Prevalence of Idiopathic Polypoidal Choroidal Vasculopathy (IPCV) in Thai Population Diagnosed with Choroidal Neovascularization (CNV)

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Background: Idiopathic polypoidal choroidal vasculopathy (IPCV) manifestation resembled those with neovascular age-related macular degeneration (AMD). Previous studies considered, IPCV was more common than neovascular AMD. This multicenter study thus intended to study IPCV in Thailand.

Materials and Methods : The medical records, fundus photos, spectral domain optical coherence tomography (OCT), fundus fluorescein angiograms (FFA) and indocyanine green angiograms (ICGA) on a series of 140 patients that were referred to ten referral centers all over Thailand were reviewed retrospectively.

Results : Of 140 patients with the clinical signs of CNV, 100 patients were diagnosed with IPCV (71.43 %), 16 patients (11.43%) were idiopathic CNVs including classic and occult forms, 12 patients (8.57%) were AMDs, 6 patients (4.29%) were Chronic central serous retinopathy (Chronic CSR), 2 patients (1.43%) were serous pigment epithelial detachment (serous PED), 2 patients (1.43%) were macroaneurysm, one patient (0.71%) was retinal angiomatous proliferation (RAP) and one patient (0.71%) was retinal pigment epithelial rip (RPE rip). Of the 107 eyes with IPCV, 79 eyes (73.83%) were found in macular area, 14 eyes (13.08%) were found temporal to vascular arcades, 10 eyes (9.35%) were found in the peripapillary area and 4 eyes (3.74%) were found both in macular and peripapillary region. We also found that IPCV was found in the same age group as other Asian studies. However, in our study, there is no sex predilection.

Conclusion : : IPCV is the common macular disease in patients with CNV in Thai population. Moreover, IPCV in Thai population is mostly found in macular area. No sex predilection was found.

Keywords : Idiopathic polypoidal choroidal vasculopahty, prevalence, choroidal neovascularization, age-related macular degeneration

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Proceeding

Introduction

In the past three decades, idiopathic polypoidal choroidal vasculopathy (IPCV) has gained its popularity among ophthalmologists worldwide as one of the new exudative maculopathies. The abnormality is believed to originate in the inner choroid.[1] The exact pathogenesis remains unclear ;however, the pathologies involve several thin-walled aneurysmal vessels which turn out to be venules in origin.[2-4] The vascular stasis which causes local edema and degeneration of choroidal tissues has been hypothesized by some that found clusters of dilated, thin-walled vessels surrounded by macrophages and fibrins.[5,6] The characteristics of IPCV usually has two compositions:

(1) dilated branching venous networks (BVNs) and (2) multiple terminal aneurysmal dilatation at the end of the venous networks.[7-11] Clinically, the signs include the presence of dilated reddish-orange, bulging polypoidal lesions in the macular and peripapillary area.[12] Also, the patients have recurrent subretinal hemorrhage, subretinal fluid and subretinal lipid which mimic the age-related macular degenerations (AMDs) and other diseases characterized by the presence of choroidal neovascularizations (CNVs).[13-16] IPCV lesions are distinctive with the use of indocyanine green angiography (ICGA) and optical coherence tomography (OCT).[17-23]

The prognosis of IPCV patients is usually better than that of AMD if proper treatment has been achieved. From several previous studies, photodynamic therapy (PDT) with verteporfin has shown to be effective.[24-38]

To the best knowledge of authors, by far there is no data about prevalence of IPCV and its characteristics in Thailand. Accordingly, this retrospective study is aimed to establish the prevalence and characteristics of IPCV in our country.

Objective

Primary objective : to study the prevalence of Idiopathic Polypoidal Choroidal Vasculopathy (IPCV) in Thai Population Diagnosed with Choroidal Neovascularization (CNV) Secondary objective : to study the characteristics of IPCV in Thai population.

Materials and Methods

Written approval for our study was granted by Khon Kaen University Ethics Committee in Human Research University, Thailand (Reference No. HE551279)

In this multicenter descriptive study, we reviewed retrospectively the medical records, fundus photos, spectral domain optical coherence tomography (OCT), fundus fluorescein angiograms (FFA) and indocyanine green angiograms (ICGA) on a series of 140 patients that were referred to ten referral centers all over Thailand from March 2013 to December 2013.

Our 10 eye centers are seven centers in the middle part and three centers in the peripheral parts of Thailand. The seven middle centers are Siriraj hospital, King Chulalongkorn Memorial hospital, Ramathibodi hospital, Rajavithi hospital, Thammasat university hospital, Pramongkut hospital and Mettapracharak hospital. The three peripheral centers are Songklanagarind hospital in Southern part, Maharaj Nakorn Chiangmai hospital in the Northern part and Srinagarind hospital in the Northeastern part of Thailand.

We included the patients with clinically subretinal or sub-pigment epithelial blood, subretinal fluid, subretinal lipid, subretinal pigment ring, irregular elevation of pigment epithelium, subretinal gray-white lesion, cystoid macular edema and sea fan pattern of subretinal small vessels. All patients must have had complete ocular history and examination with slit-lamp microscopy and posterior fundus examination. Fluorescein angiography with 5 ml of 10% Sodium Fluorescein and indocyanine green with 2 ml of 25 mg intravenously had been done.

We excluded the patients who had not been examined with FFA and ICGA, patients who had previously performed with any kinds of ophthalmic laser including focal retinal laser, PDT and transpupillary thermotherapy. Patients with angioid streak, pathologic myopia, ocular histoplasmosis, rhegmatogenous retinal detachment, tractional retinal detachment, uncontrolled glaucoma, retinal pigment epithelial tear and macular hole were also excluded. Patients whose eyes had been operated within 60 days prior to study were also excluded.

The patients were diagnosed with IPCV by angiographic findings as follows : (1) hypofluorescent halos around lesion (in the first 6 minutes) (2) polyp-like swelling (3) pulsation (can be observed by using video ICGA) (4) branching vascular networks of inner choroidal vessels (5) submacular hemorrhage and (6) late phase staining in the area of earlier hypofluorescent lesion

The prevalence equals to the ratio of patients diagnosed with IPCV over patients with clinically subretinal or sub-pigment epithelial blood, subretinal fluid,

subretinal lipid, subretinal pigment ring, irregular elevation of pigment epithelium, subretinal gray-white lesion, cystoid macular edema and sea fan pattern of subretinal small vessels. The 95% confidence interval also was calculated.

The sex in the IPCV patients were statistically compared in odds ratio and p-value was calculated by using Pearson's chi square test. Also, the mean and standard deviation of the age in IPCV patients and AMD patients were statistically calculated and compared by using two-sample T-test.

Results

Of 140 patients, 73 patients (52.14%) were male and 67 patients (47.86%) were female.

Of 140 patients, 100 patients (71.43%) were IPCV with a 95% confidence interval of 63.19% to 78.74% , 16 patients (11.43%) were idiopathic CNVs including classic and occult forms, 12 patients (8.57%) were AMDs, 6 patients (4.29%) were Chronic central serous retinopathy (Chronic CSR), 2 patients (1.43%) were serous pigment epithelial detachment (serous PED), 2 patients (1.43%) were macroaneurysm, one patient (0.71%) was retinal angiomatous proliferation (RAP) and one patient (0.71%) was retinal pigment epithelial rip (RPE rip) (Figure 1) .

Of 100 patients with IPCV, the lesions affect right eye in 44 patients (44%), affect left eye in 49 patients (49%) and affect bilaterally in 7 patients (7%). There are 107 eyes of IPCV accordingly.

Of 100 patients with IPCV, 56 patients (56%) were male and 44 patients (44%) were female. Odds ratio was 1.72 with a 95% confidence interval of 0.77 to 3.88. The difference between sexes in IPCV group was not statistically significant. (p = 0.149)

The patients with IPCV had mean ages of 61.40 (range 42 to 81, SD 8.82) while the patients with AMD in our study had mean ages of 72.08 (range 52 to 78. SD 7.23). Accordingly, the difference of these two groups was 10.68 years with a 95% confidence interval of 5.43 to 15.94. (p = 0.0001)

Of the 107 eyes with IPCV, 79 eyes (73.83%, 95%CI 65.37%-82.30%) were found in macular area, 14 eyes (13.08%, 95%CI 6.59%-19.58%) were found temporal to vascular arcades, 10 eyes (9.35%, 95%CI 3.74%-14.95%) were found in the peripapillary area and 4 eyes (3.74%, 95%CI 0.09%-7.39%) were found both in macular and peripapillary region (Figure 2).

The clinical manifestations of IPCV at presentation was subretinal hemorrhage 95 eyes (88.79%, 95%CI 81.42%-93.47%), followed by pigment epithelial detachment/orange nodules including sub-RPE hemorrhage 76 eyes (71.03%, 95%CI 61.34%-79.19%), subretinal fluid 60 eyes (56.07%, 95%CI 46.62%-65.10%), hard exudate 20 eyes (18.69%), RPE alteration 20 eyes (18.69%), intra-retinal hemorrhage 16 eyes (14.95%), retinal atrophy/scar/fibrosis 7 eyes (6.54%), vitreous hemorrhage 5 eyes (4.67%), soft drusen 4 eyes (3.74%) and subretinal fibrin 3 eyes (2.8%) (Figure 3).

By ICGA findings in IPCV patients we found that 90 eyes (84.11%) had both branching venous network and

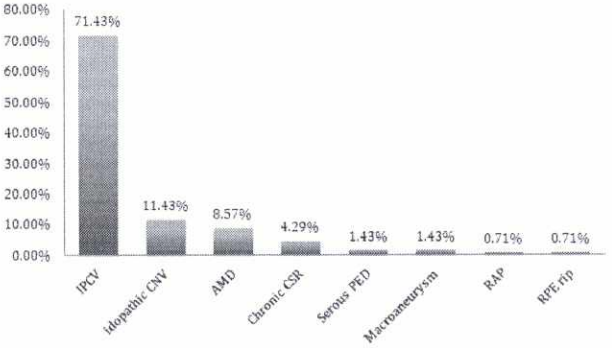

Figure 1 shows the percentage of patients in each disease (%)

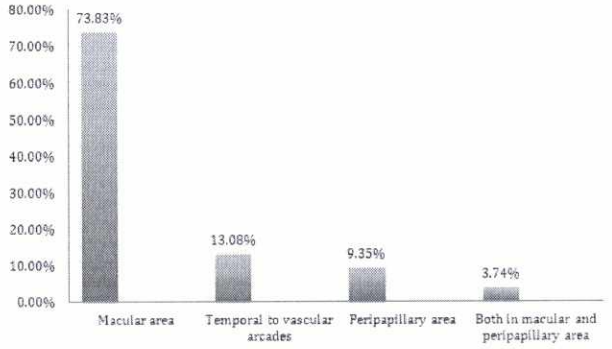

Figure 2 shows the location of IPCV (%)

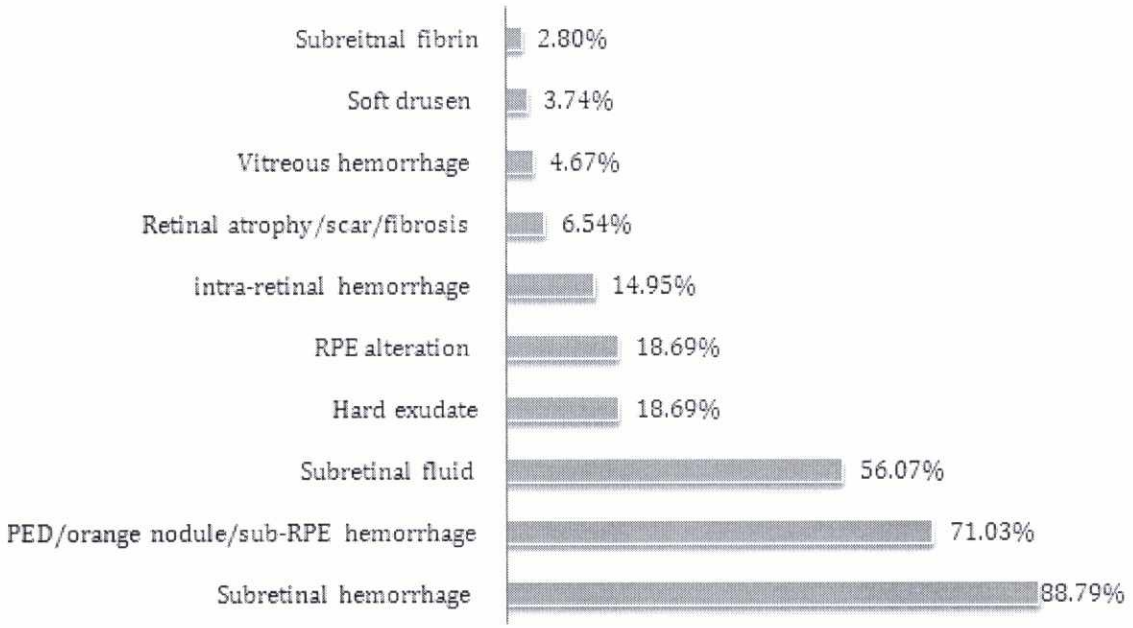

Figure 3 shows the clinical manifestations of IPCV at presentation (%)

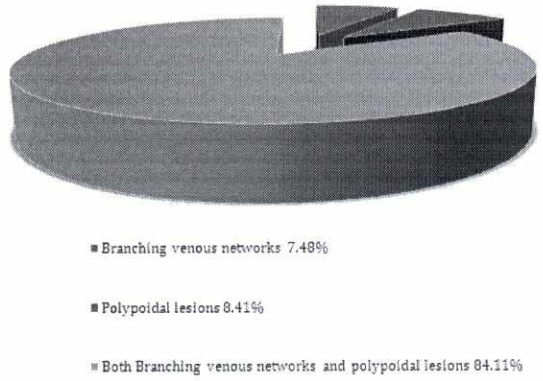

Figure 4 shows angiographic findings of IPCV (%)

polypoidal lesions, 9 eyes (8.41%) had only polypoidal lesions and 8 eyes (7.48%) had only branching venous network (Figure 4).

Some of the IPCV patients were treated with photodynamic therapy (PDT) with verteporfin. Others received anti-VEGF intravitreally before PDT was performed. The anti-VEGF used were Ranibizumab and Bevacizumab.

Discussion

IPCV is still one of the common exudative maculopathies which disturb visions substantially. If left untreated, the prognosis is still not well.[9,10,39]

In previous studies, IPCV was common in patients between the ages of 50 and 65 years.[17,40-49] In our study we also found the wider range with the mean age of 61.40 and patients with IPCV were statistically younger than those with AMDs. However, the age is not the diagnosis criteria.

Imamura Y., et al. and Ciardella A., et al. found that IPCV occurs in women as the old alternative term "multiple recurrent retinal pigment epithelial detachments in black women".[17,42] According to previous studies, IPCV is found in African-American and Caucasian women more than men.[17,41,42] However in Asian populations IPCV occurs more often in men than women.[43-49] Nevertheless, in our study, the difference between sex in IPCV group was not statistically significant.

Along with other studies, Asian descents are at risk for developing IPCV.[44-52] Moreover, IPCV was also commonly African-American descents.[53] However, there are several studies also indicated that IPCV can be found in white patients.[17,40,54-59]

About the location of the lesion, contrary to our study, several previous studies have shown that IPCV are common in the peripapillary area.[2,12,13,17,40,42,43,56,60-66]

However, in our study IPCV was most commonly found in the macular area (73.83%), followed by temporal to vascular arcades (13.08%), in the peripapillary area (9.35%) and both in macular and peripapillary region (3.74%) respectively. We also noticed that this might be because of the ethnicity but further future studies about this association are necessary to be done.

One of the limitations is that the number of populations is probably small. Moreover, our study is hospital-based, not population-based. All these limitations might lead to inaccurate prevalence number. However, to the best knowledge of the authors, this is the only article that states the prevalence of IPCV in Thailand.

Conclusion

In conclusion, from our multicenter retrospective study, the prevalence of IPCV was the most common in Thai population. In our study, IPCV is the most common macular disease in Thai population with submacular lesion. Clinical presentation of IPCV in Thai population is similar to other Asian studies in the aspect of age, area of involvement and clinical manifestations but not the gender. Moreover, IPCV in Thai population is mostly found in macular area unilaterally. Angiographically, IPCV patients have both branching venous network and polyps.

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