth, gonioscopic findings, reaction in AC, association with glaucoma, medical and surgical management was collected, number of AGMs used, duration of follow up collected.Complete slit lamp biomicroscopy, applanation tonometry, four mirror gonioscopy, dilated fundoscopy were performed to determine corneal changes, iris findings, AC reaction, PAS, intraocular pressure and Disc changes. In some patients, presence of severe corneal oedema obviated the performance of specular microscopy and PAS assessment. For diagnosing glaucoma Humphrey Visual field test was done in presence of clear cornea and sufficient vision.

Inclusion Criteria

ICE syndrome is defined on the basis of characteristic findings of beaten-bronze or hammered-silver appearance of endothelium, iris atrophy with hole and iris nodules with or without PAS.

- IOP >21 mm of Hg or </= 21mm of Hg with AGM/prior Glaucoma filtration surgery (GFS) with optic nerve head changes characteristic of glaucoma was a criterion to diagnose glaucoma in the presenting patients.
- Secondary ocular hypertension (OHT) was also included as glaucoma patients in our study.
- Patients were classified as CS, PIA and CRS on the basis of normal or mild iris atrophy with predominant corneal involvement, iris atrophy with whole formation, iris nodules respectively.
- Cornea was graded as clear oroedematous.
- PAS was graded as mild (<90), moderate (90 to 180), severe (>180-270) and extensive (>270).

Exclusion Criteria

- Axenfeld-Reiger syndrome
- Fuchs endothelial dystrophy.
- Uveitis, traumatic angle closure.
- Other primary and secondary angle closure glaucoma were excluded.

Statistical Analysis: We undertook a retrospective analysis of all study patients and obtained data pertaining to clinical characteristics. Since the data was non-parametric, we used chi squared test and student T test to analyse the data. ANOVA was also performed to look for variance. Analyses were conducted using Stata Corp 97: Stata Statistical Software Release 5 (Stata Corporation, USA) and StatXact version 2 (Cytel Software Corporation, Cambridge, MA, USA).

Results

We studied 51 eyes of 50 patients with age range of 7-65 years with median age at presentation was 40.72 years. 28 (56%) were female and 22 (44%) were male. ICE syndrome was unilateral in 49 (98%), of which right was involved in 25 (50%) and left was involved in 24 (48%). Mean IOP of involved eye was 23.70± 11.7 mm of Hg. PIA and CS variant of ICE syndrome were equal in frequency of 20 (40%). CRS was found as least variant of ICE syndrome and was 10 (20%) patients. Out of 51 eyes of study group we had 43 (84.3%) eyes were phakic and 7 (13.7%) eyes were pseudophakic. There were no aphakic eye in our study group patients. [Fig 1]44 (88%) eyes were diagnosed to have glaucoma at presentation, 1 (2%) eye was diagnosed with glaucoma during follow up.Out of 45 patients who all diagnosed as glaucoma at presentation or during follow up 20 (44.4%) patients were treated with AGMs and remaining 25(55.5%) patients were treated as glaucoma filtration surgery (GFS). [Fig 2] Mean no of AGMs used were 2.44± 1.3. Out of 25 patients treated with GFS, 20(44.4%) eyes undergone trabeculectomy and 5 (11.1%) undergone Ahmed glaucoma valve. 8(17.7%) eyes had previous GFS. [Fig 3] Number of eyes had Diode CPC were 4 (8.8%). 6 (13.3%) eyes were undergone more than 1 surgeries to control the IOP during the follow up. We have found in our study that median age of patients undergone medical and surgical management of glaucoma was 44.53 years (range 7-56) and 46.26 years (range 13-60). (Table 1)

Table 1: Clinical Profile of Ice Syndrome Patients

Gender	N (%)
Male	22 (46%)
Female	28 (56%)
Laterality	
OD	25 (50%)
OS	24 (48%)
OU	1 (2%)
Mean IOP (mm of hg)	23.70±11.7
Mean no. of anti glaucoma medications	2.44±1.3
Lens status of eyes	
Phakic	43 (84.3%)
Pseudophakic	7 (13.7%)
Aphakic	0
Follow up period (months)	8.4±6.4

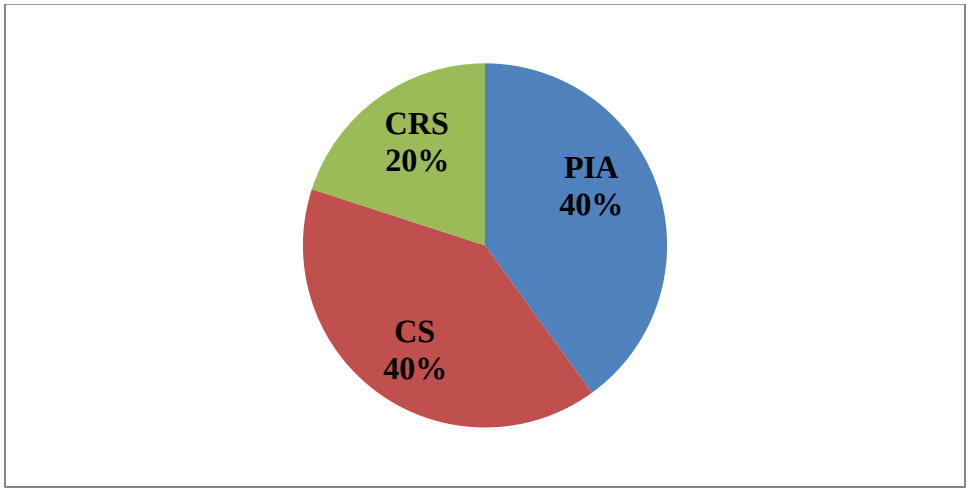

Fig 1: Variants of ice syndrome

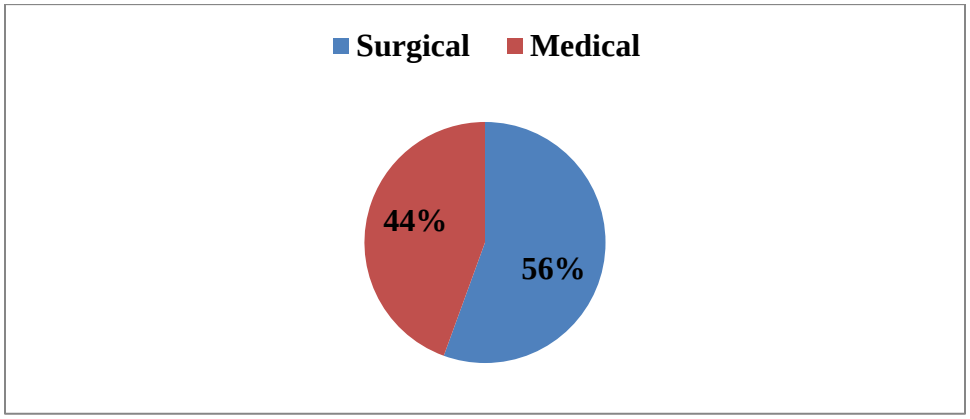

Fig 2: Treatment

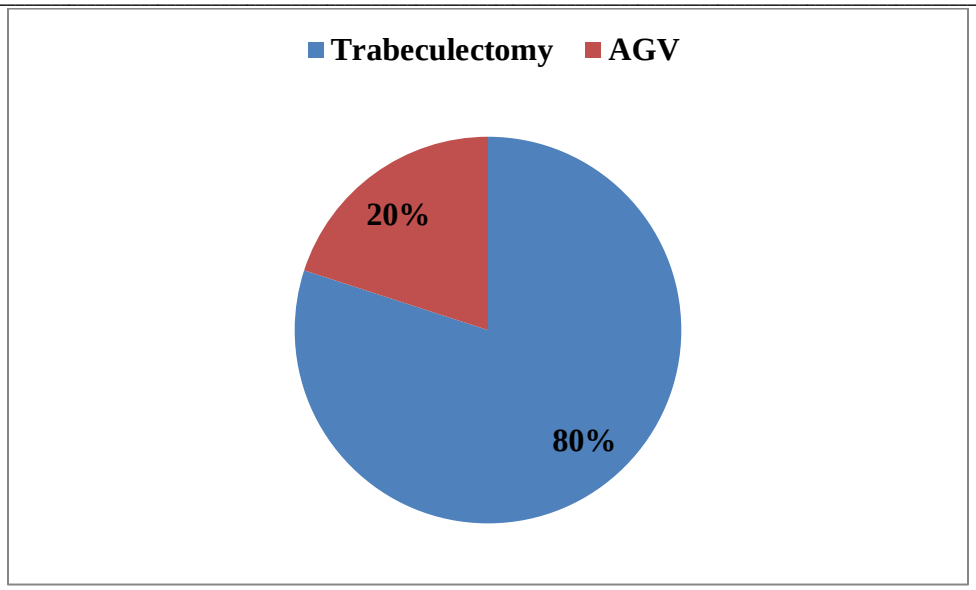

Fig 3: Type of surgical management

Discussion

In our study PIA and CS variants are of equal frequency (40%). CRS (20%) was the least common variant of ICE syndrome which was similar to Chandran et al in which PIA (52%) was the predominant variant followed by CS and the least was CRS. [13]. Laganowski et al case series also showed the similar frequency.

Wilson et al[10] reported that Cogan-Reese was the commonest in Caucasian population, whereas Teekhasaenee et al reported that Cogan-Reese syndrome was common in the Thai population [14]. This is suggestive of the existence of an ethnic variation in the presentation of the disease [7].

ICE syndrome is typically unilateral in presentation. In our study 98% of the patients were having unilateral involvement, which was similar to previous studies also. Glaucoma is strongly associated with ICE syndrome. The prevalence of glaucoma in our study was 90%. It ranges from 46% to 82%. In our study 44.4% of glaucoma patients were managed with antiglaucoma medications. In other published series it ranged from 12-40%[8].These differences can be possibly explained by the variations in the severity of the disease and the evolution of anti-glaucoma medications over time. Chandan et al studied a case series of 202 patients and found an inter eye variance in the presentation of ICE syndrome[9].

Unilaterality was exhibited in 183 (90%) subjects, and bilaterality in 20 (10%) subjects. Progressive iris atrophy (115; 52% eyes), Chandler syndrome (87; 39% eyes) and Cogan-Reese syndrome (21; 9% eyes) were found in decreasing order of frequency. 156 eyes (70%) had glaucoma at initial presentation and the average intraocular pressure in glaucomatous eyes was 24 (16, 38) mm Hg. Glaucoma filtration surgery for IOP control was required in our study was 55.5% patients[10].

Conclusion

In our study, ICE syndrome was unilateral, and middle-aged females were more commonly affected. Glaucoma is strongly associated with ICE syndrome. Surgical management is required in majority of the affected patients. ICE patients may require more than 1 surgery for IOP control. They are commonly associated with corneal complications which may require penetrating keratoplasty.

References

1. Glaucoma. Basic and Clinical Science Course (BCSC). American Academy of Ophthalmology 2010-2011:142-4.
2. Sacchetti M, Mantelli F, Marenco M, et al. Diagnosis and management of iridocorneal endothelial syndrome.Biomed Res Int. 2015; 2015:1-9.
3. Campbell DG, Shields MB, Smith TR. The corneal endothelium and the spectrum of essential iris atrophy. Am J Ophthalmol1978; 86(3):317-24.

4. Wilson MC, Shields MB. A comparison of the clinical variations of the iridocorneal endothelial syndrome. Arch Ophthalmol 1989; 107(10):1465-8.
5. Eagle RC, Font RL, Yanoff M, et al. Proliferative endotheliopathy with iris abnormalities.The irido corneal endothelial syndrome. Arch Ophthalmol. 1979; 97(11):2104-11.
6. Chandler PA. Atrophy of the stroma of the iris, endothelial dystrophy, cornealoedema, and glaucoma.Trans Am Ophthalmol Soc1955; 53:75-89.
7. Chandran P, Rao HL, Mandal AK, et al. Glaucoma associated with iridocorneal endothelial syndrome in 203 Indian subjects. PloS One 2017; 12(3): e 0171884.
8. Levy SG, Kirkness CM, Moss J, et al. The histopathology of the iridocorneal-endothelial syndrome. Cornea 1996; 15(1):46-54.
9. Shields MB, Campbell DG, Simmons RJ. The essential iris atrophies.Am J Ophthalmol 1978; 85(6):749-59.
10. Kim DK, Aslanides IM, Schmidt CM, et al. Long-term outcome of aqueous shunt surgery in ten patients with iridocorneal endothelial syndrome. Ophthalmology 1999; 106(5):1030-4.

Conflict of Interest: Nil
Source of support:Nil