

RETINOPATHY: VARIABLE CLINICAL SPECTRUM AND POST-ENDARTERECTOMY CHANGES

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Background: The Carotid Artery Insufficiency Retinopathy (CAIR) is an uncommon sign of carotid artery obstruction. It is mainly found in patients with complete occlusion or severe obstruction of Internal Carotid Artery (ICA). Retinopathy is caused by progressive and chronic hypoxia to ocular tissues. The purpose of the study is to describe the variable presentation of CAIR in patients with internal carotid artery stenosis and to assess the resolution of retinopathy in patients who had carotid endarterectomy. **Methods:** Records of the patients with confirmed internal carotid artery stenosis were reviewed. Patients' demographic data and way of presentation to ophthalmologist was recorded. Associated systemic vascular diseases were also recorded on the proforma. Records of the patients with confirmed internal carotid artery stenosis were reviewed. **Results:** Thirteen eyes of 10 patients were included in study with male to female ratio of 9:1. Patients' clinical presentation ranged from scattered blot haemorrhages to ocular ischemic syndrome. Patients presented with retinopathy at different stages. The presentation of retinopathy varied from scattered blot haemorrhages to ocular ischemic syndrome. Endarterectomy resolved CAIR in 2 out of 3 patients, with one patient having bilateral resolution. **Conclusion:** CAIR should be suspected if retinopathy is unilateral. On the other hand patients with asymptomatic Carotid artery stenosis should be examined for signs of ocular ischemia. All patients with CAIR should be investigated for cardiovascular diseases. Endarterectomy in selected patients can resolve CAIR.

Keywords: Carotid artery insufficiency retinopathy (CAIR), Internal carotid artery (ICA), Retinopathy

INTRODUCTION

The term Carotid Artery Insufficiency Retinopathy (CAIR) is more accurate than previously used Venous Stasis Retinopathy. Other terms used for this condition are ocular ischemic syndrome, hypoperfusion retinopathy, hypotensive retinopathy and ischemic coagulopathy.

The CAIR is an uncommon sign of carotid artery obstruction.¹ It is mainly found in patients with complete occlusion or severe obstruction of internal carotid artery (ICA). Retinopathy is caused by progressive and chronic hypoxia to ocular tissues.^{2,3} The Clinical picture varies from retinal haemorrhages, venous dilatation and tortuosity, macular oedema and eventual neovascular proliferation. Retinal findings may be similar to diabetic retinopathy except that they are unilateral. Retinal arteriovenous communications proximal to extensive areas of complete vascular closure were reported by Bolling *et al.*⁴ Fluorescein angiography commonly reveals delayed choroidal and retinal filing, while electroretinography generally demonstrates a reduction in the amplitude of a- and b- waves.⁵

MATERIAL AND METHODS

We reviewed the ophthalmic records of the patients with confirmed internal carotid artery stenosis. Patient's demographic data and way of presentation

to the ophthalmologist was recorded. We also recorded the associated systemic vascular diseases. We also looked at resolution of retinopathy in patients who had carotid endarterectomy.

RESULTS

Thirteen eyes of 10 patients were included in this study. We had 9 male patients and 1 female patient with male to female ratio of 9:1. Mean age of all patients was 69.6±8.3 years. All patients of CAIR presented with retinopathy at different stages. The presentation of retinopathy varied from scattered blot haemorrhages to ocular ischemic syndrome. Initial diagnoses made were retinal branch arterial occlusion (BRAO, 2), arteriolar emboli (2), atypical central retinal vein occlusion (2), suspected orbital mass (1), amaurosis fugax (1), coincidental blot haemorrhages (3). Ocular ischemic syndrome (OIS) was suspected in one of the eyes presenting with sudden visual loss due to BRAO. Four out of 10 patients were asymptomatic. Best corrected visual acuity was 6/18 or better in 11 eyes. The 2 eyes with rubeosis irides had vision worse than 6/60.

Associated vascular diseases known at presentation were diabetes (70%), hypertension (80%), stroke/transient ischemic attack (50%), and ischemic heart disease (30%). Complete or severe occlusion (>70%) of the internal carotid artery was found in 70% on the ipsilateral side of the affected eye. Three patients underwent carotid endarterectomy

(2 had bilateral procedures). Out of the 3 patients who underwent endarterectomy, resolution of retinopathy was found in 3 eyes of 2 patients at 11 months follow up after endarterectomy.

DISCUSSION

Obstruction of carotid artery reduces the ocular blood supply. This reduced blood supply manifests clinically in different ways like central retinal artery occlusion, branch retinal artery occlusion, cilioretinal artery occlusion and CAIR. CAIR describes the clinical features secondary to severe carotid artery stenosis. Carotid artery stenosis causes ocular hypoxia which in turn is responsible for signs and symptoms of CAIR. Visual complaints include decrease visual acuity and history of amaurosis fugax.⁶ Visual acuity varies from normal to severely reduced. Decrease in vision could be because of hypoxia itself or arterial occlusions. Ocular or periocular pain is present in 40 per cent patients. It is usually a dull ache over eye brow and can be because of ischemia or neovascular glaucoma. The retinopathy is unilateral and ipsilateral to more severely compromised carotid artery. It includes venous dilatation without tortuosity, microaneurysms, retinal haemorrhages, arterial narrowing and neovascularization of retina and disc and macular edema.⁷ Anterior segment signs include iris neovascularization and mild iritis. Fundus fluorescein angiography (FFA) reveals delayed transit time, patchy filling of choroids and delayed arteriovenous phase.

The results of the North American Symptomatic Carotid Endarterectomy (NASCET) and European Carotid Surgery (ECST) trials have guided treatment of patients with severe symptomatic internal carotid artery (ICA) stenosis. However, trials assessing asymptomatic severe ICA stenosis have not addressed the ocular effects. In addition, the ocular ischemia seen in 100% ICA occlusion has not been addressed in trials looking at both symptomatic and asymptomatic ICA stenosis. Patients with carotid artery disease may present with ipsilateral, monocular symptoms and signs. Asymptomatic retinal emboli, transient monocular visual loss and central retinal artery occlusion can occur. CAIR is associated with severe hypoperfusion of the eye which results in progressive ischemia and this reflects severe ICA disease.

In our study associated vascular and cardiac diseases known at presentation were common. Diabetes was present in 70%, hypertension in 80%, stroke / transient ischemic attack in 50% and ischemic heart disease in 30% of the patients in our study. These systemic findings confirm the fact that affected patients usually have advanced multivessel disease and all the patients with CAIR should be screened for common vascular diseases. Diabetes is present in high

percentage of cases which makes it difficult to make the diagnosis of CAIR. The reason for this is that retinopathy signs are almost similar in both conditions. If the diabetic retinopathy is asymmetrical, then the more severely affected side can have additional CAIR. Though clinically both conditions look same but on fundus fluorescein angiography CAIR shows increased arm to eye time, patchy filling of choroids in severe cases and increased arteriovenous phase. Other FFA findings like microaneurysms and neovascularization of retina and optic disc are similar in both conditions. In our study patient's presentation was with branch arterial occlusion, arteriolar emboli, atypical central retinal vein occlusion, suspected orbital mass, amaurosis fugax and coincidental blot haemorrhages. 40% of the patients were asymptomatic. This shows the non typical nature of presentation of this rather rare condition. CAIR is associated with severe hypoperfusion of the eye which results in progressive ischemia and this reflects severe ICA disease. The ocular signs of CAIR are so similar to diabetic retinopathy and nonischemic central retinal vein occlusion, that it becomes difficult to make clinical diagnosis.

Our study raises some important issues. The first issue is that carotid artery insufficiency retinopathy has a wide spectrum of clinical presentations, with ocular ischemic syndrome representing the end-stage of the disease. Therefore, patients with 100% ICA stenosis should be referred for ophthalmology follow up given the risk of rubeotic glaucoma. The second point is the insidious course of retinal hypoperfusion. CAIR is usually present in patients with asymptomatic ICA stenosis. The question therefore is that should asymptomatic patients found to have ocular signs of CAIR be investigated for carotid artery stenosis. The third point is the recurrently reported association between cardiovascular disease states, diabetes and CAIR. A high index of suspicion is therefore required in treating patients with retinopathy and co-existing systemic cardiovascular diseases.⁸ Early diagnosis increases the chance of success of carotid endarterectomy and therefore the reversibility of early retinopathy.⁹ Finally, the fourth point is that mortality and morbidity related to asymptomatic ICA stenosis has been deemed low^{10,11} enough not to justify surgery. However ocular morbidity was not considered in these studies.

CONCLUSION

We think ocular morbidity should and be studied prospectively in patients with severe asymptomatic ICA stenosis. By such prospective study we will be able to know that will this be justifiable to do endarterectomy to stop ocular ischemia from developing, which in turn will stop CAIR.

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