Purtcher's Like Retinopathy in a Patient with Malignant Hypertension

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Abstract: Malignant arterial hypertension is a serious cardiovascular disorder with fatal consequences if untreated. Ocular morbidity includes severe visual impairment and even blindness. A case of malignant hypertension is reported, which had features of purtcher's retinopathy. The Keith Wagner Barker classification is used to describe the severity of hypertensive retinopathy changes in the retina. This classification may be inadequate in the context of malignant hypertension where the systemic hypertension is due to a secondary cause.

Keywords: Purtcher's like retinopathy, malignant hypertension, cotton wool spots.

CASE REPORT

A 28-year-old white male was referred by his general practioner, to the emergency eye department with a history of headache and visual disturbances of 1-2 weeks duration. He was reported as being in good general health with no abnormality of note. He had a significant previous medical history of migraine, but denied any ocular problems. At presentation his visual acuity was 6/6 right eye and Counting Fingers in the left eye. His anterior segment examination and intraocular pressure was normal in both eyes by applanation tonometry. On dilated fundus examination, he had multiple cotton wool spots with few scattered superficial and deep retinal haemorrhage's clustered around the posterior pole mimicking a purtcher's like retinopathy (Figure **1** & **2**). He denied any form of trauma. The retinal veins were full, and there were no AV crossing changes. The optic nerve head was pink, with clear margins and no oedema. Physical examination was normal. Although his general practioner had referred the patient with normal blood pressure recording, this was rechecked on two occasions and was found to be elevated at 210/120 mm Hg. A diagnosis of malignant hypertension was made and the patient was referred to the physicians for urgent management of his systemic hypertension. A regime of oral atenolol, nefedipine and labetolol infusion resulted in stabilization of his blood pressure over a period of one week. Systemic investigations for urinary catecholamine was negative but serum creatinine was elevated and his urine showed 3+proteins. There was cardiomegaly in his chest X-ray and ECG was consistent with left ventricular strain.

Figure 1: Right Eye multiple cotton wool spots and retinal haemorrhages around posterior pole. Note absence of disc oedema.

Figure 2: Left Eye posterior pole showing similar appearance with absence of optic disc oedema.

Renal biopsy was undertaken, and it showed evidence of IgA nephropathy, confirming the secondary nature of his systemic hypertension.

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DISCUSSION

Purtcher's retinopathy was first described in the earlier part of 20th century as a syndrome of blindness associated with head and chest trauma. Subsequently a diverse range of systemic diseases, have been reported to produce retinal appearance resembling purtcher's retinopathy. These include systemic vasculitis [1], fat [2] & amniotic fluid embolization [3] and acute pancreatitis [4]. Although our patient had a fundus appearance of purtcher's like retinopathy, there was no known systemic or local cause to explain his retinal signs. Fundus appearance of chronic hypertensive retinopathy like AV crossing changes or signs of malignant hypertension like optic disc oedema or macular star was absent. Systemic hypertension as a cause of his retinal signs was not suspected immediately, which was complicated by the fact that his primary care physician had reported that to be normal on referral. In this context, it is important to point out, that the Keith-Wagner-Barker classification (Table **1**) was described in relation to the course and prognosis of essential hypertension. The retinal manifestations in grade 1&2 of this classification are as a result of sustained and long standing chronic arterial hypertension whereas grade 3&4 hypertension is caused by acute hypertension. Grade 3 patients have high and sustained hypertension and in addition have an associated impairment of cardiac and renal function. Grade 4 hypertension, also called as malignant hypertension can develop either in a patient with known chronic hypertension, or more commonly secondary to renal parenchymal damage. Optic disc oedema is an important feature in the ophthalmoscopic diagnosis, of this stage, in addition to Cotton wool spots, retinal haemorrhages and hard exudates. In the absence of disc oedema, hard exudates and AV crossing changes one would not suspect malignant hypertension as a cause of the retinal finding leading to a delay in the diagnosis and prompt management. It would appear from this case that absence of optic disc oedema, probably does not rule out malignant hypertension.

Brown *et al.* showed that even a single cotton wool spot in an otherwise normal fundus, indicates a systemic disease like systemic hypertension [5]. Although cotton wool spots may be associated with many conditions, it should definitely alert the clinician to check the patient's blood pressure, and to recheck if reported as being normal previously.

Fluorescein angiography has shown generalized delayed filling of the choroidal vascular bed especially in the macular and Foveal region. This may explain the poor initial vision at presentation in the left eye of our patient. The acute choroidal vascular insufficiency can lead to focal infarction of the overlying RPE, manifesting clinically as pale, white, punctate lesions.

Hayreh *et al.* [6] describe focal intraretinal periarteriolar transudates as a specific retinal lesion in malignant hypertension. These are focal leakages from the pre capillary arterioles that lie in the deeper layers of the retina. Cotton wool spots develop late in the disease and are thought to be due to local ischaemia. The nature of the white patch (cotton wool spots) seen clinically is due to, disruption of the orthograde and retrograde flow of axoplasmic substances, with accumulation of debris from cell organelles.

Systemic arterial Hypertension is a serious public health problem in the developed world and ranks as the fourth largest mortality risk factor, accounting for nearly 6% of all deaths [8]. Survey in U.S.A have shown nearly 30% of hypertensive patients being unaware of their condition. Nearly 1% of the population with systemic hypertension will develop hypertensive urgency needing prompt treatment but not necessarily hospitalization [9]. Visual complaints are often the initial symptom in patients with malignant hypertension. Ophthalmologists have a great responsibility in recognizing this condition and promptly referring them for medical management to reduce the mortality and the morbidity associated with this common public health problem.

Table 1:

Keith-Wagner-Barker classification:
Grade 1 Retinal vessels are narrowed and straightened.
Grade 2 Retinal arteries begin to indent/nick the retinal veins.
Grade 3 Cotton Wool Spots, retinal haemorrhages, lipid exudates
Grade 4 grade 3 changes plus optic disc swelling

CONCLUSION

Malignant hypertension can present without retinal findings of a optic disc oedema or macular star. Serial blood pressure measurement is of paramount importance in evaluating patients with extensive cotton wool spots and a retinal picture mimicking purtcher's like retinopathy. The Keith Wagner Barker classification though useful, should not be unduly relied upon in evaluating patients with acute systemic hypertension and may be misleading in the context of malignant hypertension. To the best of our knowledge this is the first reported case of systemic hypertension as one of the causes of purtchers like retinopathy.

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